سی امین کنگره سالانه انجمن جراحان کودکان ایران

همزمان با چهل و چهارمین کنگره سالانه جامعه جراحان ایران

برنامه و خلاصه مقالات

23 – 27 اردیبهشت 1402

Program & Abstract

13th – 17th May 2023

30th Annual Congress of Iranian Society of Pediatric Surgeons

44th Annual Congress of Iranian Association of Surgeons

اعضاء هیئت مدیره انجمن جراحان کودکان ایران

دکتر احمد خالق نژاد طبری	رئیس انجمن
دکتر سید جواد نصیری	دبیر انجمن
دکتر هوشنگ پورنگ	خزانه دار
دکتر فرید اسکندری	بازرس
دکتر محسن روزرخ	عضو هیئت مدیره
دکتر منصور شیخ	عضو هیئت مدیره
دکتر فریبا جهانگیری	عضو علی البدل
دکتر سید جواد سیدی	عضو علی البدل
دکتر مهرداد ایزدی	بازرس علی البدل

کمیته علمی سی امین کنگره انجمن جراحان کودکان ایران

دبیر علمی کنگره: دکتر غلامرضا ابراهیمی سراج

دکتر غلامرضا ابراهیمی سراج

دکتر نصراله استادیان

دکتر فرید اسکندری

دکتر بهار اشجعی

دکتر محمود اشرفی امینه

دکتر سعید اصلان آبادی

دکتر حسین الماسی

دکتر امید امان اللهی

دکتر محمد امید

دکتر سهیل اوصیا

دکتر احمد آهنگران

دکتر مهرداد ایزدی

دکتر داود بادبرین

دکتر سید عباس بنانی

دکتر سید سهیل بن رضوی

دکتر علی بهادر

دکتر هوشنگ پورنگ

دکتر امیر پیروز

دکتر مهران پیوسته

دکتر مسعود جمشیدی

دکتر مرجان جودی

دکتر محمد جوانمردی

دکتر فریبا جهانگیری

دکتر مهرداد حسین پور

دکتر احمد خالق نژاد طبری

دکتر حیدرعلی داوری

دکتر صلاح الدین دلشاد

دکتر محمد هادی رفیعی

دکتر محسن روزرخ

دکتر فتح الله روشن ضمیر

کمیته علمی سی امین کنگره انجمن جراحان کودکان ایران

دبیر علمی کنگره: دکتر غلامرضا ابراهیمی سراج

دکتر سید محمد علی رئیس السادات

دکتر محمود سعیدا

دکتر جواد سیدی

دکتر رضا شجاعیان

دکتر منصور شیخ

دکتر ناصر صادقیان

دکتر صادق صادقی پور

دکتر مهدی صرافی

دکتر سعید طرلان

دکتر شهنام عسکر پور

دکتر عبدالحمید عمویی

دکتر جواد غروبی

دکتر حمیدرضا فروتن

دکتر منوچهر قربانپور

دکتر مریم قوامی عادل

دکتر مهدی کلانتری

دکتر ولی اله محرابی

دکتر مرتضی محمودی

دکتر احمد محمدی پور

دکتر فرشید مقدم

دکتر مهرداد معمارزاده

دکتر منصور ملائیان

دکتر سید عبدالله موسوی

دکتر لیلی مهاجرزاده

دکتر میرنظام میرچرخچیان

دکتر علیرضا میرشمیرانی

دکتر مسعود ناظم

دکتر هدایت الله نحوی

دکتر سید جواد نصیری

دکتر رضا نظرزاده

دکتر عباس هادیپور

دکتر مهران هیرادفر

هیئت اجرایی سی امین کنگره جراحان کودکان ایران

دکتر احمد خالق نژاد طبری

دکتر سید جواد نصیری

دکتر هوشنگ پورنگ

دکتر فرید اسکندری

دکتر محسن روزرخ

دکتر منصور شیخ

دکتر فریبا جهانگیری

دکتر جواد سیدی

دکتر مهرداد ایزدی

دکتر غلامرضا ابراهیمی سراج

خانم نسیم کاظمی‌فر

In the name of God

President message

Dear colleagues and respected guests on behalf of executive board of Iranian Society of Pediatric Surgeons, it is our pleasure to welcome all of you to the Thirty Annual Congress of Iranian Society of Pediatric Surgeons, that being held at Razi Congress Center, Iran University of Medical Sciences from 13 to 17 of May 2023.

This year congress is being organized with collaboration of the Pediatric Surgery Centers, from Universities of Medical Sciences of all provinces of Iran and Pediatric Surgery Research Center of Shahid Beheshti University of Medical Sciences.

It will be a 5 days program in the subject of **"Surgery in Childhood Cancer"**. During 5 days we will have, 7 guest speakers and 6 panel discussions, and 66 oral presentations.

I should express my special thanks and welcome to our guest speaker from different University of Medical Sciences who kindly have accepted our invitation to give lecture in our Annual Congress. I also appreciate the support of Iranian Association of Surgeons that helps us to hold our annual Congress.

I wish all guest speakers, presenters and participants of congress a pleasant and memorable stay in Tehran, Iran where has 2500 years strong and rich history and enjoy their visit of capital Tehran one of the super city of the world and other cities of Iran like Isfahan half of the world and Shiraz with its ancient Persepolis and Caspian sea of north and Persian Golf of south.

I hope this year congress to be academically, educationally and socially rewarding for all our pediatric surgeons, pediatric surgery fellows and pediatric surgical nurses and other specialty from different part of Iran and other countries.

Ahmad Khaleghnejad Tabari M.D.

Professor of Pediatric Surgery & Pediatric Urology

President of Iranian Society of Pediatric Surgeons

Chairman of the Congress

بنام خداوند جان و خرد

خداوند را شاکریم که موفق شده‌ایم پس از سه سال وقفه در برنامه حضوری کنگره به دلیل پاندمی کورونا، در سال 1402 سی امین کنگره سالانه انجمن جراحان کودکان را همزمان با چهل و چهارمین کنگره سالانه جامعه جراحان ایران از تاریخ 23 لغایت 27 اردیبهشت در سالن همایش‌های رازی دانشگاه علوم پزشکی ایران داشته باشیم.

امسال مثل سال‌های قبل با همراهی و همکاری خوب همه همکاران جراحی کودکان از سراسر دانشگاه‌ها در موضوع انتخابی کنگره شاهد استقبال پر شور همه همکاران جراحی کودکان و سایر رشته‌های مرتبط با جراحی در سرطان‌های کودکان بودیم.

جای تشکر دارد که همکاران جراح کودکان و سایر رشته‌ها و پرستاران کودکان، مقالات و گزارشات موارد جالب خود را در زمان تعیین شده بصورت الکترونیک از طریق ایمیل ارسال نموده‌اند و این موضوع در سال جاری بصورت صد در صد مجازی انجام شده و این امر رسیدن انجمن به هدف نهایی را تأمین نموده است. جا دارد در سال‌های آینده جهت یکسان سازی پذیرش مقالات برنامه نرم افزاری آن را فعال نماییم تا فرمت آن برای همه مقالات در کتابچه چاپی و الکترونیک یکسان گردد.

در کنگره امسال هفت سخنران مهمان و شش میزگرد در رابطه جراحی در سرطان‌های کودکان داریم و سعی شده از افراد با تجربه، همکاران جراحان کودکان علی الخصوص همکاران جوان و سایر رشته‌های مربوطه در میزگردها استفاده شود.

از مجموعه نزدیک به 120 جراح کودکان فعال تعداد 66 مقاله و گزارش موردی ارسال شده که نشانه تلاش عالمانه شما همکاران عزیز در کشور است و امید داریم دوستان در اسرع وقت، این خلاصه مقالات را با تکمیل داده‌های آن به صورت مقاله تکمیل شده برای چاپ در مجله رسمی انجمن ارسال نمایید. این اقدام شما کمک می‌کند تا مجله انجمن هم بصورت مرتب چاپ تا در کوتاه‌ترین مدت بتوانیم در سایت‌های علمی داخل و خارج نمایه گردد. لازم می‌دانم از تلاش‌های هیئت مدیره انجمن و کمیته علمی و کمیته اجرایی کنگره و بخصوص دبیر علمی کنگره جناب آقای دکتر غلامرضا ابراهیمی سراج و سرکار خانم ها کاظمی‌فر و قنبری که در برنامه‌ریزی کنگره تلاش نموده‌اند، تشکر نماییم و از مرکز تحقیقات جراحی کودکان دانشگاه علوم پزشکی شهید بهشتی و جامعه جراحان ایران که در امر برگزاری کنگره ما را پشتیبانی و همراهی نموده‌اند، قدردانی نماییم.

امید است در این پنج روز کنگره با مطالب علمی آمورنده در رابطه با موضوع کنگره و ارائه آن توسط همکاران جراح کودکان از سراسر کشور و تبادل اطلاعات در زمینه *"جراحی در سرطان‌های کودکان"* بتوانیم راهنمای بالینی واحدی را برای کودکانی که نیازمند عمل جراحی هستند، تدوین و به مسئولین سلامت کشور ارایه نمائیم.

امید داریم همکاران با حضور فعال خود انجمن را در راه رسیدن به اهداف بلند مدت آن یاری نمایند.

دکتر احمد خالق نژاد طبری
رئیس انجمن جراحان کودکان ایران
رئیس سی امین کنگره جراحی کودکان ایران

Dear Colleagues,

It is a great pleasure that I have the chance to invite you to the 30th **Annual Congress of the Iranian Society of Pediatric Surgeons and 44th annual congress of Iranian Association of surgeons** in Tehran, Iran.

Focus of the current Congress will be on clinical and experimental research regarding **"Surgery in Childhood Cancer**". There are going to be helpful speeches and presentations emphasizing the future of our field. Scientific sessions will be complemented by numerous special lectures and expert sessions. It will be a five days program in the subject of Surgery in Childhood Cancer. During these days we will have Seven guest speakers, Six panel discussions, and 66 oral presentations. The exponential output of knowledge and data in our field requires communication between colleges and researchers from all subspecialties of pediatric surgery. **Annual Congress of the Iranian Society of Pediatric Surgeons will** provide again an excellent communication platform during the scientific sessions and social activities of the congress. We are very grateful to our congress president and executive board for arranging this society. Let us enjoy the promising program together with colleagues and friends from all over the Iran. I am looking very much forward to seeing you in this congress.

Scientific Secretary
Gholamreza Ebrahimisaraj M.D.

همکاران گرامی

بسیار مفتخرم که شما بزرگواران را به سی امین کنگره سالانه انجمن جراحان کودکان ایران، همزمان با چهل و چهارمین کنگره سالانه جامعه جراحان ایران دعوت نمایم.

در این کنگره موضوع مورد بحث درباره **" جراحی در سرطان‌های کودکان"** خواهد بود و در این راستا تلاش خود را متمرکز بر فراهم کردن سخنرانی‌های مفید، مقالات سودمند و میزگردهای آموزنده جهت ارتقاء هر چه بیشتر سطح علمی مان نموده‌ایم.

در طول کنگره هفت سخنرانی توسط سخنرانان مهمان، شصت و شش مقاله و شش میزگرد در رابطه با **جراحی در سرطان‌های کودکان** ارائه خواهند گردید.

بهترین راه برای رسیدن به هدف والای دانش افزایی در حیطه کاری ما، ارتباط با سایر همکاران، صاحب نظران و تبادل دانسته‌ها و تجربیات در این زمینه است. به منظور رسیدن به این هدف با ارزش، انجمن جراحان کودکان ایران با فراهم نمودن جلسات علمی ماهانه و کنگره‌های سالانه جهت ایجاد برنامه‌ای مدون و جامع، ما را برای بالا بردن تجربیات علمی و عملی یاری نموده است.

در این جا از تلاش‌های اعضای هیئت مدیره و رئیس انجمن جراحان کودکان ایران که در برنامه‌ریزی کنگره مرا همراهی نموده‌اند، تشکر نموده و ساعات خوش و مفیدی را در کنار سایر همکاران، از سراسر ایران عزیز آرزومندم.

دکتر غلامرضا ابراهیمی سراج
دبیر علمی سی امین کنگره سالانه انجمن جراحان کودکان

برنامه کنگره

روز اول: شنبه 1402/2/23

تاریخ	ساعت	سخنران	موضوع
1402/2/23	8 الی 13	سالن عمومی جامعه جراحان	سالن عمومی جامعه جراحان
1402/2/23	8 الی 10		مراسم افتتاحیه و سپاس
1402/2/23	10 الی 10:30		سخنران مدعو
1402/2/23	10:30 الی 11		استراحت
1402/2/23	11 الی 13		میزگرد عمومی جامعه جراحان
1402/2/23	13 الی 14		ناهار و نماز
1402/2/23	14 الی 15:30	سالن انجمن جراحی کودکان	سالن انجمن جراحی کودکان
1402/2/23	14 الی 14:15	دکتر غلامرضا ابراهیمی سراج	خیرمقدم
1402/2/23	14:15 الی 14:30	دکتر احمد خالق نژاد طبری	سخنرانی ریاست انجمن
1402/2/23	14:30 تا 15:30		مجمع انجمن جراحی کودکان ایران

روز دوم: یکشنبه 1402/2/24

تاریخ	ساعت	سخنران	موضوع	صفحه
/2/24 1402	7:55 الی 8:00		تلاوت قران	
/2/24 1402	8:00 الی 10:30	هیات رییسه: دکتر ولی اله محرابی، دکتر حسین شفایی، دکتر محمود اشرفی امینه، دکتر هوشنگ پورنگ	جلسه علمی اول	
/2/24 1402	8:00 الی 8:15	دکتر محمد رضا طلوع	تاریخچه جراحی سرطان کودکان	1
/2/24 1402	8:15 الی 8:30	**سخنران مدعو: دکتر پیمان عشقی**	مبانی و اهداف طب تسکینی حمایتی در سرطان کودکان	2
/2/24 1402	8:30 الی 8:45	دکتر علیرضا میرشمیرانی	بررسی ندول‌های تیرویید در یک مرکز	3
/2/24 1402	8:45 الی 9:00	دکتر سعید اصلان آبادی	مورد هامارتوم مادرزادی ژنیتال	4
/2/24 1402	9 الی 9:15	دکتر سعید اصلان آبادی	توده اینترالومینال ایلئوم ترمینال	5
/2/24	9:15 الی 9:25	دکتر مهدی پرویزی مشهدی	معرفی یک مورد انسولینوما در نوجوان با تضاهر تشنج	6

1402				
/2/24 1402	9:25 الی 9:35	دکتر مهدی پرویزی مشهدی	بررسی SLNB در تومور ویلمز	7
/2/24 1402	9:35 الی 9:45	دکتر مهدی پرویزی مشهدی	معرفی یک مورد تومور ویلمز اکسترارنال	8
/2/24 1402	9:45 الی 10	دکتر منصور ملاییان	معرفی یک مورد تومور ویلمز با تهاجم عروقی	9
/2/24 1402	10 الی 10:15	دکتر سعید اصلان آبادی	بررسی غربالگری در تومور ویلمز	10
/2/24 1402	10:15 الی 10:30	دکتر جواد غروبی	بررسی بیست ساله تومور ویلمز در بیمارستان مفید	11
/2/24 1402	10:30 الی 11		استراحت	
/2/24 1402	11:00 الی 13:00	هیات رییسه: دکتر علیرضا میرشمیرانی، دکتر منصور ملاییان، دکتر مهران پیوسته، دکتر فرید اسکندری	جلسه علمی دوم	
/2/24 1402	11 الی 11:15	دکتر لیلی مهاجرزاده	معرفی یک مورد تومور ویلمز با درگیری قلب	12
/2/24 1402	11:15 الی 11:30	دکتر فریبا جهانگیری	تظاهر اورژانس در مزوبلاستیک نفروما، گزارش یک مورد نادر	13

/2/24 1402	11:30 الی 11:45	دکتر داوود بادبرین	مقایسه پروتکل درمانی NWTS و SIOP در تومور ویلمز	14
/2/24 1402	11:45 الی 12	دکتر امیرحسین لادن	معرفی یک مورد تومور ویلمز با گسترش عروقی	15
/2/24 1402	12 الی 12:15	دکتر داوود بادبرین	معرفی یک مورد تومور ویلمز دوطرفه	16
/2/24 1402	12:15 الی 12:30	دکتر آزیتا پرویزی‌زاده	معرفی یک مورد مزونفروبلاستوما در کودک 4 ماهه	17
/2/24 1402	12:30 الی 12:45	دکتر امیرحسین لادن	بررسی متون و آخرین تغییرات پروتکل‌های درمان تومور ویلمز	18
/2/24 1402	12:45 الی 13	دکتر فاطمه ملک	معرفی یک مورد تومور ویلمز با متاستازهای گسترده	19
/2/24 1402	13:00 الی 14		ناهار و نماز	
/2/24 1402	14 الی 16	گرداننده: دکتر سعید اصلان آبادی اعضا: دکتر حمید رضا فروتن، دکتر مهران پیوسته، دکتر داوود بادبرین، دکتر سید مهدی آل حسین، دکتر فاطمه ملک، دکتر بهار معینی، دکتر نفیسه مرتضوی	میزگرد تومور ویلمز	

روز سوم: دوشنبه 1402/2/25

تاریخ	ساعت	سخنران	موضوع	صفحه
/2/25 1402	7:55 الی 8:00		تلاوت قران	
/2/25 1402	8:00 لی 10:30	هیات رییسه: دکتر صلاح الدین دلشاد، دکتر جواد غروبی، دکتر محمد هادی رفیعی، دکتر احمد آهنگران	جلسه علمی سوم	
/2/25 1402	8:00 الی 8:15	**سخنران مدعو: دکتر احسان مرادی**	تومورهای سیستم عصبی در کودکان	20
/2/25 1402	8:15 الی 8:25	دکتر بهار اشجعی	مقایسه نتایج FNA و بیوپسی اکسزیونال در لنفادنوپاتی کودکان	21
/2/25 1402	8:25 الی 8:35	دکتر بهار اشجعی	مقایسه نتایج وج بیوپسی و بیوپسی اکسزیونال در لنفادنوپاتی کودکان	22
/2/25 1402	8:35 الی 8:45	دکتر بهار اشجعی	بررسی نتایج پاتولوژی توده‌های شکمی در کودکان	23

24	تازه های مدیریت تومورهای سالید کودکان	دکتر فریبا جهانگیری	8:45 الی 9	**/2/25 1402**
25	معرفی یک مورد نوروبلاستوم با تهاجم نخاعی	دکتر منصور ملاییان	9 الی 9:15	**/2/25 1402**
26	بررسی جراحی در نوروبلاستوم stage چهار	دکتر سعید اصلان آبادی	9:15 الی 9:30	**/2/25 1402**
27	معرفی یک مورد تظاهر نوروبلاستوم به صورت توده گردن	دکتر محسن روزرخ	9:30 الی 9:45	**/2/25 1402**
28	معرفی یک مورد نوروبلاستوم آدرنال راست	دکتر امید امان الهی	9:45 الی 10	**/2/25 1402**
29	معرفی یک مورد نوروبلاستوم در شیرخوار 6 ماهه با پاتولوژِی متفاوت قبل و بعد از جراحی	دکتر سعیده مجیدی	10 الی 10:15	**/2/25 1402**
30	معرفی یک مورد نوروبلاستوم با درگیری توراکوابدومن	دکتر محمود خوش خبر	10:15 الی 10:30	**/2/25 1402**
	استراحت		10:30 الی 11	**/2/25 1402**
	میزگرد نوروبلاستوما	گرداننده: دکتر محسن روزرخ اعضا: دکتر سید جواد نصیری، دکتر محمد رضا طلوع، دکتر بهار اشجعی، دکتر محمد فرانوش، دکتر میترا قالیبافیان، دکتر میترا خلیلی، دکتر یلدا نیلی پور	11 الی 13	**/2/25 1402**

/2/25 1402	13 الی 14		ناهار و نماز	
/2/25 1402	14:00 الی 16:15	هیات رییسه: دکتر حیدرعلی داوری، دکتر جواد سیدی، دکتر امید امان الهی، دکتر سید عبداله موسوی	جلسه علمی چهارم	
/2/25 1402	14:00 الی 14:15	**سخنران مدعو: دکتر علی جعفریان**	پیوند در سرطان‌های کودکان	31
/2/25 1402	14:15 الی 14:30	**سخنران مدعو: دکتر آرش ملکی**	تومورهای استخوانی	32
/2/25 1402	14:30 الی 14:45	دکتر هادی متقی پیشه	معرفی تظاهرات ناشایع سرطان در کودکان	33
/2/25 1402	14:45 الی 15	دکتر عبداله موسوی	بررسی یک مورد تومور فیبروبلاستیک التهابی با علایم ادراری	34
/2/25 1402	15 الی 15:15	دکتر امین حاج اسماعیلی	معرفی یک مورد بازیدیوبلومایکوزیس	35
/2/25 1402	15:15 الی 15:30	دکتر نورا بیگدلی	معرفی یک مورد کارسینوم برونکوژنیک ریه	36
/2/25 1402	15:30 الی 15:45	دکتر نورا بیگدلی	معرفی یک مورد تومور بیضه	37
/2/25	15:45 الی 16:00	دکتر محمد خضروی	یک تظاهر ناشایع لنفوم بورکیت	38

1402				
/2/25 **1402**	16 الی 16:15	دکتر آرین کریمی روزبهانی	بررسی اثربخشی کموامبولیزاسیون در تومورهای رتروپریتوئن کودکان	39
/2/25 **1402**	16:15 الی 16:30	دکتر آرین کریمی روزبهانی	معرفی یک مورد عفونت سایت جراحی استئوسارکوم با دیفتری	40

روز چهارم: سه شنبه 1402/2/26

تاریخ	ساعت	سخنران	موضوع	صفحه
/2/26 1402	7:55 الی 8:00		تلاوت قران	
/2/26 1402	8:00 الی 10:30	هیات رییسه: دکتر مهرداد ایزدی، دکتر شمس الدین عاملی، دکتر عباس هادی پور، دکتر صادق صادقی پور	جلسه علمی پنجم	
/2/26 1402	8:00 الی 8:15	دکتر محمد حسین سلطانی	لاپاراسکوپی توده های آدرنال در کودکان	41
/2/26 1402	8:15 الی 8:30	دکتر احمد خالق نژاد طبری	بررسی نتایج پورت گذاری در کودکان	42
/2/26 1402	8:30 الی 8:45	دکتر محسن روزرخ	بررسی مزایای تعبیه پورت زیر فاشیای پکتورال	43
/2/26 1402	8:45 الی 9	دکتر مریم پناهی	بررسی 3 ساله دسترسی‌های عروقی در بیمارستان کودکان مفید با استفاده از سونوگرافی عروق گردن	44
/2/26 1402	9 الی 9:15	دکتر محمد خضروی	بررسی تعیین محل نوک کاتتر در کودکان	45
/2/26	9:15 الی 10:45	گرداننده: دکتر ناصر صادقیان	دسترسی عروقی در سرطان کودکان	

1402		اعضا: دکتر احمد خالق نژاد طبری، دکتر فریبا جهانگیری، دکتر حمیدرضا حقیقت خواه، دکتر نیکی تدین، دکتر شیوا نظری		
/2/26 1402	10:45 الی 11		استراحت	
/2/26 1402	11:00 الی 13:00	هیات رییسه: دکتر شهنام عسکرپور، دکتر مسعود ناظم، دکتر امیر پیروز، دکتر حلیم بردی طعنه	جلسه علمی ششم	
/2/26 1402	11 الی 11:15	**سخنران مدعو: دکتر امیر علی حمیدیه**	کاربرد ژن درمانی در انکولوژی کودکان	46
/2/26 1402	11:15 الی 11:30	دکتر ناصر صادقیان	بررسی 10 ساله تومورهای تخمدان	47
/2/26 1402	11:30 الی 11:45	دکتر حلیم بردی طعنه	معرفی یک مورد توده مثانه در سیستیت ائوزینوفیلیک	48
/2/26 1402	11:45 الی 12	دکتر منصور ملاییان	بررسی تومور ژرم سل لگن در کودکان و نوزادان	49
/2/26 1402	12 الی 12:15	دکتر جواد غروبی	بررسی 10 ساله تومورهای ژرم سل در بیمارستان مفید	50
/2/26 1402	12:15 الی 12:30	دکتر مهدی صرافی	بررسی 10 ساله تراتوم ساکروکسیژیال در بیمارستان مفید	51

/2/26 1402	12:30 الی 12:45	دکتر عادل منصوری	معرفی یک مورد تومور ساکروکسیژیال با تظاهر بی‌قراری در شیرخوارگی	52
/2/26 1402	12:45 الی 13	دکتر هدی ایلخانی پاک	معرفی یک مورد تومور ژرم سل ریه در کودک 3 ساله	53
/2/26 1402	13:00 الی 14		ناهار و نماز	
/2/26 1402	14 تا 15:45	گرداننده: دکتر منصور شیخ اعضا: دکتر سهیل اوصیاء، دکتر مهدی صرافی، دکتر مجتبی موسوی خوشدل، دکتر مهرزاد مهدی زاده، دکتر ثمین علوی، دکتر فاطمه محجوب	میزگرد تومورهای ژرم سل	

روز پنجم: چهارشنبه 1402/2/27

تاریخ	ساعت	سخنران	موضوع	صفحه
/2/27 1402	7:55 الی 8:00		تلاوت قران	
/2/27 1402	8:00 الی 9:15	هیات رییسه: دکتر منصور مولاییان، دکتر مسعود جمشیدی، دکتر سعید طرلان، دکتر سید محمد علی رئیس السادات	جلسه علمی هفتم	
/2/27 1402	8:00 الی 8:15	دکتر پرستو ملایی توانا	معرفی یک مورد لوسمی با تظاهر تومور نازوفارنکس	54
/2/27 1402	8:15 الی 8:20	دکتر خشایار اتقیایی	مالفورماسیون لنفاتیک شکمی	55
/2/27 1402	8:20 الی 8:30	دکتر خشایار اتقیایی	معرفی یک مورد لیپوسارکوم اندام	56
/2/27 1402	8:30 الی 8:45	دکتر احمد خالق نژاد طبری	معرفی یک مورد رابدومیوسارکوم مادرزادی شانه	57
/2/27 1402	8:45 الی 9	دکتر علی تدین	معرفی یک مورد PNST در بیمار نوروفیبروماتوز	58
/2/27	9 الی 9:15	دکتر غلامرضا ابراهیمی سراج	معرفی یک مورد کندروسارکوم بافت نرم	59

1402				
/2/27 **1402**	9:15 الی 10:45	گرداننده: دکتر حمید رضا فروتن اعضا: دکتر احمد خالق نژاد طبری، دکتر مریم قوامی عادل، دکتر مسعود جمشیدی، دکتر پرستو ملایی توانا، دکتر معین الدین صفوی	میزگرد رابدومیوسارکوم	
/2/27 **1402**	10:45 الی 11		استراحت	
/2/27 **1402**	11:00 الی 11:45	هیات رییسه: دکتر هدایت اله نحوی، دکتر مهرداد معمارزاده، دکتر رضا نظرزاده، دکتر پیروز فرهود	جلسه علمی هشتم	
/2/27 **1402**	11 الی 11:15	**سخنران مدعو: دکتر حسن ابوالقاسمی**	تجربه درمان پالیاتیو سرطان بزرگسالان و اجرایی کردن آن برای کودکان	60
/2/27 **1402**	11:15 الی 11:30	دکتر لیلی مهاجرزاده	بررسی 10 ساله هپاتوبلاستوم در بیمارستان مفید	61
/2/27 **1402**	11:30 الی 11:45	دکتر غلامرضا ابراهیمی سراج	هپاتوبلاستوم در شیرخوار 7 ماهه	62
/2/27 **1402**	11:45 الی 13:30	گرداننده: دکتر علی بهادر اعضا: دکتر مهرداد حسین پور، دکتر لیلی مهاجرزاده، دکتر حسام الدین اقلیمی، دکتر نادر ممتازمنش، دکتر زهرا قمی، دکتر مریم کاظمی اقدم	میزگرد هپاتوبلاستوم	
/2/27	13:30 الی 14		ناهار و نماز	

1402				
/2/27 1402	14:00 الی 15:30	هیات رییسه: دکتر علی بهادر، دکتر احمد خالق نژاد طبری، دکتر حمید رضا فروتن	جلسه علمی نهم	
/2/27 1402	14 الی 14:15	**سخنران مدعو: دکتر بی بی شهین شمسیان**	پیوند سلول‌های بنیادی خون‌ساز در تومورهای سالید کودکان	63
/2/27 1402	14:15 الی 14:30	دکتر مریم کاظمی اقدم	گزارش 25 ساله تومورهای سالید در بیمارستان مفید	64
/2/27 1402	14:30 الی 14:45	دکتر مهرداد حسین پور	بررسی ملانوم در کودکان	65
/2/27 1402	14:45 الی 15	دکتر علی تدین	معرفی یک مورد بازیدیوبلومایکوزیس شکمی	66
/2/27 1402	15 الی 15:15	دکتر غلامرضا ابراهیمی سراج	سخنرانی پایانی	
/2/27 1402	15:15 الی 15:30	دکتر احمد خالق نژاد طبری	سخنرانی پایانی	

History of Cancer, Ancient and Modern Methods of Healing

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Abstract

A review of the first written descriptions and cancer reports shows that ancient physicians and surgeons made gradual progress in understanding cancer. Over the past 30 years, researchers have made significant progress in understanding the biological causes of human cancers. Genetic disorders through inheritance and environment lead to new changes in cell growth control.

• With the help of significant advances in bioinformatics and molecular methods, a lot of information has been obtained which will help in early diagnosis of cancer and timely screening for some cancers helps in early diagnosis. In recent years, molecular genetic studies have explained the basis of the mechanism for the development and development of cancers. In general, scientists today have concluded that by early detection and complete removal of cancer cells before cancer spreads, they can achieve the best results.

• Cancer is the second leading cause of death in the world after cardiovascular disease. Half of men and one-third of women in the United States will experience cancer in their lifetime. Today, millions of people continue to live due to early detection and treatment of cancer.

Cancer is not a new disease.

• The earliest description of cancer dates back to 1600 BC in the ancient Egyptian civilization. At the time, although the term cancer was not used, it was described from the notes left of the eight patients with breast cancer. Egyptian doctors burned the cancer site with the help of a fire drill, although this method increased the patient's lifespan slightly, but in their notes they wrote that cancer is incurable …

Key Words: History of Cancer, Screening, Chemotherapy, Radiotherapy

Palliative and Supportive Care on Pediatric Cancer in Iran

Eshghi P, Khanali Mojen L, Rassouli M

Abstract

Cancer is the second leading cause of death among the children under the age of 14 in Iran. It is fatal in 1,037 out of every 1567 children with cancer which is a high rate in comparison with advanced countries. Due to the increasing incidence of the disease by 2020, and considering the young population of Iran (World country Index, 2015), securing health for this group is one of the most essential needs of Iran's health system.

Despite medical advances, the development of therapies, and consequently the increase in survival rates, children with cancer continue to suffer physically, psychologically, socially and spiritually from a wide range of symptoms associated with the disease and the side effects of the treatment. It affects the quality of life not only for the child, but also for the whole family as the primary care provider. In such a situation, one of the essential objectives of the health care system is to provide palliative care services , services that are required to be applied as major components of the health model is essential which can be used as a guide for the development of related activities . Therefore, considering the variety of palliative care systems of all countries and must be presented as comprehensive services in the global action plan published in 2014 according to the World Health Organization. (WHO, 2016). The Program of Action for Cancer Therapy (PACT) of the international Atomic Energy Agency and the WHO experts recommendations on the matter confirm this necessity as well. This policy is only carried out through governmental support and is very important in countries with limited resources. In Iran, health care system is managed by the governmental sector and the need to integrate this program into the health care system is well-understood by the policy-makers. This is a fact confirmed by the establishment of the Palliative Care taskforce in the Ministry of Health and Medical Education and also the draft of palliative care action.

The provision of palliative care in the world is carried out in a variety of ways. Designing it as a health care model is essential which can be used as a guide for the development of related activities. Therefore, considering the variety of palliative care models for children it is not possible to set of fixed and predetermined goals for it (Cancer Research Center, 2012) and the detection and application of the appropriate model requires considering the specific conditions of each society and consequently, the policies of the health care system of that society.

In Iran, in spite of the fact that the health policy makers pay attention to this issue and have considered it in the National Palliative Care Action Draft Plan, the classification of palliative care services and establishment of the referral system are considered as challenges, and the children and their families still have a low quality of life and their access to resources is limited (Rassouli et al., 2017). Therefore, providing a conceptual model for the provision of these services is necessary in Iran, and its applicability requires considering and applying the beneficiaries' views in which the child and the family play an important role as a care unit. On the other hand, considering the standards of palliative care in Europe, based on providing care services according to the needs of the patient and the family (the Committee of the EAPC task force, 2007), and considering satisfaction as one of the results of these services, it is necessary to identify the needs of the child and the family to provide care based on their needs. To this end, using the beneficiaries' opinions based on available resources and service delivery models can be helpful. This study aims to explain the beneficiaries' perceptions on providing a conceptual framework based on the health care service delivery system in palliative care for children with cancer in Iran.

Thyroid Nodules in Childhood: a Single Institute Experience

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Abstract

Objective:

Thyroid nodules are rare in children. Multiple diagnostic modalities are used to evaluate the thyroid mass. The aim of this study was to determine results of management of thyroid nodules in children with special attention to the role of fine needle aspiration biopsy (FNAB) in diagnosis:

Methods:

Thirty-two children who underwent surgery for thyroid nodules in Mofid Children's Hospital within 10 years (1996t0 2005) were retrospectively studied From clinical records we obtained data about demographic characteristics, clinical manifestations, ultrasonography (USG) findings, and FNAB results, pathological reports, surgical therapy and complications Data was analyzed statistically for association with thyroid cancer:

Findings:

Twenty-five patients (78.1%) were girls, and 7 (21.9%) boys: Mean age was 10.9 (range 8 to 14) years: 24 (75%) patients had benign and 8 (25%) malignant tumors 18 (56.25%) nodules were located in the right lobe. Statistical analysis revealed sensitivity, specificity, accuracy, and positive and negative predictive values as follows: 80%, 65%, 63%, 25%, and 86% for USG; 35%, 41%, 40%, 18%, and 66% for RNS; 91%, 94%, 90%, 74%, and 96% for FNAB respectively.

Conclusion:

Clinical judgment as determined by serial physical findings with USG continues to be the most important factor in the management of thyroid nodules in children: FNAB is the most accurate method of investigation and its accuracy is improved by USG guidance.

Soft Tissue Tumors Accompanying Giant Melanocytic Nevus; Case Report of a Congenital Genital Hamartoma Underlying a Giant Melanocytic Nevus

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1. Tabriz University of Medical Sciences
2. Ardabil University of Medical Sciences

Abstract

Giant melanocytic nevus (GMN) is a rare condition that defined as melanocytic lesion recognized at birth, larger than 20 cm in diameter, and they occur in about 1 per 200,000-500,000 newborns.

Melanoma represents the most common malignancy arising in the context of GMN. The lifetime risk of developing malignant melanoma in patients with GMN is 5% to 10%. Melanoma could arise at the cutaneous as well as extracutaneous sites. Other soft tissue tumors such as lipomas, schwannomas, neurofibromas and harmatomas are benign associated tumors. Whereas, malignant soft-tissue neoplasm such as liposarcoma, rhabdomyosarcoma, ganglioneuroblastoma and undifferentiated spindle cell neoplasms may also occur. Clinically, a suspicion of benign versus malignant lesion beneath the giant congenital nevus prompted its surgical removal.

We report a 4 years old girl with bathing trunk congenital GMN accompanying a inguinal and genital mass mimicking scrotum and sex differentiation disorder. The underling mass was excised and pathology examination revealed the hamartoma diagnosis. This case report highlights the role of prompt surgical excision and histopathological examination.

An Extensive Multicentric Inrtaluminal Mass of Terminal Ileum Mimicking Meckel's Diverticulum.

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2. Ardabil University of Medical Sciences

Abstract

Heterotopic gastric mucosa (HGM) can be located in various parts of the gastrointestinal tract. As a rare anomaly in the small intestine, it can become complicated by intussusception, obstruction, gastrointestinal bleeding, and even peritonitis leading to death. The present study is a case report from a 12-years-old boy who was presented with hematochezia and abdominal pain for a couple of days. Tagged Red blood cell (RBC) scan and Technetium scan revealed gastrointestinal bleeding at the lower abdomen, highly suggestive for the diagnosis of Meckel's diverticulum. Subsequently, exploratory laparotomy revealed contiguous and some scattered mucosal lesions with multiple polyps in various sizes in the terminal ileum. Meckel's diverticulum was absent and the patient was treated by resection and primary anastomosis. The resected tissue revealed extensive ectopic gastric mucosa and polypoid tissues. The patient recovered uneventfully and was discharged four days after the surgery. The symptoms did not recur within six month after his surgery. Our case demonstrated that although multiple polypoid gastric heterotopias in the terminal ileum is extremely rare, it should be considered as one of the differential diagnoses of gastrointestinal tract bleeding.

Insulinoma in a Teenager with Seizures

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Abstract

Insulinoma is an extremely rare pancreatic neuroendocrine tumor that originates from insulin- secreting islet cells, with an incidence of one to four cases per million each year. Here, we report a case of a 16-year-old boy presenting with seizures who was diagnosed with insulinoma and successfully treated with surgical resection.

Keywords*:* Insulinoma, Hypoglycemia, Surgery

Sentinel Lymph Node Biopsy in Pediatric Wilms Tumor

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Abstract

Background:

Although the sentinel lymph node Biopsy (SLNB) is well stablished in solid tumors among adults but the experience on SLNB in pediatrics is still limited. In this article we report our experience of sentinel lymph node detection that is applied on pediatric solid renal tumors.

Methods:

Twenty 1–16 year old children with non-metastatic primary Wilms tumor regarding the radiological studies were enrolled. At the time of radical nephrectomy, radio tracer injection was carried out after renal vein, artery and ureter ligation. Sentinel node detection and sampling was performed in every location with radiotracer count of 3 times more than background. Finally lymph node sampling was completed following the standard current discipline in Wilms tumor surgery.

Results:

A single SLN was detected in 16 patients. 4 patients had more than one SLN. The most common site of SLN detection was inter aortocaval space. Histopathologic studies revealed tumor involvement in 3 sentinel nodes (15%). All other lymph node samples were also studied and LN involvement was not detected in any of the cases with tumor free sentinel lymph node (no false negative case). Multiple LN involvement was reported in two patients with positive SLN in which, other involved lymph nodes were removed with the tumor during radical nephrectomy.

Discussion:

Intraoperative SLNB is a safe and feasible tool to improve the accuracy of staging in pediatric Wilms' tumor. We suggest to ligate renal artery and vein prior to radiotracer injection to diminish the background confounding effect.

Extrarenal Wilms' Tumor: Challenges in Diagnosis, Embryology, Treatment, and Prognosis

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Background:
Wilms' tumor is one of the most common childhood solid malignancies, which classically arises from primitive metanephric cells, but exceptionally it may arise in places other than kidneys. Extrarenal Wilms' tumor is a rare but challenging entity, considering its diagnosis histopathology, staging, treatment, and prognosis. Diagnosis of extrarenal Wilms' tumor is always postsurgical, which may jeopardize treatment planning and consulting with parents in the first step.

Methods:
This article is a comprehensive search in scientific major database including Medline, Scopus, EMBASE, SciELO, Cochrane Database and google scholar up to April 2022 with the key words of "Wilms tumor" and "Extra renal". All published or unpublished (pre-print or even pre-proof accepted literature in English) were included. Those articles that reported or discussed about extra renal Wilms tumor were eligible to include in this narrative review. Two researchers reviewed all selected articles to extract data about neonatal extra renal Wilms tumor.

Results:
The histopathology of Wilms' tumor is very confusing. While most authors believe that it arises from primitive ectopic nephrogenic rests, teratoid Wilms' tumor leads to the debate whether this tumor is neoplastic or embryonic. Staging of extrarenal Wilms' tumor is also a challenge when we consider the National Wilms' Tumor Study (NWTS) recommendations; all these tumors should be considered as stage II or higher as they are beyond the renal capsule. This will mandate chemotherapy for all patients while most of the reported cases have a favorable histology, and long-term tumor-free survival has been reported even with exclusive surgery in some case reports. Although treatment strategies for extrarenal Wilms' tumor are the same as those for renal Wilms' tumor, different locations and neighboring organs may invoke special considerations and scenarios while planning for surgery and adjuvant therapies. Consulting with the parents is also a problem, considering the rarity of the disease and limited publications. In this chapter, we discuss all these topics in detail after a systematic review of extrarenal Wilms' tumor cases to date in order to provide a clear perspective for confronting this rare disease.

Discussion:
ERWT is considered a rare childhood malignancy with atypical presentations. The pathogenesis of ERWT becomes clearer by the popular theory, which suggests the heterotopic metanephric blastema as the precursor of ERWT while the diagnosis, staging, and treatment remain challenging. NWTS protocols are applied for ERWTs due to the rarity of the disease and lack of systematic data. We reviewed 87 reported childhood ERWT cases and observed favorable histology in most cases, which made the prognosis good and comparable to that of classic Wilms' tumor with the same stage and histology.

Wilm's Tumor

Surgical Concepts in Cava Vein Thrombosis

Mollaeian M, Eskandari F, Sadid D, Tolou M. R

Bahrami Children's Hospital, Tehran

Abstract

The incidence of Wilm's tumors with cava thrombosis ranges from 4% to 8% within the different international trials. Tumor thrombosis extension can vary from the renal vein up to the right atrium and is classified according to the staging system of Daum this stagin system is helpful for decision- making regarding treatment intensity.

It remains unclear if primary or delayed surgery is the optimal treatment in cases of Wilm's tumor with intravascular extension. Both SIOP and VOG treatment guidelines recommend primary chemotherapy if the tumor thrombus extends into the inferior vena cava vein or higher. Emergency surgery is indicted only in much selected cases (for example, cardiac failure or respiratory insufficiency caused by lung embolism.

Advances of primary chemotherapy are shrinkage of the tumor and regression of thrombus extension. This can lead to a reduced invasiveness of surgical procedure, especially with regard to the need for cardiopulmonary bypass (CPB). Another point is the reduction in vital tumor cell components, which could be demonstrated in the NWTS-4 trial.

A relevant parameter in the judgment of thrombus extension is the level of diaphragm. Most surgeons use a CPB in cases of tumor extension above the level of the diaphragm. Lodge et al. published a surgical strategy without CPB in cases of tumor thrombus extension above the diaphragmatic level with the aim to prevent side effects of hypothermia and anticoagulation. Nevertheless, the risk of cardiac failure and uncontrolled bleeding must not be neglected. Another important aspect is the time pressure for the complete removal of the thrombus from the vessel level wall, which is impossible to determine preoperatively. In some constellations the thrombus can be easily dissected from the vessel wall, whereas in others an adherence of invasion of the vessel wall is observed. Complications rates for surgical treatment have been reported with a wide range from 0-44% including intraoperative death. Altogether, survival rates of children with intravascular tumor extension are over 85%.

How and when Wilm's Tumor should be Screened in Syndromic Patients and their Family Members?

Aslanabadi S[1], Badebarin D[1], Farhadi E[1], Hasanzadeh Ghavifekr N[2]

1. Tabriz University of Medical Sciences
2. Ardabil University of Medical Sciences

Abstract

Wilm's tumor is the most common malignant kidney tumor of childhood reaching about 6% in prevalance.

In first years of 20th century, total survival rate of Wilm's tumor in childhood was as low as 5%, but in recent years it has reachead about more than 90% in which screenig plans might play a role.

The risk of developing Wilm's tumor in general population in 1: 10000, but the incedence of Wilm's tumor in some syndromes like beikwith-wiedman syndrome is very high and may occur earlier; or in congenital anolalies either isolated or as part of special syndromes occur in about 10% of children with Wilm's tumor.

WAGR syndrome (Wilm's tumor, Genitourinary malformation, and Mental Retardaion) is a rare genetic syndrome with defects in 11p13 and children with this syndrome are at 30% higher risk of developing Wilm's tumor.

So we must have special strategy for cases with some syndromes and congenital anomalies; for example in children with hemihypertrophy or aniridia, serial sonographic studies must be done between ages 3 to 10 years old.

Long-term Outcome in Children with Wilms' Tumor; Experience of a Single Center for Two Decades

Mohajerzadeh L[1], Khaleghnejad Tbari A[1], Rouzrokh M[1], Shamsian Sh[2], **Ghoroubi J**[1], Sarafi M[1], Amonollahi O[1], Ebrahimisaraj Gh[1]

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2 Pediatric Congenital Hematologic Disorders Research Center, Research Institute for Children's Health, Shahid Beheshti University of Medical Sciences, Tehran, Iran

Abstract

Background:

Wilms' tumor (nephroblastoma) is the major renal cancer in children. Objectives: The aim of this study was to assess the individuality of Wilms' tumor and the consequences of management attained in our referral subspecialty center.

Methods:

In this study, we composed the data of children with Wilms' tumor in 2 decades; 55 cases between 1992 and 2002 and 49 patients between 2006 and 2016 were diagnosed with Wilms' tumor. Demographic characters, a form of presentation, tumor stage, related underlying disease, histopathology consequences, type of management, and the survival rates were assessed.

Results:

In the first decade, 24 patients were females and 31 were males (M/F = 1.2); in the other groups, 30 were females and 19 were males (M/F = 0.61). The mean age was 45.2 months at the time of diagnosis for the first group and the mean age was 36 months for the other group. In the first decade, the surgical stage after the operation was as follows: stage I (32.7%), stage II (16.36%), stage III (38.1%), stage IV (9%), and stage V (1.8%) who did not operate. In second decade, 49 patients were as follows: stage I (14.3%), stage II (40.8%), stage III (24.5%), stage IV (10.2%), and stage V (10.2%). In 54.5% of the first group, histology was favorable, and in 43.6% of the first group, histology was unfavorable; in the second group, 95.4% were the favorable type. The patients were managed based on protocols of the National Wilms' Tumor Study. In the first decade, relapse-free was 71% and 4-year survival rates were estimated at 86%, and in the second decade, pulmonary metastasis was observed at 28.6%, liver metastasis in 2.3%, recurrence in 5%, and 4-year survival rates were estimated at 90%.

Conclusions:

This study demonstrated development in the management of children with Wilms' tumor in recent 20 years, with comparable relapse-free and survival rates to the National Wilms' Tumor study. But with more adjustment in treatment protocols, the superior outcome will be attainable.

Keywords: Wilms' Tumor, Relapse-free Survival, Nephroblastoma

Wilms' Tumor with Tumor Thrombus Extension to the Right Atrium: with Long Term Follow-up after Cardiac Approach

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Abstract

Introduction:

Wilms' tumor is the most common kidney cancer in children, but Wilms' tumors that extend directly into the right side of the heart are very rare and at this stage, they necessitate the cardiac surgeons' intervention.

Case Presentation:

The patient was a 24 months old girl who was hospitalized due to fever and abdominal pain that had started about 2 weeks before admission. Patient's full abdominal exams revealed a large mass in the right kidney with a thrombus tumor in the entire inferior vena cava which was extended into the Right Atrium. Patient completed about 8 weeks of neoadjuvant chemotherapy. Next to chemotherapy no changes was distinguished in renal mass and Tumor Thrombus. As, the thrombosis in the atrium was loose and there was a high risk of pulmonary embolism, so emergency intervention was required. Two pediatric and cardiac teams, simultaneously performed the operation without Cardiopulmonary Bypass and cardiac arrest. Both renal mass and the thrombus tumor were entirely removed. Patient followed up for 5 years after surgery and no medical complications, tumor recurrence, or metastasis were observed throughout that period.

Conclusions:

Although surgical intervention and tumor resection leads to good prognosis, multimodality management and multidisciplinary approach gives the best consequences.

Keyword: Wilms'Tumor, Tumor Thrombus, Cardiopulmonary Bypass

Acute Peresentation of Congenital Mesoblastic Nephroma, A Case Report

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Background:

Congenital mesoblastic nephroma (CMN) is a rare pediatric renal tumor with low malignant potential that most commonly occurs early in infancy and the Complete surgical removal is curative in most cases.

Case presentation:

A mature male neonate was born, at 37 weeks of gestation, in whom an abdominal mass was detected in her last prenatal ultrasonography in favor of mesoblastic nephroma. Delivery was done by cesarean section and the baby was born in a good condition, with Apgar scores of 9. Birth weight was 3590 gm.

A palpable abdominal mass was detected during physical examination at birth and the postnatal ultrasonography showed a huge solid-cystic heterogenous mass originated from left kidney which was extended to right side of abdoman and pelvis with mild free peritoneal fluid. After admission to the NICU, the expansion of abdominal mass in association with an ongoing drop in hemoglobin level and hemodynamic instability was occurred. So after resuscitation and hemodynamic stabilization, an emergency operation was performed and massive hemoperitoneum and ruptured renal mass with ongoing bleeding was detected and left radical nephrectomy was done. After 2 weeks he was discharged.

NWTS vs SIOP

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Abstract

Wilms' tumor is the most common malignant tumors of the kidney in children .The treatment of Wilms' tumor can be considered as the paradigm for multimodal treatment of malignant solid tumors in childhood .Major research and randomized controlled trials performed by several co-operative groups have made the future of Wilms' tumor patients very bright.

The two major groups which have tremendous contributions in the management of Wilms' tumor are National Wilms' Tumor Study (NWTS) and the Societe Internationale D'oncologie Pediatrique (SIOP).

The largest numbers of patients have been studied in the several trials of both NWTS and SIOP groups though the UK group has also been active in the trials of Wilms' tumor. Both treatment approaches yield almost equivalent clinical outcomes though a valid debate continues about the relative merits of each approach.

In our country, the incidence of advanced and metastatic Wilms' tumor is considerably higher than in the western literature as the patients usually present in the late stages with large tumors and many a times with metastases.

In our circumstances, where more than basic investigations may be difficult to perform, probably it is more appropriate and feasible to follow the SIOP protocol for management of Wilms' tumor with the added advantage of preventing intra-operative tumor spillage with upfront chemotherapy.

درمان و فالوآپ تومور ویلمز متاستاتیک و Locally Advanced همراه گسترش عروقی در پسر ۴ ساله

دکتر امیرحسین لادن

استادیار جراحی کودکان دانشگاه علوم پزشکی زنجان

خلاصه:

گزارش یک مورد کودک پسر ۴ ساله با تشخیص اولیه تومور ویلمز کلیه راست با گسترش تا وریدهای کبدی و درگیری وسیع لنف نودهای پاراآئورتیک و متاستاز منفرد ریوی که ابتدا تحت کموتراپی نئوادجوانت قرار گرفت و با حذف متاستاز کبدی و کاهش درگیری وریدی به ورید IVC تحت نفرکتومی رادیکال همراه با خارج کردن ترومبوز از IVC و لنفادکتومی گسترده پاراآئورتیک قرار گرفت و فالوآپ ۱/۵ ساله بیمار.

Management of Bilateral Wilms' Tumors: Two Cases Report

Badebarin D, Aslanabadi S, Farhadi E, Hasanzade N

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Background:

Wilms' tumor remains the most common renal tumor in children (6% of all pediatric malignancies) and present as one of the most challenging tasks for pediatric surgeons as its management requires an advanced procedure. The ultimate goal in these two cases is to preserve as much renal parenchyma as possible whilst still achieving complete tumor resection.

Presentation of cases:

Here we present two bilateral Wilms' tumor cases, a 5 year's old boy (case 1) and a 7 year's old girl (case 2). This patient underwent neoadjuvant chemotherapy regimen, followed by unilateral partial nephrectomy and radical nephrectomy for other side. Adjuvant radiotherapy was performed following the surgery. Follow-up imaging 6 months afterward revealed, none of which created any problem for the case 1. In case 2 a small local recurrence reported in MRI was later performed on the 8th month after the surgery.

Results:

According to SIOP and NWTSG classification, the patients presented as stage V of the disease. The patients was on neoadjuvant chemotherapy (Regimen I) as recommended by NWTSG. This strategy was shown to be effective, as the tumor on the kidneys was reduced to less than 60-70% of the initial size. A routine follow-up using chest x-ray, abdominal ultrasonography (USG), and contrast studies such as MRI and MSCT scan, was performed in our reports.

Discussion:

From our experience, the combination of neo-adjuvant chemotherapy, renal salvage surgery and adjuvant radiotherapy is a feasible, safe and effective option for bilateral Wilms' tumor cases.

Introducing a 3.5 Month - old Infant with Mesoblastic Nephroma Tumor

Parvizizadeh A

Abstract

A 3.5-month-old infant with a right inguinal hernia accidentally reported a mass in the left kidney in the abdominal US. An image of a mass-like hypoechoic area with a maximum diameter of 40 mm was seen in the middle bridge and lower bridge of the left kidney, which is towards the perinephric space and also it has bulged into the pilocallis system. It is reported that it is more likely to be mesoblastic nephroma. In the next US that was done at Mohammad Kermanshahi, the possibility of Wilms tumor was reported for the patient. The patient underwent a CT scan, which revealed a hypodense mass with internal enhancement. The size 32×42 was seen in the lower bridge of the left kidney; it has internal vascularity and lack of calcification, and Wilms tumor was the first diagnosis. Evidence of vascular invasion was not seen; several lymph nodes were seen in the para-aorta. The patient was diagnosed with Wilms' tumor and underwent surgery. A nephrectomy with adrenal preservation and para-aortic lymphadenectomy was performed. The pathological report of classic subtype mesoblastic nephroma was without Girota involvement and reactive lymph nodes in IHC.

Ki67: positive in 5% of stromal cells

WT1: positive

CD56: negative

Literature Review and the Latest Updates in Wilm's Tumor Treatment Protocols

Ladan A

Abstract

CCN Guidelines for Wilm's Tumor focus on the screening, diagnosis, staging, treatment, and management of Wilm's tumor (WT, also known as nephroblastoma). WT is the most common primary renal tumor in children. Five-year survival is more than 90% for children with all stages of favorable histology WT who receive appropriate treatment. All patients with WT should be managed by a multidisciplinary team with experience in managing renal tumors; consulting a pediatric oncologist is strongly encouraged. Treatment of WT includes surgery, neoadjuvant or adjuvant chemotherapy, and radiation therapy (RT) if needed. Careful use of available therapies is necessary to maximize cure and minimize long-term toxicities, we are going to discuss the latest evidence based changes in approaching to patients affected by Wilm`s tumor.

A Massive Metastatic Wilm's Tumor Case

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Abstract

A 12-year-old female with a massive tumor in the left kidney was referred to our department. The initial tumor enlarged, so it was inoperable, and a biopsy under the guidance of sonography confirmed the pathology of an unfavorable Wilms tumor.

The initial tumor was 234×194×60 mm, with multiple nodules in the lung and liver.

The patient received the chemotherapy regime consisting of alternative courses of Adriamycin and Cyclophosphamide interchangeable with Carboplatin and Etoposide for nine total courses.

The CT scan of the patient showed complete resolution of liver nodules, shrinkage of kidney tumor, and decreasing number of nodules.

The patient underwent surgery for nephrectomy nine months after treatment initiation; pathology favored an unfavorable Wilms tumor.

After a delay related to patient personal matters, she received Radiotherapy for Six courses.

then received a novel regime for refractory solid tumors consisting of Temozolomide, Irinotecan

Bevacizumab and vincristine for four courses.

The CT scan showed complete tumor resolution, then FDG avid whole-body Pet scan confirmed complete resolution.

Now it is more than six months after the patient is off therapy. The patient's Biochemistry and complete cell count are within normal range, and the patient is healthy.

Keywords: Wilms Tumor, Unfavorable, Radiotherapy

A Review to Childhood Brain Tumors

Moradi E

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Abstract

Unfortunately, many subtypes of children brain tumors continue to have unfavorable long-term prognoses. Brain tumors are the most frequent solid tumors in pediatric population. High-resolution genomic, epigenetic, and transcriptomic profiling, which has provided insights for improved tumor categorization and molecularly targeted therapies, has nevertheless resulted in notable advancements in our comprehension of the molecular basis of these tumors over the past few years. While previously cancers like medulloblastomas have been divided into standard- and high-risk categories, it is now understood that these tumors comprise four or more genetic subsets with unique clinical and molecular characteristics. The same is true for high-grade glioma, which was once thought to be a single high-risk entity but is now understood to consist of several subsets of tumors that vary in terms of patient age, tumor location, prognosis and other factors. The circumstance is substantially worse, for ependymoma, at least nine subsets of tumors have been identified, making it complex. On the other hand, the majority of pilocytic astrocytomas appear to be caused by genetic alterations that affect a single, treatable molecular target. As a result, treatment in the modern era is changing from the historical standard of radiation and conventional chemotherapy to a more nuanced approach in which these modalities are applied in a framework that takes risks into account and molecularly targeted therapies are implemented to supplement or, in some cases, replace conventional therapy.

مقایسه نتایج FNA و بیوپسی در لنفادنوپاتی کودکان

دکتر بهار اشجعی

دانشیار گروه جراحی کودکان، دانشگاه علوم پزشکی تهران، بیمارستان مرکز طبی کودکان

مقدمه:

لنفادنوپاتی بر اساس تعریف به حالتی گفته می‌شود که در آن لنف نودهای طبیعی یک یا چند قسمت در بدن فرد به دلیل التهاب و یا سایر علل پاتولوژیک و بیماری بزرگتر از معمول بشود. در این مطالعه ما تصمیم گرفتیم به بررسی نتایج پاتولوژی بیوپسی‌های اکسیزیونال و گوه‌ای انجام شده و آسپیراسیون سوزنی از لنفادنوپاتی کودکان مراجعه کننده به مرکز طبی کودکان بپردازیم. به این ترتیب شاید بتوانیم به نتایجی برسیم که بر اساس اندازه و محل لنف نود از انجام بیوپسی‌های غیرضروری پرهیز کنیم و در صورت یکسان بودن نقش دو نوع بیوپسی با آسپیراسیون سوزنی در تعیین پلن درمانی، اصراری بر انجام اکسیزیون یا بیوپسی انسیزیونال در مواردی که انجام آن برای بیمار خطراتی مثل صدمه به عروق و اعصاب دارد، یا استفاده از بیهوشی جنرال دارد، نداشته باشیم.

روش انجام:

این یک مطالعه گذشته نگر می‌باشد و در آن تمام کودکانی که از شهریور سال ۱۳۹۴ تا شهریور سال ۱۳۹۹ در بیمارستان مرکز طبی کودکان تحت بیوپسی لنف نود قرار گرفته‌اند، بررسی شدند. تمام بیماران تحت آسپیراسیون سو.نی همزمان با بیوپسی قرار گرفتند. روش جمع آوری اطلاعات بر اساس مراجعه به پرونده بیماران است و در مواردی که اطلاعات پرونده ناقص بوده با تماس تلفنی با خانواده بیماران اطلاعات خواسته شده کامل گردیده است.

نتایج:

در مجموع غدد لنفاوی 55 بیمار در جمعیت کودکان در طول دوره مورد مطالعه تحت بیوپسی و آسپیراسیون قرار گرفتند (12 مورد اینسیژنال،43 مورد اکسیژنال). اکثر موارد (47 مورد؛ 85٪) خوش خیم بودند. نتایج به چهار دسته کلی به عنوان لنفادنیت واکنشی (32 مورد؛ 58 درصد)، لنفادنیت گرانولوماتوز (11 مورد؛ 20 درصد)، لنفادنیت عفونی (5 مورد؛ 9 درصد) و بدخیمی (7 مورد؛ 12/72 درصد) طبقه‌بندی شدند. 7 مورد بدخیمی شناسایی شده در این جمعیت شامل سه مورد لنفوم هوچکین (HL) و چهار لنفوم غیر هوچکین (NHL) بود. در تمامی موارد لنفادنیت واکنشی آسپیراسیون سوزنی به تشخیص پاتولوژی کمک کرد و در سایر موارد جهت تشخیص قطعی نیاز به بررسی نمونه بیوپسی بود.

نتیجه گیری:

علت اصلی LAP لنفادنیت واکنشی است. آسپیراسیون سوزنی در این بررسی فقط در تشخیص لنفادنوپاتی واکنشی کمک کننده بود و به خصوص در تشخیص بدخیمی‌ها نیاز به بررسی نمونه بیوپسی شد. این موارد تنها با بیوپسی قابل تشخیص و مدیریت هستند. بیوپسی می‌تواند نقش کلیدی در تریاژ موارد مناسب داشته باشد، به طوری که این بیماران برای تشخیص و یا ارجاع به مراکز بالاتر مورد بررسی بیشتر قرار می‌گیرند.

واژه‌های کلیدی: لنف آدنوپاتی، اطفال، بیوپسی، اکسیزیونال، گوه‌ای، آسپیراسیون سوزنی، FNA

مقایسه نتایج وج بیوپسی و بیوپسی اکسیزیونال در لنفادنوپاتی کودکان

دکتر بهار اشجعی

دانشیار گروه جراحی کودکان، دانشگاه علوم پزشکی تهران، بیمارستان مرکز طبی کودکان

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در مجموع غدد لنفاوی 55 بیمار در جمعیت کودکان در طول دوره مورد مطالعه تحت آسپیراسیون قرار گرفتند (12 مورد اینسیژنال،43 مورد اکسیژنال). اکثر موارد (47 مورد؛ 85٪) خوش خیم بودند. نتایج به چهار دسته کلی به عنوان لنفادنیت واکنشی (32 مورد؛ 58 درصد)، لنفادنیت گرانولوماتوز (11 مورد؛ 20 درصد)، لنفادنیت عفونی (5 مورد؛ 9 درصد) و بدخیمی (7 مورد؛ 12/72 درصد) طبقه بندی شدند. 7 مورد بدخیمی شناسایی شده در این جمعیت شامل سه مورد لنفوم هوچکین (HL) و چهار لنفوم غیر هوچکین (NHL) بود. نتایج بررسی وج بیوپسی انسیزیونال از نظر آماری با نتایج اکسیزیونال قابل تطبیق بود و همخوانی داشت.

نتیجه‌گیری:

علت اصلی LAP لنفادنیت واکنشی است. اکثر این موارد تنها با بیوپسی قابل تشخیص و مدیریت هستند. بیوپسی می‌تواند نقش کلیدی در تریاژ موارد مناسب داشته باشد، به طوری که این بیماران برای تشخیص و یا ارجاع به مراکز بالاتر مورد بررسی بیشتر قرار می‌گیرند. در مواردی که که اکسیزیونال بیوپسی به دلیل بزرگی لنف نود و یا چسبندگی عروقی خطرناک هست می‌توان از وج بیوپسی انسزیونال به عنوان روش تشخیصی قابل اطمینان جهت شروع درمان کمک گرفت.

واژه‌های کلیدی: لنف آدنوپاتی، اطفال، بیوپسی، اکسیزیونال، گوه‌ای

بررسی نتایج پاتولوژی توده‌های شکمی در کودکان

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مقدمه:

تعیین پاتولوژی توده‌های شکمی اطفال همواره نقش مهمی در مدیریت بالینی آنها داشته و با وجود اینکه تفکیک میان انواع پاتولوژی‌های بدخیم قدم مهمی در هدایت پزشکان اطفال و جراحان به سمت برنامه درمانی مناسب است، بیشتر مطالعات به ارزشیابی ابزار تشخیصی از نظر شناسایی بدخیمی محدود بوده است.

هدف:

در این مطالعه به صورت گذشته‌نگر ارزش تشخیصی بیوپسی از طریق کور نیدل را در شناسایی بدخیمی و تعیین نوع پاتولوژی تومور شناسایی شده در توده‌های شکمی اطفال سنجیدیم.

روش اجرا:

از میان اطفالی که از فروردین *90* تا دی ماه *98* با شکایت توده شکمی مراجعه کرده بودند، پرونده *67* بیمارانی که تحت کور نیدل قرار گرفته و بنا بر جواب پاتولوژی بدخیمی برای آنها تشخیص داده شده مطالعه شد و از مدارک ایشان جنسیت، سن، تظاهرات بالینی، سابقه خانوادگی، گـزارش پاتولوژی نمونه بیوپسی کور نیدل، جواب پاتولوژی نمونه جراحی و گزارش سی تی اسکن شکم و لگن ثبت شد.

برای تعیین تاثیر تشخیصی کور نیدل بیوپسی، گزارش پاتولوژی نمونه‌های کور نیدل بیوپسی را با گزارش پاتولوژی نمونه جراحی در *25* بیماری که تومور ایشان با جراحی برداشته شده بود، مقایسه کردیم. از میان *25* بیمار ذکر شده، در مدارک *18* بیمار گزارش سی تی اسکن شکم و لگن موجود بود که در مقایسه دقت این مدالیته تشخیصی با کور نیدل بررسی شد.

نتایج:

در تمام گزارش‌های پاتولوژی، میزان بافت نمونه برداری شده توسط بیوپسی کور نیدل برای تفسیرهای پاتولوژی کافی قلمداد شده بود و در هیچ یک از بیمارانی که تحت کور نیدل بیوپسی قرار گرفته بودند، عوارض مربوط به کور نیدل دیده نشد.

در *25* بیمارمورد بررسی، کور نیدل با ارزش اخباری مثبت *96* درصدی قادر به تشخیص بدخیمی بوده و در تشخیص نوع پاتولوژی تومور در *76* درصد موارد موفق عمل کرده است؛ چنانچه در این زمینه، توان تشخیصی قطعی در تفکیک بین نوروبلاستوم، ویلمز و هپاتوبلاستوم داشته است. هم چنین در مقایسه با سی تی اسکن بیوپسی کور نیدل هم در شناسایی بدخیمی هم در تشخیص نوع تومور (صحت *83* درصدی در کورنیدل بیوپسی در مقایسه با صحت *61* درصدی سی تی اسکن) بهتر عمل کرده بود.

نتیجه‌گیری:

با توجه ارزش اخباری مصبت قابل قبول در شناسایی بدخیمی و صحت تشخیصی قابل توجه در تعیین نوع پاتولوژی تومورها در عین نداشتن عارضه، ما کور نیدل بیوپسی را به عنوان روشی حساس، دقیق و امن برای تشخیص ماهیت و ویزگی توده‌های شکمی اطفال پیشنهاد می‌کنیم.

کلمات کلیدی: نور تیدل بیوپسی، سرطان اطفال، تومور توپر، توده شکمی، اطفال، تشخیص

Updates in Pediatric Solid Tumors Management

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Background:

According to WHO, cancer is one of the leading cause of death for children and adolescents. The cancer incidence rates have been rising for the past few decades. Long non-coding RNAs (lncRNAs) are a group of transcripts with longer than 200 nucleotides that lack the coding capacity and play vital regulatory roles in cancer initiation and development in both adults and children. In particular, many lncRNAs are stable in cancer patients' body fluids such as blood and urine, suggesting that they could be used as novel biomarkers. In support of this notion, lncRNAs have been identified in liquid biopsy samples from pediatric cancer patients.

Neuroblastoma:

New immunotherapeutic techniques and nuclear medicine-targeted therapies have emerged and are demonstrating promising response rates for patients at high risk. Data indicate that blood-borne exosomal hsa-piRNA-1089 is a diagnostic marker for NB and assessing metastasis and Increased D-dimer level was a poor predictor of neuroblastoma, especially in the high-risk group.

In summary:

Elucidation of the function of lncRNAs and association with diverse subtypes of childhood cancer, and development of novel lncRNA-based approaches for diagnostics and targeted therapy hold considerable potential, despite the challenges ahead.

Hepatoblastoma:

Advances in surgical (Indocyanine green-guided surgery and liver partition and portal vein ligation for staged hepatectomy) and medical treatment (Cisplatin-containing chemotherapies) provide better outcomes for children with HB, and identifying novel targets may lead to the development of future targeted therapies and immunotherapies.

Intraspinal Extension of Neuroblastoma

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Abstract

In a subset of patients with para spinal neuroblastoma, tumor growth may extend into the spinal canal (dumbbell tumor). If neurologic symptoms result, urgent treatment is required to prevent permanent injury to cord. Each of the three main therapeutic modalities (surgery, radiation, and chemotherapy) has been used. A POG report showed similar rates of neurologic recovery in patients treated with surgery or chemotherapy, but significant orthopedic sequel were seen more commonly in patients treated with surgery. Although chemotherapy is probably considered most appropriate for the initial managements of these patients, improvements in neurological techniques, including the use of laminotomy to access the intraspinal tumors may allow reconsideration about the optimal approach, especially in patients with acutely progressive symptoms.

The appropriate approach for patients with asymptomatic intra spinal tumor extension is also uncertain. For patients with low or intermediate – risk disease, the risks of attempting to remove the intra spinal component of a para spinal tumor likely outgrowth the benefits. This situation is usually arises in patients with thorasic primary tumors. The intra thorasic component is resected, and gross residual disease remains in in the spinal canal. Care should be taken to minimize operative complications such as leakage of cerebrospinal fluid or uncontrollable intra spinal bleeding. As residual foraminal disease rarely shows to a symptomatic size, the importance of conservative therapy in this circumstance should be emphasized. In the absence of metastatic disease or unfavorable tumor biology, these patients will be classified as L1 (INSS stage 2 A/B), low risk, and have a very favorable prognosis with no further therapy. For patients with high-risk disease, the importance of resecting gross intra spinal disease is uncertain.

Influence of Complete Surgical Excision on the Survival of Patients with Stage 4 Neuroblastoma

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Abstract

Neuroblastoma is the third most common solid malignancy of childhood with the overall incidence of

1 per 100,000 children (median age of 17 months). Unfortunately about 40-50% of patients have metastatic disease at the time of diagnosis.

The recent therapeutic advances as the multimodality approach to stage 4 high grade tumor consists of induction chemotherapy; therapy to the site of the primary tumor (surgical excision and radiotherapy); high-dose therapy (HDT) with stem-cell rescue (SCR); and residual disease therapy, including immunotherapy.

Complete excision could been achieved in about 70-77% of patients after induction chemotherapy.

According to the recent treatment modalities, in patients with stage 4 neuroblastoma who have responded to induction therapy, complete resection of primary tumor is associated with improved survival and local control. But optimal extent of surgery in stage 4 patients is still a matter of debate. Comparing survival after true complete resection (total macroscopic tumor removal) with survival after gross total resection (about 95 % tumor removal), the significant difference were not shown.

Complete excision lead to higher peri-operative morbidity and mortality with only slightly better results. Nevertheless despite all previous studies, the possibility that complete resection may be advantageous over gross tumor resection has to be acknowledged.

Pediatric Neuroblastoma with Initial Presentation as a Neck Mass: Case Report in 12 Years Old Girl

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Background:

Neuroblastoma is the most common solid extracranial malignancy of childhood and the most common malignant tumor in infants. The overall incidence of neuroblastoma is 1 per 100,000 children in the United States, thereby accounting for 7-10% of all malignancies diagnosed in patients younger than 15 years of age. Yet neuroblastoma is responsible for approximately 15% of all childhood cancer deaths. Neuroblastoma is a heterogeneous disease. Tumors can spontaneously regress or mature, or display a very aggressive, malignant phenotype. Because of these unique characteristics, neuroblastoma has been of great interest to both clinicians and basic science researchers. Progress in molecular cellular biology in the past 40 years has contributed greatly to a better understanding of this disease. Unfortunately, this progress has not significantly altered the clinical outcome foe patients with high- risk disease. Although the prognosis for these patients has improved somewhat in the past three decades. The long- term outcome remains very poor.

Methods:

The patient is 18 years old girl with recurrent neuroblastoma. The first symption 5 years ago was a neck mass with pathology of neuroblastoma after chemotherapy and 3 surgury now General condition of patient is good and in recovery period.

Results:

After treatment and chemotherapy and third surgery patient in recovery period now. In well general condition.

Discussion:

This disease need a multidisciplinary approach to achieve a best result contain oncologist team that well trained, good surgery team, and pathologist.

Neuroblastoma in a 3 years Old Boy

Amanollahi O

Abstract

A 3 years old boy referred to our department due to abdominal pain and mass, after diagnostic test including imaging, sonography and ct.scan he was candidate for surgery with diagnosis of neuroblastoma of right adrenal. In operation we noticed that origin of tumor was neurological tissue of lumbar spine underside of inferior vena cave in a hard and bad position for removal. Finally complete excision of tumor has been done successfully and sample sent to pathology that ganglioneuroblastoma was the result. Patient discharged 2 days after in very good position and referred to oncology department for complementary assessment.

A Huge Neuroblastoma in a Six-month-old Infant with Different Pathology after Resection

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Abstract

A six-month old infant was consulted due to a huge abdominal mass. Her parents noticed abdominal distention when she had two months old and during evaluation a retroperitoneal mass at right adrenal gland was found. Pathology report of needle biopsy showed neuroblastoma. She underwent four cycle chemotherapy however despite chemotherapy the mass enlarged vigorously.

She underwent laparotomy for tumor resection. A huge mass which weighted about 1300 grs was resected completely. The final pathology and immunohistochemistry was reported as a mature teratoma.

Keywords: Neuroblastoma, Teratoma, Pediatric Cancer

Ganglioneuroma with Shortness of Breath and Thoracoabdominal Involvement in a 12 Years Old Boy: A Case Report

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Abstract

The most benign tumor is the ganglioneuroma, which is composed of gangliocytes and mature stroma. Ganglioneuromas (GNs) are wholly differentiated neuronal tumors that do not contain immature elements and potentially occur anywhere along with the peripheral autonomic ganglion sites. The patient is a 12-year-old boy who referred to Imam Khomeini Hospital in Ahvaz with complaints of cough, cold and shortness of breath. The patient underwent chest radiography and a very large mass was observed on the left side of the chest, which caused the heart to shift to the right side, and the patient underwent echocardiography and CT scan for further investigation. Based on abdominal and pelvic CT, a heterogeneous hypodense mass with dimensions of approximately 135x105 mm was observed in the left upper region, which caused a severe deviation of the heart and mediastinum to the right side, an elevation of the left hemi-diaphragm, and a downward displacement of the left kidney. Also, it was reported that the above mass primarily favors benign lesions of adrenal origin. The samples were sent to the laboratory for pathology examination. Based on the biopsy report, a large heterogeneous area was observed in the left half of the abdomen and thorax, which extended upward to near the upper part of the left hemithorax. The mentioned area is not moved by breathing and is located in the retroperitoneum. The pathology of the patient was ganglioneuroma. Then the patient underwent surgery with a midline incision extending to the 6-7 intercostal space (thoraco-abdomen). Finally, the patient was discharged with good general condition after 3 weeks of hospitalization. Ganglioneuroma is a rare benign tumor, usually asymptomatic. The main treatment for that is complete surgical excision. In case of tumor resection, chemotherapy and radiotherapy are not indicated.

Keywords: Ganglioneuroma, Thoracoabdominal Involvement, Thoracoabdominal Resection, Child

Pediatric Liver Transplantation for Cancers

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Abstract

Liver transplantation is the treatment of choice for end stage liver disease in all age groups. In pediatric age subgroup, however, metabolic diseases and congenital biliary anomalies are the most common indications for liver transplantation. Acute liver failure with or without underling liver disease another common cause for liver transplantation.

Primary liver malignancies are one of the uncommon indications for liver transplantation in children. The usual liver malignancy in pediatric liver transplantation is hepatocellular carcinoma (HCC) which often develops on underlying liver disease. The most common cause of HCC in children is Tyrosinemia, a congenital defect in tyrosine metabolism. Another underlying disease that may end up with HCC is galactosemia which is indication for liver transplantation.

The other primary liver malignancy that may be treated with liver transplantation is unresectable non-metastatic hepatoblastomas. Hemangioendothelioma is another low grade malignancy. These tumors are resected if possible, but liver transplantation is an option for unresectable ones.

In this lecture, we present the results of pediatric liver transplantation in Imam Khomeini Hospital Complex, Tehran; and discuss the indications for liver transplantation in children.

تومورهای استخوانی

دکتر آرش ملکی

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چکیده:

توده های استخوانی معمولا با شکایت درد مراجعه میکنند که در طی رادیوگرافی متوجه درگیری استخوان میگردیم. توجه به علایم از جمله ماهیت درد (الگو و مدت بروز درد) و شکل رادیولوژیک ضایعه در تشخیص و بدنبال آن درمان موثر خواهد بود. در بیشتر مواقع برای تشخیص قطعی توده های استخوانی، انجام بیوپسی لازم است و البته قبل از آن باید کلیه اقدامات پاراکلینیک لازم شامل ام ار ای و سی تی اسکن انجام گیرد. انجام اقدامات کلینیک و پاراکلینیک لازم سبب تصمیم گیری مناسب شده و این مساله به نفع بیمار خواهد بود. در این فرصت در کنار بررسی اقدامات لازم در تومورهای استخوانی، به بررسی چند تومور استخوانی شایع میپردازیم.

Rare Surgical Presentations of Cancer in Childhood

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Background:

Most childhood tumors will first present to a physician; some tumors will present in an atypical manner and may mimic a surgical condition. The diagnosis may be missed if the surgeon is not aware of the possibility of cancer. It is important that the surgeon who is not experienced in the management of childhood cancer is aware that an apparently benign condition could be a manifestation of an underlying malignancy, like acute abdominal pain, intussusception, gastrointestinal bleeding …

On the other hand, many complications could occur in pediatric malignancies as a common cause for surgical consultation on the oncology ward. Soft tissue complications, compartment syndrome, thoracic complications such as superior vena cava syndrome, fungal lung lesions, catheter related thrombosis, gastrointestinal and genitourinary complications are among the common problems which could occur in pediatric malignant disorders.

Discussion:

Early and correct diagnosis of surgical manifestation and complications could be life saving for the patient and can prevent spending extra money and decreased mortality and long term morbidity among pediatric patients.

Inflammatory Myofibroblastic Tumor with Urinary Tract Presentation in 15 - year - old - boy: A Case Report

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چکیده

Inflammatory myofibroblastic tumor یک تومورخوش خیم بسیار نادر است که از عضلات صاف، سلول‌های میوفیبروبلاست بافت همبند و سلول‌های التهابی منشا می‌گیرد. محل شایع درگیری ریه، اوربیت و صفاق بوده و مشاهده آن در سیستم ادراری بسیار نادر است. می‌تواند بدون علامت یا تهاجمی موضعی و حتی همراه با متاستاز دوردست باشد. بدخیمی بسیار نادر است. در نمای بالینی، رادیولوژیکی و هیستوپاتولوژیکی بسیار شبیه لیومیوسارکوم و رابدومیوسارکوم است. عود این تومور پس از جراحی 37-11٪ و شانس متاستاز دوردست 11٪ است.

اسامی دیگر آن عبارتند از:

Inflammatory pseudotumor, Fibrous xanthoma, Plasma cell granuloma, Pseudosarcoma

در این مقاله مورد بسیار نادری از این بیماری معرفی می‌شود. بیمار پسر 15 ساله‌ای هستند که به علت درد شکم، دیزوری و هماچوری مراجعه نمودند. در سونوگرافی و سی تی انجام شده کلیه‌ها و حالب نرمال بودند ولی جدار مثانه 11 میلی متر ضخامت داشت. همچنین یک ساختار کیستیک با جدار ضخیم و حاوی کانون‌های اکوژن کلسیفیه به ابعاد 32 در 38 میلی‌متر در قدام مثانه گزارش شد که در بررسی کالر داپلر پر خون بود. تشخیص بر اساس گزارش سی تی آبسه کیست اوراکوس ذکر شد. بیمار تحت عمل جراحی قرار گرفت که در بررسی پاتولوژیکی Inflammatory Myofibroblastic Tumor عنوان گردید. بیمار با حال عمومی خوب ترخیص و با یک سال پیگیری مشکلی نداشته است.

Pediatric Gastrointestinal Basidiobolomycosis Mass: Case Report

Hajesmaielie A

Background:

Basidiobolomycosis is a rare fungal infection in which gradually enlarging granulomas form that mainly causes subcutaneous infections and rarely gastrointestinal disease in immunocompetent hosts.

Methods:

We are reporting a 7 years old Iranian girl that admitted because of ileocolic intussusception. We found a mass in cecum (like malignancy) that RT hemicolectomy had done. Her pathology was eosinophilic colitis with basidiobolomycosis. she returned to the hospital with generalized abdominal pain like pancreatitis and bowel obstruction about 20 days after the first operation. She had two masses in body and tail of pancreas and Lt para colic space. So, fungal medical therapy started and she had operated that a 5×7 cm mass was in splenic flexure of colon and a 5×3 cm mass was in body and tail of pancreas.

Results:

We resected enteric masses because of bowel obstruction but because of a mass in whole of pancreas we did not pancreatectomy, so antifungal therapy had started for about 6 months.

Discussion:

These fungal masses are very aggressive with high recurrent rate that mimicking malignancy, so combination of surgical resection and longtime antifungal therapy is very effective.

Lung Bronchogenic Carcinoma in 9 Years Old Girl

Bigdeli N

Background:

Our patient is a 9 years old girl with suspicious history of FBA and chocking for 3 month ago. She had also cough and hemoptesis from that time. In first evaluation in HRCT a mass was seen in root of LUL bronchous.

By suspicious of FBA rigid and fiexible bronchoscopy was done that a fragile and hemorrhagic mass was seen in that location of HRCT. So patient was candidated for sonography or CT scan guided biopsy that was not done. So open surgery was done.

By left posterolateral incision thorachotomy was done.A huge mass in upper lobe was seen that was expanded from LUL bronchial to the LMB and root of LLL bronchous.So sleeve lobectomy was done that it is a rare procedure in thoracic surgery field.

Conclusion:

Regardless of rarity of more malignansis in children we should note them in our differential diagnosis and all of diagnostic modality should be done and empirical thrapy must be avoided.

Testicular Tumor in 15 Years Old Boy

Bigdeli N

Abstract

My patient is a 15 years old boy with feelin of enlarging of right testis. In first physical examination (inspection and palpitation) didn't find any evidence of mass or abnormality and only for reassurance I wanted a sonography.

Sonography introduced a solid 0.5 × 0.5 mm mass with calcification was seen in upper pole of right testis (in testicular tissue).

So patient was candidate for surgery and concurrent frozen-section that unfortunately introduced seminoma. So right orchiectomy and left orchiopexy was done and onchologic consultation was requested.

Conclusion:

In pediatric surgery every chief complain from everybody (child or her / his parents) should be noted and all of effective diagnostic modality should be done until R/O of serious condition. Only normal examination not to be sufficient for absolute decision or final assessment.

Abdominal Mass and Hematuria; a Rare Early Presentation of Burkitt Lymphoma

Khazravi M

Abstract

Introduction:

Burkitt is a subtype of NHL, known for its characteristic rapid growth; it can be duplicated in 26-66 hours. Its more prevalent in boys and M:F rate is 4:1. most involved patients are 5-9 years old with mean age of 8 y. Burkitt's was first introduced in 1958 in Uganda (Africa) characterized by its rapid growing mass then after it was categorized to three subtypes: endemic, sporadic & immune deficiency related Burkitt's. The early presentation of abdominal and pelvic Burkitt's is often the palpable mass and one of following symptoms: abdominal pain, abdominal distention, GI obstruction, appendicitis, and oral feed intolerance even jaundice is reported in some cases due to biliary tree compression. In some cases renal failure happens secondary to mass effect or paraneoplastic events such as cryoglobulinemia or glomerulonephritis.

Presentation:

A 2.5 years old child was referred to our institute with chief complaint of gross hematuria during recent 3 weeks and a rapid growing abdominal mass noticed since the last week. The mass was apparently palpable at the examination and even the enlargement was noticeable during primary workup in few days. Ultrasonography at day three after admission showed a hypoechoic lobulated mass measuring 93×88×73 mm extending from the dome of bladder to umbilical area and few adjacent lymphadenopathies (mean size 23×14. the final diagnosis was Burkitt lymphoma.

Conclusion:

We here presented a case of rapid growing abdominal mass & gross hematuria as a new picture of Burkitt's lymphoma. Without other known early symptoms such as Gi obstruction, pain, etc. Other researches never reported gross hematuria as the first symptom.in this case after the primary imaging and stabilizing the patient, total resection of tumor and reconstruction was done and adjuvant chemotherapy cured the patient with no recurrence and complications.

Efficacy of Transarterial Chemoembolization (TACE) with Intra-arterial Chemotherapy (IAC) for the First Time in Pediatric Inoperable Retroperitoneal Tumors in Iran: A Report of 3 Cases

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Background:

Inoperable retroperitoneal tumors are those that are unable to be removed surgically because of their location in the abdominopelvic area or because there are near main vessels or spinal cord. Retroperitoneal tumors can cause a diagnostic dilemma and present several therapeutic challenges because of their rarity, relative late presentation and anatomical location, often in close relationship with several vital structures in the retroperitoneal space. Complete surgical resection is the only potential curative treatment modality for retroperitoneal solid tumors and is best performed in high-volume centers by a multidisciplinary team. The ability completely to resect a retroperitoneal tumor grade remains the most important predictors of local recurrence and disease-specific survival.

Case Presentation:

We introduce 3 children with neuroblastoma tumor, who were initially unable to undergo surgery due to the involvement of the aorta and mesenteric vessels or close proximity to the thoracolumbar spinal cord, and each of them underwent TACE-IAC 2-3 times and was able to undergo surgery.

Results:

Trans-catheter arterial chemo-embolization (TACE) and Intra-arterial chemotherapy (IAC) is a minimally invasive technique performed by interventional radiologists or vascular interventionists that delivers chemotherapy drugs with embolization, injected through a catheter, into the feeding arteries of tumors directly supplying the tumor. Although TACE-IAC is used frequently in adult intraperitoneal tumors such as hepatocellular carcinoma, neuroendocrine tumors or ocular melanomas, to our knowledge and experience it has not yet been performed on pediatric tumors. Also, in our cases it was done on retroperitoneal tumors for the first time.

Discussion:

The result was completely successful for all three patients; it is recommended that this method be considered in large and inaccessible retroperitoneal pediatric tumors.

Cutaneous Diphtheria Complicated Oncologic Reconstruction Surgery in Osteosarcoma

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Background:

Cutaneous diphtheria is usually complicated pre-existing cutaneous lesions, including traumatic abrasions, surgical wounds, burns, insect bites, pyoderma, eczema, impetigo, and dermatitis, which causes a breach in the skin surface; however, it also could be appeared on previously healthy skin. Cutaneous diphtheria rarely develops into an invasive disease in immunocompetent patients. Here, we report a case of lower limb osteosarcoma complicated with post-surgical cutaneous diphtheria.

Case Presentation:

A 15-year-old female, a known case of right lower limb osteosarcoma with a history of reconstruction surgery after tumor resection, visited for SSI on April 01, 2021. She has a history of open reduction and internal fixation of the right tibia using a cadaveric bone graft in a rural setup elsewhere. During the 14th course of chemotherapy, she developed localized surgical site cellulitis, which progressed gradually to complete wound dehiscence after about 4 weeks. On admission, a large skin defect over the anterior aspect of the right tibia with an exposed black cadaveric bone (about 3 cm below the knee) was found. Despite primary surgical debridement, a progressive necrotic ulcer developed around the skin defect 3 weeks later.

Discussion:

Cadaveric bone was replaced with a metal prosthesis after clinical improvement about 6 weeks after anti-diphtheria treatment. The patient's chemotherapy continued successfully without further complications.

Conclusion:

cutaneous diphtheria could be missed due to nonspecific clinical presentation. So, any chronic non-healing ulcer should arouse the suspicion of rare etiologies such as cutaneous diphtheria. Skin ulcers not responding to conventional antibiotic treatment should be investigated for uncommon organisms such as C.diphtheriae.

Clipless Laparoscopic Adrenalectomy in Children less than 15 Years Old: A Single Center Experience

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Background:

Laparoscopy is the gold standard approach for management of some adrenal masses in adult cases. Still there have not been many findings in case of children. We present our experience with clipless laparoscopic adrenalectomy in pediatric cases for the first time.

Methods:

From January 2009 to January 2020, sixteen laparoscopic adrenalectomies were performed in patients 1-15 years old. The first port (10 mm) was inserted using open approach above the umbilicus and three 5 or 3 mm trocars were inserted under direct vision. On the left side, the colon was mobilized medially, then the renal vein exposed. Adrenal vein was coagulated using bipolar cautery after separating from renal vein. No endoscopic clips were used.

Results:

Ten girls and six boys with the mean age of 12.4 years old (ranging from 1 to 15 years old) underwent laparoscopic adrenalectomy. The mean operative time was 151 ± 47 (80- 240) minutes. The mean size of adrenal lesions in greatest diameter was 6.9 ± 2.4 cm (2.2 to 10). The mean hospital stay was 3.7 days (2-5) and average follow-up time was 36 months (18-48).

Conclusion:

Laparoscopic adrenalectomy in children and young adults is effective and safe if the cases are selected appropriately. Clipless laparoscopic approach by an expert surgeon has acceptable outcomes.

Applying Totally Implantable Venous Access Devices (TIVAD) in Children

Khalegnejad Tabari A, **Mohammadi D**

Abstract

Background:

During recent years and paralleling the advances in the treatment of patients requiring chem- otherapy or long-term total parenteral nutrition (TPN), it has been necessary to provide a chronic central venous access with a low complication rate and long-term availability (months or even years).

In our country, this procedure is performed and its technique is refined, but its advantages and complica- tions have not been analyzed and reported.

Materials and Methods:

The records of 120 patients who had undergone TIVAD placement in Mofid children's hospital, Tehran from 1999 to 2005 were retrospectively reviewed. Outcomes and compliance of parents and therapeutic team were evaluated.

Results:

There were 120 patients, 68 boys (56.6%) and 52 girls (43.3%); with the age range of 3 months to 13 years old. The following postoperative complications were encountered; withdrawal occlusion in 4 patients (3.3%), intraluminal fibrin sheath in one patient (0.8%), severe neutropenia in 3 patients (2.4%), complete intraluminal occlusion of the catheter in one patient (0.8%), fever and chills in 2 patients (1.6%), and catheter dislodgement in only one patient (0.8%). All parents and members of the therapeutic team were pleased with the TIVAD (100% acceptance).

Conclusions:

TIVAD placement can be performed in infants and children of all ages. In cases where a chron- ic venous access is needed, the use of this device is appropriate, because of its low complication rate and long-term applicability.

Keywords: Neoplasms, Total parenteral nutrition, Venous access

Totally Implantable Subpectoral vs. Subcutaneous Port Systems in Children with Malignant Diseases

Rouzrokh M, **Mohammadi D**

Abstract

Background:

For many years, subcutaneous therapeutic port system was known as a major route to access central veins. However, significant complications have been reported through recent years. One of the most important complications of subcutaneous port implantation is skin necrosis. In order to decrease this complication, we would like to introduce subpectoral fascia port implantation through this study. A

Methods:

Five hundred and twenty four patients with a, variety of neoplastic diseases underwent port implantation, from March 2003 to March 2008 (60 months). All suitable size catheters were put in the superior vena cava through the internal jugular vein under general anesthesia. The ports were placed in the subcutaneous pocket (SCP group) in 342 patients and in the subpectoral fascia pocket (SPFP group) in 182 patients. Data were analyzed using Chi-square test and survival analysis for time (Kaplan-Meier).

Results:

A total of 538 devices were placed for 524 patients in two groups (14 patients received a second device after removal of the first one, due to failure of the first implantation). Mean follow- up period was 508 days (8-2025 days).

One, due common complications observed in the SPFP group were as follows: wound infection (7 cases, 3.8%), catheter obstruction (7 cases, 3.8%), catheter displacement (6 cases, 3.2%), port related infection (5 cases, 2.7%), and pocket hematoma (2 cases, 1.1%).

Common complications observed in the SCP group were as follows: catheter displacement (12 cases, 3.5%), skin necrosis (11 cases, 3.21%), port exposure (9 cases, 2.6%), port related infection (8 cases, 2.3%), catheter obstruction (8 cases, 2.3%), and port rotation (3 cases, 0.9%).

Conclusion:

The results showed that port implantation in the subpectoral fascia pocket had a lower rate of skin complications than the subcutaneous pocket implantation. According to this study, this procedure was not complicated by skin necrosis over the port, port exposure or port rotation.

Comparison between Open and Ultrasonography Guided Venous Access Ports in Children with Malignancy

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Background:

Long-term central venous access is used in children for various reasons specially for chemotherapy. Vessels diameter in children are smaller and more prone to injury and complications such as thrombosis. Different methods are used for implantation of port-a-cath in children. We aimed to compare the complications of insertion of central venous access ports between two methods of open and ultrasound guided.

Methods:

All children referred to pediatric surgery department of a children hospital from April 2018 to March 2020 for implantation of port-a-cath were included. Right jugular vein was the target vein and patients were randomly divided between two methods of open insertion by lateral neck exploration and ultrasound real-time guided percutaneous and the reservoir was fixed in subpectoral fascia pouch. Patients were followed up for early and late complications or malfunction for at least 6 months.

Results:

We included 76 patients less than 18 years of age: 24 patients with ultrasound guided method (median 3 years) and 52 patients with open exploration method (median 6 years). We observed no statistically significant difference between two groups with respect to sex, underlying disease, and complications. Most patients had hematological malignancies including ALL (52.9%), AML (19.1%) and the rest had solid organ malignancies. Early complications were observed in 2 (3.8%) in the open and 1 (4.2%) in the US- guided group (P=1). Late complications were observed in 9 (17.3%) patients in the open group and 1 (4.2%) in the US guided group. Infection was observed in 9.6% and malfunction in 5.8% of the open group leading to earlier removal of the catheter. There was not any complication indicative of infection in the US-guided group.

Discussion:

US-guided method can be suggested for routine use as a safe method of insertion of port venous access in children.

Evaluation of the Catheter tip Position among Children with Totally Implantable Port during 2015 in Dr. Sheikh Children's Hospital

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Abstract

Introduction:

Totally implantable port insertion is a common procedure in pediatrics surgery and the main concern is the position of catheter tip, which is directly related to its complications, failure and durability. The best position is in superior vena cava (SVC)/ right atrium (RA) junction which is compatible with carina or T5-T6 vertebral level in chest x-ray. In our center we routinely use surface anatomy to estimate the adequate length for the tip to reach SVC/RA junction. In this study we compared the precision and accuracy of this method by post-operative chest x-ray study.

Materials and Methods:

As a retrospective study we evaluated the accuracy of surface anatomical land marks to estimate the catheter tip position, considering the carina or vertebra in chest x-ray which represents the SVC/RA junction. Forty eight patients were included and their records were reviewed in Dr. Sheikh Children's hospital in Mashhad.

Results:

Considering the carina as the best radiographic land mark, we had accurate tip position only in 29.2%, over insertion in 45.8% and under inserted catheter tip in 10.4%. Considering the vertebral bodies as radiographic land mark, 50% were over inserted, 35.4% accurate and 14.6% under inserted.

Conclusion:

Regarding our high rate of catheter tip mal-position arising from considering surface anatomy alone, we suggest not only to rely on surface anatomy but use imaging modalities such as portable X-ray, fluoroscopy or ultra-sonography within the operation room while inserting implantable port devices.

Keywords: Pediatrics, Implantable Catheter, Superior Vena Cava

Cell and Gene-based Immunotherapy: Overview and Products

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Abstract

The era of Regenerative Medicine as future medicine is upon us. Thousands Ideas to market have demonstrated profound, durable and potentially curative effects that are already improving human quality of life for patients who have no other available therapeutic options. Until 2022 products reports in regenerative medicine field included three main branches of gene therapy, cell therapy and tissue engineered based therapy. Cell and Gene therapy developers raised the vast majority of investment, generating $21B in 2021, respectively. However, investment in Cell and Gene-based companies is growing at a faster pace than cell therapy financing. There is no reason except breakthrough treatment for cancer patients and their promising results. According to the latest reports, there are numerous clinical trials in different phases and achieved unprecedented bench to bedside clinical success. So, related market authorized products are increasing subsequently. There are six FDA approved CAR T cell therapy products. Also, there are a superiority in the number of this type of candidate products in the banner year of regulators' Regenerative Medicine Advanced Therapy (RMAT) designation such as FDA and EMA.

بررسی ده ساله توده‌های تخمدان در کودکان مراجعه کننده به بیمارستان کودکان مفید

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زمینه و هدف:

توده‌های تخمدانی طیف وسیعی از آسیب شناسی از تومورهای بدخیم بسیار تهاجمی تا کیست‌های خوش خیم را دربرمی‌گیرند. تقریباً در حدود 2٪ از تمام بدخیمی‌های زنان در کودکان رخ می‌دهد، به نحوی که 60-70٪ از این ضایعات در تخمدان به وجود می‌آیند. لذا در این مطالعه به بررسی توده‌های تخمدانی در کودکان مراجعه کننده به بیمارستان کودکان مفید تهران طی سال 1381 تا 1391 پرداختیم.

روش مطالعه:

این مطالعه یک بررسی توصیفی - مقطعی بود که به صورت سرشماری بر روی 57 کودک مبتلا به توده تخمدانی تحت عمل جراحی مراجعه کننده به بیمارستان کودکان مفید طی سال‌های 1381 تا 1391 انجام گرفت. متغیرهایی همانند علایم بالینی، سن زمان تشخیص و بستری، نوع جراحی، یکطرفه یا دوطرفه بودن، یافته‌های حین جراحی و نتایج سونوگرافی، سی تی اسکن و سیتولوژی و پاتولوژی از پرونده‌ها استخراج شد و در فرم اطلاعاتی ثبت گردید. نتایج توسط نرم افزار SPSS نسخه 18 آنالیز شد.

یافته‌ها:

57 دختر (40/2 ± 57 ماه با دامنه یک روزه تا 15 سال) که تحت 64 عمل جراحی تخمدان (24 مورد Salpingo – Oophorectomies، 10 مورد Oophorectomies ، 21 مورد Cystectomies تخمدان و 2 مورد بیوپسی تخمدان) قرار گرفته بودند، بررسی شدند. 50 کودک مبتلا به توده تخمدان یکطرفه (49٪/1 سمت راست و 38/6٪ سمت چپ) بودند. 26 نفر (45٪/6) درد شکم حاد، 20 نفر (35٪/1) لمس توده شکمی، 3 نفر (5٪/3) تب، 3 نفر (5٪/3) تهوع و استفراغ، 21 نفر (33٪) پیچ خوردگی تخمدان داشتند. 8 نفر (15٪) تومور بدخیم و 4 نفر (8٪) تومور خوش خیم داشتند. هیچ تفاوت معنی‌داری بین میانگین سن مبتلایان به توده‌های خوش خیم (8/2 ± 2/6 سال و بدخیم 6/1 ± 5/3 سال) مشاهده نشد.

نتیجه‌گیری:

تومورهای تخمدان در کودکان نادر هستند. اغلب آنها در مطالعه حاضر خوش خیم بودند و ریسک بدخیمی با سن افزایش می‌یافت. پیگیری بیماران باید شامل تاریخچه دقیق و معاینات فیزیکی، سونوگرافی، تومور مارکرها، ارزیابی داخل شکمی و پیگیری بدخیمی باشد. درمان جراحی در ضایعات خوش خیم و بدخیم مداخله‌ای محافظه کارانه است، گرچه در موارد بدخیمی، بیماران باید بعد از عمل جراحی تحت شیمی درمانی قرار بگیرند.

واژه های کلیدی: توده تخمدان، کودکان و تومور بدخیم

Successful Treatment of Large Bladder Mass Due to Eosinophilic Cystitis: A Rare Case

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Abstract

Background:

Eosinophilic cystitis (EC) is a rare bladder disease in children, which is often misdiagnosed with urinary tract infection, bladder tumor, and parasitic infection. The disease usually presents with a spectrum of urological symptoms including increased frequency, hematuria, and suprapubic pain. Herein, we present a rare case of a large bladder mass due to EC.

Case description:

Our case was a 28-month-old male presenting with urinary retention, urgency, inability to urinate, and agitation. Bladder biopsies demonstrated massive eosinophilic infiltration of the bladder, confirming the diagnosis of EC. Treatment with prednisone was initiated in a dose of 2 mg/kg for 2 weeks, 1 mg/kg for 2 weeks, and 0.5 mg/kg for 2 weeks. The symptoms diminished within 8 weeks of steroid treatment. After 2 months, bladder wall thicknesses decreased, and on long-term follow-up, all complaints were fully resolved.

Conclusion:

Oral steroids can be an effective and non-invasive choice for the treatment of huge bladder masses due to EC.

Pelvic Germ Cell Tumor in Children

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Abstract

GCTs are rare and accounting for 3% of childhood cancer, arising from the aberrant migration of common progenitor cells. These cells first appear in endoderm of embryonic yolk sac then migrate to developing gonads. Normally germ cells are only found in the ovaries and testes. GCTs can arise in both extra gonadal and gonadal location; the extra gonadal sites predominate in children.

The majority of GCTs is benign and include mature and immature teraromas.

Malignant GCTs notably include yolk sac tumors (endodermal sinus tumor), embryonal carcinoma, choriocarcinoma, gonadoblastoma and mixed malignant GCTs. Serological tumor biomarkers such as AFP and B-hCG are used for diagnosis and monitoring of recurrence. Malignant GCTs can develop metastasis to regional LN and lung.

The intra-abdominal undescended testes and MGD and Kline filter syndrome are associated with increased risk of GCTs.

Sacrococcygeal teratomas are the most common congenital neonatal tumor. SCTs are more common in females. The tumor is developes at the base of spine in the coccyx region from pluripotent cells. The vast majority of SCTs is diagnosed in utero or at birth and are most commonly benign (90-90%) mature and immature teratomas. Detection outside the newborn period is more commonly associated with malignancy.

Other sites for extra gonadal GCTs are mediastinal and retroperitoneum and vaginal and uterine cavity and some rare sites.

Pediatric Germ Cell Tumors; A 10-year Experience

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Abstract

Background:

The aim of this study was to evaluate the outcome of germ cell tumors in patients admitted to our center during a ten year period.

Methods:

In a retrospective descriptive study, patients with the pathological diagnosis of germ cell tumor (GCT) were included. All records were evaluated and patients followed by personal visit in clinic or phone call. Data regarding age, sex, tumor site, bio-chemical assay, pathology, treatment and outcomes were gathered. For qualitative variables we computed frequency and percentage and for quantitative variables, mean and standard deviation. Survival analysis was performed using Kaplan-Meier. All statistical analyses were performed by SPSS version16.0.

Methods:

Forty four patients consisted of 32 girls (72.7%) and 12 boys (27.3%). Their median age was 23 months. The most common pathological tumor types were 18 (40.9%) mature teratomas and 14 (31.8%) yolk sac tumors. Extra gonadal tumors were more prevalent (32 cases) and consisted of 21 (47.7%) sacrcoccygeal, 7 (15.9%) retroperitoneal, 2 (4.4%) mediastinal and 2 (4.4%) cervical tumors. In gonadal tumors 9 patients had ovarian and 3 patients' testicular involvement. Staging at the time of diagnosis revealed stage one in 23 (52.3%) cases. All patients were treated surgically and the most common procedure was total resection in 41 (93.2%) patients. Fifteen (34.1%) patients received chemotherapy. In follow-up 31 (77.5%) patients were in complete remission, 9 (22.5%) had died, and 4 cases did not appear to follow-up visits. The median survival was 16 months (IQR 4-49 months). The highest mortality rate was found in patients with yolk sac tumors (8 of 13 cases).

Conclusion:

The patients with extra-gonadal GCT and a high AFP level have the worst prognosis and lower survival rate. Combination of surgery and chemotherapy can lead to a better prognosis.

Sacrococcygeal Teratoma: a 20-Year Single-center Experience Report

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Background:

Sacrococcygeal teratoma (SCT) is a relatively rare surgical concern in the pediatric population. Due to the limited number of reported cases and the lack of large-scale studies, the diagnosis, management, and prognosis of SCT are not well understood. In this report, we present a retrospective review of over 20 years of SCT management at a single center.

Methods:

We retrospectively reviewed all children who underwent surgical treatment for SCT between 2011 and 2023. Demographic information, prenatal diagnosis, age at diagnosis, primary treatment, recurrence rates, and outcomes were analyzed.

Results:

A total of 25 children, including 18 females (72%) and seven males (28%), were included in the study. The average age at diagnosis was 22.5 ± 21.6 months, and only one patient (4%) was diagnosed prenatally. The mean age of surgical intervention was 23.0 ± 29.2 months, which was significantly correlated with the age at diagnosis ($p < 0.001$, $r = 0.985$). Mature teratomas were the most common type of tumor (52%), followed by yolk sac tumors (24%) and germ cell tumors (12%). Moreover, 13 (52%) of patients treated with surgical resection alone, while seven (28%) received chemotherapy after surgery. Also, 5 (20%) of cases were treated with chemotherapy without surgical intervention. Averagely, the patients followed up for 24.8 months after the initial diagnosis. Recurrence and mortality rates were 32% and 4% respectively.

Conclusion:

Our study provides valuable insights into the management of sacrococcygeal teratoma (SCT) in pediatric patients. Mature teratomas were the most common type of tumor observed in our study. Surgical resection remains the primary treatment modality for SCT, with chemotherapy being used as an adjunct therapy in some cases. While the overall recurrence rate was relatively high, early mortality was rare, emphasizing the importance of long-term follow-up in these patients. Our findings highlight the need for continued research into SCT management to improve outcomes for affected children.

Keywords: Sacrococcygeal Teratoma; Pediatric Surgery; Surgical Resection; Chemotherapy; Recurrence

Sacrococcygeal Teratoma Presenting with Irritability & Urine Retention in a Female Infant

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Background:

Sacrococcygeal teratoma (SCT) is the commonest tumor in neonates. SCT type IV presents a diagnostic problem as it is often concealed so that its symptoms are evident only if it causes a mass effect on adjacent organs. Urinary retention is a common presentation. The aim of this presentation is to report a case of acute urinary retention in an infant with SCT type IV which made a diagnostic challenge because of its concealed nature and emphasize on the importance of early diagnosis and management by having a high index of suspicion in such cases during early physical examination since in addition to potential of malignancy it can cause urinary tract obstruction leading to permanent damage to the kidneys.

Case Presentation:

We report a 2-month-old female infant presented with irritability, abdominal distention, and difficulty in passing urine and stool. She had been admitted and worked-up by our pediatrician colleagues whose physical exam and also sonographic evaluation did not reveal any significant findings except for overdistended bladder and mild bilateral hydronephrosis. She had developed rising blood urea nitrogen and serum creatinine, which has normalized after urethral catheterization and relief of the urinary obstruction. On pediatric surgery consultation, the infant was noted to have a presacral mass that was palpable on DRE and confirmed by an MRI scan. A combined abdominal and posterior sacral approach was used for the resection of the tumor with complete resolution of symptoms.

Conclusion:

Sacrococcygeal teratoma is the most common tumor that occurs in newborns. Its clinical presentation varies depending on the size and location of the tumor. There must be a high index of suspicion during the physical exam of infants presenting with obstructive urinary or bowel symptoms to rule-out a concealed, non- clinically apparent type IV SCTs, since early diagnosis and treatment are essential for a good prognosis.

Keywords: Sacrococcygeal Teratoma, Urinary Retention, Irritability, Physical Exam

Type III Pleuropulmonary Blastoma in a 3-Year-Old Girl: A Case Report

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Abstract

Pleuropulmonary blastoma (PPB) is a rare malignant embryonal tumor of the lungs that appears to arise during organ development and is usually diagnosed in the pediatric population under the age of 5 years. Pleuropulmonary blastomas fall under the larger aegis of the DICER1-related disorders. It has poor prognosis with three different subtypes: cystic [type I], combined cystic and solid [type II] and solid [type III]. Children with PPB often present with nonspecific symptoms, and the primary treatment include surgery and chemotherapy. In this report, a 3-years-old girl with a history of outpatient treatment for infections from two weeks ago with fever and shortness of breath went to the emergency ward of Abuzar Hospital in Ahvaz, and due to the reduction of pulmonary thorax and air fluid level in chest radiography and CT scan, she underwent a tube thoracostomy, and drainage of two liters of pus. Finally, the patient underwent a left thoracotomy, and complete necrosis of the left lung and multiple cystic lesions were evident. A total left pneumonectomy was performed and after a few days the patient was discharged with a good general condition. One month after discharge, she returned with severe shortness of breath and left pneumothorax. Then, in the emergency ward, she underwent a tube thoracostomy, and then a thoracotomy, where a very fragile and bleeding vegetative mass was seen in the pulmonary hilum, and it was not possible to complete resection. The biopsy specimen was removed and the bronchial stump was covered with an intercostal muscle flap. With definitive pathology and IHC, both surgeries reported type III pleuropulmonary blastoma. The child was a candidate for chemotherapy, which according to the delayed chest CT (three weeks after the last operation) showed involvement and seeding of the entire left hemithorax. Chemotherapy was considered palliative.

Keywords: Pleuropulmonary Blastoma, Chemotherapy, DICER1, Thoracostomy, Pediatric

Blindness as the First Presentation of ALL in a Child with Nasopharyngeal Mass

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Introduction:

A wide range of masses develop in the nose, nasal cavity, and nasopharynx in children. These lesions may be benign or malignant. The most cause for malignant masses in these regions are rhabdomyosarcoma and nasopharyngeal carcinoma. Optic nerve infiltration by leukemic cells is very rare in pediatric ALL (acute lymphoblastic leukemia). Blindness due to optic nerve involvement as the first presentation in these patients is extremely rare. About 3.6% of children with ALL could present with eye symptoms.

Case report:

We describe an atypical manifestation of ALL in a child with nasopharyngeal mass and blindness from three month ago. Radiologic imaging shows a large mass in nasopharynx with abnormal signal in sphenoidal bone and involvement of Sella and suprasellar region in favor of RMS or metastatic neuroblastoma. Endoscopic biopsy of nasopharyngeal mass was performed for three times and the pathology report was chronic inflammation. First CBC and bone marrow aspiration was perfect and there was no involvement in reticuloendothelial system. Chest and abdominal CT Scan was normal and there was no increase in urine methanephrins level. Ophthalmologic findings were pallor in head of bilateral optic nerves and no evidence of optic nerve involvement in orbital MRI was detected. EBV IgM level was upper than normal range but EBV PCR (polymerase chain reaction) was negative in histologic sample of the mass. Three weeks later, the CBC show bicytopenia and the spleen size was enlarged. BMA repeated and the diagnosis was acute lymphoblastic leukemia. First CSF (cerebrospinal fluid) was normal. Immunological evaluations were done. Chemotherapy started for him but the nasopharyngeal mass was completely resolves before starting chemotherapy. Sphenoidal bone involvement decrease after six-month chemotherapy but not completely resolves. There was no change in visual acuity after three month of starting treatment.

Conclusion:

ALL can initially present with ocular symptoms without any changes in CBC or bone marrow aspiration.

Dumble Cystic Lymphangioma as an Underlying Cause of Vague Abdominal Complaints in Pediatrics

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Abstract

Introduction:

Lymphatic Malformations (LM) refer to rare hamartomatous benign lymphatic ectasias with an incidence of 1: 250,000. They almost involve children more than adults, and among them, infants under one age are mostly affected. Macrocytic LM is found to be more than 2 cm in diameter or 2×2 cm^2 in volume. The proper treatment for mesenteric LM is complete surgical excision unless there is vital structure involvement.

Case Presentation:

We report a mesenteric macrocytic LM in a 2-year-old girl complaining of vague abdominal discomfort and persistent vomiting, in which ultrasonography revealed a cystic mass with seromucous components. She then underwent an exploratory laparotomy. The operation and the follow-up duration were uneventful.

Discussion:

LMs are rare benign lesions of vascular origin with lymphatic differentiation, according to the latest International Society for the Study of Vascular Anomalies (ISSVA 2018). Under light microscopy, their thin-walled endothelium and lymphatic tissue characterize these malformations.

These mobile lesions are incidentally found or appear with intestinal obstruction or acute abdomen scenarios.

Conclusion:

Although benign, the LMs have the potential for invasion and recurrence. Thus, the examiner physician must keep such intra-abdominal lesions in mind.

Myxoid Liposarcoma in the Extremities: A Case Report

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Abstract

Liposarcoma is among the most common sarcomas in the adult population. Conversely, its incidence in the pediatric population is rare. The most common subtype affecting the extremities is myxoid liposarcoma. The preferred method of surgery for limb myxoid liposarcoma is limb-preserving surgery.

In the current study, we report on a 4-year-old boy who was referred to our department with a chief complaint of a gradual increase in mass located at the lateral aspect of his left arm for 6 months.

The primary physical examination revealed a 5 cm by 6 cm firm mass at the lateral aspect of the deltoid muscle and 3 - 4 discrete lymphadenopathies in the left axillary area.

T1-weighted magnetic resonance imaging (MRI) delineated a multiloculated cystic mass with fine septa and circumscribed borders.

In a single procedure, the patient underwent radical resection. The mass was described intraoperatively as having a multinodular lipoid texture and measuring 4 cm to 5 cm without vascular involvement. The microscopic assessment of the mass elucidated uniformly round and oval-shaped cellular sheets in the myxoid. Approximately 20% of the cells were rounded. The hyperchromatic nucleus with the capillary network was notable. The immunohistochemistry assessment was positive for S100, Vimentin, CD34, and CD31, while Cytokeratin and Desmin markers were negative. Ultimately, the pathological evaluation suggested myxoid liposarcoma.

For follow-up, a biennial chest X-ray and whole-body computed tomography scan are performed during the first year and annually for the next three years. The oncology team administered the consequent treatment.

Congenital Rhabdomyosarcoma of Shoulder

Khaleghnejad Tabari A, Mirshemirani A, Rouzrokh M, Nariman Sh, Hassas Yeganeh Sh, Gharib A, Khaleghnejad Tabari N, **Behnia N**

Abstract

A 16-day-old female was referred with congenital swelling on her right shoulder. On examination, there was a hard, round, ecchymotic, nontender, slightly movable, warm and shiny 10x15 cm mass on the right axillary pits which was extended to the right side of neck and chest wall. The mass separated the shoulder from the chest wall causing paralysis of right hand. Chest X-ray, ultrasound and MRI with contrast demonstrated a soft tissue mass suspected to be a hemangioma. The mass rapidly increased in size despite aggressive steroid therapy with rupture and bleeding. On the 45th post natal day the baby was taken to operating room to control the bleeding and if possible total excision of the mass. The mass was separated easily from the surrounding tissue and was excised along with right upper extremity. At the end of surgery the baby had cardiac arrest, and apparently died of Disseminated Intravascular Coagulation (DIC). The final pathology report was Rhabdomyosarcoma (RMS).

Keywords: Rhabdomyosarcoma; Congenital; Newborn; Shoulder

Huge Intrathoracic Neurofibromatosis-1AssociatedMalignant PeripheralNerveSheathTumor ina10Year'sOldGirl,"AcaseReport"

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Background:

Neurofibromatosis 1 (NF1) is an autosomal dominant tumor predisposition syndrome in which affected individuals have a greatly increased risk of developing malignant peripheral nerve sheath tumors (MPNSTs). These cancers are difficult to detect and have a poor prognosis. They commonly invade axial sites and rarely do they occur in the thorax. Herein, we present the case of an enormous multilobulated intrathoracic malignant peripheral nerve sheath tumor that was successfully resected for palliative purpose.

Methods:

The patient presented with only diminished tolerance to physical activity with no other obvious symptoms. Standard chest radiography revealed a well-defined opacity of subcostal intensity, occupying two thirds of the right hemithorax, forming a common body with the mediastinal shadow. Thoracic computed tomography (CT) identified a 21/11 cm solid mass that compresses the right lung and the right main bronchus with both a solid component and a central liquid area. Open surgery was performed in order to remove the tumor, which was 20.5/12.5/9 cm in size and weighed 1,830 g, well defined, with no invasion of the adjacent organs, having a solid-fibromatous aspect as well as a central necrotic area. The origin of the tumor was confirmed from the posterolateral part of the forth intercostal nerve. Pathology examination and immunohistochemistry confirmed the diagnosis of a benign Schwannoma.

Results:

The patient underwent Rt. Posterolateral thoracotomy that well tolerated, and is asymptomatic in post-operative period for 3 months.

Discussion:

Benign intrathoracic Schwannomas are asymptomatic for long periods and the main therapeutic option is complete surgical resection. The surgical approach, either open or video-assisted is dictated by the localisation of the tumor, local extension and most importantly the size of the neurogenic mass.

A Unique Presentation of Chondrosarcoma in Soft Tissue

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Abstract

Background:

Chondrosarcoma is a collective term for a group of tumors that consist predominantly of cartilage and that range from low-grade tumors with low metastatic potential to high-grade, aggressive tumors characterized by early metastasis. The typical appearance of a dedifferentiated chondrosarcoma is an area of punctuate opacities surrounded by a permeating, destructive lytic lesion.

Methods:

We evaluated a 4 years old girl which had been referred to us with pubic area induration. Imaging studies revealed a heterogenous hypo-echo mass in pubic and another soft tissue mass located in labia. Bx was done and the pathologic features was compatible with enchondroma. Radical resection of the mass was accomplished and the permanent histologic and IHC findings was in favor of chondrosarcoma Infiltrative chordroid neoplasm with atypia and foci of myxoid changes was found in soft tissue mass.

Conclusion:

Despite various investigations, it may be difficult to differentiate a benign cartilage lesion from a slowgrowing, low-grade chondrosarcoma. Secondary chondrosarcoma can occur in a previously benign cartilaginous lesion.

تجربه درمان پالیاتیو سرطان بزرگسالان و اجرایی کردن آن برای کودکان

Abolghasemi H

مراقبت تسکینی حمایتی به مجموعه مراقبت هایی اطلاق می گردد که به منظور بهبود کیفیت زندگی بیمارانی که به یک بیماری جدی یا تهدید کننده زندگی مانند سرطان مبتلا هستند، انجام می شود. مراقبت تسکینی رویکردی برای مراقبت است که شخص را به عنوان یک کل مخاطب قرار می دهد. هدف از این رویکرد، کمک به درمان، در کوتاه ترین زمان ممکن، رفع علائم و عوارض جانبی بیماری و درمان آن، علاوه بر آن رسیدگی به هر گونه مشکلات روانشناختی، اجتماعی و عاطفی در بیمار و خانواده اوست. مراقبت تسکینی را مراقبت راحت، مراقبت حمایتی و کنترل علائم نیز می نامند. بیماران ممکن است در بیمارستان، یک کلینیک سرپایی، یک مرکز مراقبت طولانی مدت یا در خانه تحت نظر پزشک از مراقبت تسکینی برخوردار شوند. در وضعیت کنونی که درمان سرطان توسط بیمه ها مورد حمایت میباشد، مراقبت های تسکینی تا حدودی در قالب خدمات درمانی به بیمار ارائه میشود، اما نقایص زیادی در این خدمات وجود دارد. لذا از یک دهه پیش موسسه کنترل سطان ایرانیان یا مکسا قبول نمود این وظیفه به جا مانده را که اکثر خدمات آن تحت حمایت بیمه ها و دولت نیست به صورت کاملاً رایگان پوشش دهد؛ لذا شعباتی در اصفهان و تهران تاسیس نمود و بعدها آن را به چند شهر دیگر گسترش داد. خدمات متعدد از جمله مهمترین آنها خدمات تسکینی در منزل میباشد که با مراقبت های پایان حیات به بیماران توانسته است بطور قابل توجهی بار مراجعه به واحدهای مراقبت ویژه را کم کند. علیرغم وجود خیریه محک که امور زمین مانده کودکان سرطانی را پوشش میدهد خدمات حمایتی تسکینی کودکان کماکان بدون پوشش است و انتظار میرود با هماهنگی که بین موسسه خیریه محک و مکسا برقرار میشود این امر مهم که نقش اساسی در کیفیت زندگی کودکان سرطانی دارد در کشور اجرایی شود.

Epidemiological Aspects, Clinical Features and Treatment Outcome in Children Suffering Hepatoblastoma

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Introduction:

Hepatoblastoma (HBL) is the most common primary liver tumor within childhood. Entire tumor resection is basis for ultimate cure for HBL and supports the only pleasant chance of long-standing diseasefree survival.

Objectives:

In the current survey, we attempted to evaluate the long-term survival of children with HBL with surgical resection in a referral children hospital in Iran within the last decade.

Materials and Methods:

This retrospective descriptive study was conducted on all children who suffered HBL and undergone surgery between 2006 and 2016. Reviewing the recorded hospital files led to a sample of 30 eligible patients. The baseline characteristics of the patients were all collected by reviewing the files.

Results:

In total, 30 consecutive children (21 male and 9 female) suffering HBL were described. Of those, 40.0% aged less than 12 months and only 6.7% aged higher than 36 months. The most common clinical manifestations were asymptomatic abdominal mass (in 66.7%) followed by fever (in 10.0%) and pain (in 10.0%). In more than half of the patients (53.3%), right lobe involved, while left lobe involved in 16.7%. based on histological report, marginal involvement was found in 75.0% of children

Evaluation of Hepatoblastoma in a 7 Months Old Infant (Case Report)

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Background:

Hepatoblastoma is the most common form of liver cancer in children. The disease usually affects children younger than 3 years. Hepatoblastoma is usually diagnosed as an asymptomatic abdominal mass. Surgical techniques and adjuvant chemotherapy have markedly improved the prognosis of patients with hepatoblastoma.

Methods:

We evaluated a 7 months old infant with abdominal distention and gastroenteritis. Imaging studies revealed a solid mass in right liver lobe which turned out to be hepatoblastoma using incisional Bx. After neoadjuvant chemotherapy, the patient undergone Laparotomy + Right liver lobectomy + Cholecystectomy.

Results:

The permanent tissue diagnosis turned out to be poorly differentiated neuroblastoma instead of hepatoblastoma.

Discussion:

Signs and symptoms of neuroblastoma vary with site of presentation. Generally, symptoms include abdominal pain, emesis, weight loss, anorexia, fatigue, and bone pain. Chronic diarrhea is a rare presenting symptom secondary to tumor secretion of vasoactive intestinal peptide secretion. Approximately two thirds of patients with neuroblastoma have abdominal primaries. In these circumstances, patients can present with an asymptomatic abdominal mass that usually is discovered by the parents or a caregiver. Symptoms produced by the presence of the mass depend on its proximity to vital structures and usually progress over time.

Hematopoietic Stem Cell Transplantation (HSCT) in Solid Tumors of Children

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Abstract

Bone Marrow Transplantation (BMT) is called, recently as Hematopoietic Stem Cell Transplantation - HSCT (with introduction of other Stem Cell Sources including peripheral Blood Stem Cell (PBSC) & Cord blood (CB). Now HSCT is a part of the treatment protocols in some of the disease or the only treatment option in some others. EBMT Survey for 32 years (1990- 2021) reported that patients receiving more than 850,000 transplants.

Indications of HSCT in Children

- Heriditary, Aqcuired
- Malignant disorders (Hematologic & solid tumor)
- Non Malignant: Hematologic, Non Hematologic

Type of HSCT:

- ALLOGENIC; Cells from another person (Sibling, MRD, Unrelated Donor, Parent or relative)
- AUTOLOGOUS: (Own cells)
- SYNGENIC: Identical Twin

Sources of HSCT

- Bone Marrow (BM)
- peripheral Blood (PB)
- Cord Blood (CB)

Solid tumors are about 60% of pediatric neoplasms. During last years there are specific progress in diagnosis & management of this group of neoplasm, duo to understanding of prognostically important biological and clinical features.

In solid tumors more effective treatment, include often a combination of Chemotherapy, Surgery, & Radiation therapy with 5 year EFS about 70%. Some of the patients in solid tumor group don't have response to this treatment, and over the last 3 decades accepted treatment modality, include Consolidation (High dose Chemotherapy & Autologous HSCT) in children with high risk solid tumors; in CR / VGPR / PR or relapsed after conventional chemo regimen is used. Auto HSCT as stem cell rescue allows for escalation of chemotherapy doses above those limited by myeloablation.

The most common solid tumors which are candidate for Consolidation include:

- Neuroblastoma Stage IV, Stage 3 NMYC Positive…
- Relapsed / Refractory; Ewing Sarcoma
- Germ cell tumors
- Brain Tumors; (Medulloblastoma…)

No other extracranial solid tumor indication of ASCT has been established as standard management for high risk cases. Studies show that in tandem HSCT (such as double HSCT in neuroblastoma) there is no advantage over single HDT/HSCT. Also the results of Allogenic HSCT in solid tumors is Experimental, investigational, and / or unproven.

It is important that using of High dose Chemotherapy & Autologous HSCT in children of solid tumors except of high risk Neuroblastoma, needs Carefull assessment of RISK & BENEFIT. Future directions will likely include combinations of therapies, biomarkers to assess success, the creation of more antibody-drug conjugates, and further breakdown of regulatory barriers.

Survey on Childhood Solid Malignant Tumors in Cases Admitted to Mofid Pediatric Hospital from 1996-2022: a Single-Center Study

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Background:

As children comprise a considerable proportion of our population, the importance of local epidemiologic research and geographic and racial differences can't be disputed on childhood malignancies.

Methods:

In this descriptive retrospective study, we extensively reviewed the medical records of patients younger than 27 years of age, diagnosed with solid malignant tumors, from 1996 to 2020, using the last version of International Classification of Childhood Cancers.

Results:

In our study the order of incidence of solid malignancies was relatively similar to the other national studies, with lymphomas and Central Nervous System (CNS) tumors as the most common, followed by Sympathetic Nervous System (SNS) tumors, soft tissue sarcomas and renal tumors. The peak age of diagnosis was between 1 and 4 years old. In our study, the overall male to female ratio was 1.38, with a trend towards male dominance in the older age groups. We also observed a disturbing trend of childhood solid malignancies. The total number of cases almost doubled from 2009 [54 (6.9%) to 2010 (96) (12.2%)] .This trend was particularly detected in CNS and SNS tumors. Further analysis showed that malignant CNS tumors had played a more pronounced role in this change. We added the result of further 12 years (to 2020) to the previous study.

Discussion:

Changes in trends of some tumor categories have illustrated a desperate need to further research in regional and national levels. Also the gathered data can be used to make more accurate programs for a better control of cancer and to help policymakers to allocate more evidence-based resource for hospitals.

ملانوم در کودکان

(عوامل خطر، پیش آگهی، تشخیص و تازه‌های درمان)

دکتر مهرداد حسین پور

دانشگاه علوم پزشکی اصفهان

مقدمه:

ملانوما هر چند جزو سرطان‌های ناشایع در کودکان محسوب می‌شود، لیکن شایعترین سرطان پوست در کودکان می‌باشد و در حدود چهار درصد از موارد سرطان در سنین پانزده تا نوزده سال را در بر می‌گیرد.

روش و مواد:

در این مطالعه مروری با توجه به ناشایع بودن بیماری بررسی عوامل خطر، روش‌های تشخیص و تازه‌های درمان به تفصیل مورد بررسی قرار گرفته است.

نتایج:

عوامل خطر شامل خال ملانوسیتیک بزرگ، گزرودرما پیگمانتوزوم، نقص‌های ایمنی، رتینوبلاستوم مادرزادی، نژاد بور و مو قرمز، تعداد بالای خال‌های ملانوسیتیک می‌باشند.

عوامل پیش آگهی شامل سن، مرحله بیماری، سندرمیک بودن بیماری، محل ملانوم می‌باشد.

روش‌های تشخیصی شامل نمونه برداری از محل و بررسی غدد لنفاوی ناحیه بوده و روش‌های درمان نیز جراحی و استفاده از شیمی درمانی با دستورالعمل‌های جدید می‌باشد.

51

نتیجه‌گیری:

درمان ملانوم در کودکان با توجه به ناشایع بودن آن باید بر اساس آخرین دستورالعمل‌های به روز موجود در دنیا انجام گیرد.

Abdominal Basidiobo; Omycosis, an Abdominal Fungal Mass, Mimics Malignancy, Leads to Whipple's Procedure and Right Hemicolectomy, "A case report"

Tadayon A

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Background:

Basidiobolus ranarum is a fungus found in the dung of amphibians, reptiles and insectivorous bats. Basidiobolomycosis is a chronic subcutaneous infection of the trunk and limbs caused by B. ranarum. The disease is a well-known infection in the tropical areas. It usually presents with subcutaneous or gastrointestinal lesions and rarely with systemic affection. Recently, the etiologic role of B. ranarum in gastrointestinal infections has been increasingly recognized.

Methods:

Here, we retrospectively reviewed the records of one patient with aggressive gastrointestinal basidiobolomycosis, from sowtern of Iran.

Results:

The patient presented with prolonged fever and other manifestations suggestive of either chronic infection (such as tuberculosis) or malignancies (such as lymphoma). The diagnosis of gastrointestinal basidiobolomycosis was established on histological ground (granulomatous reaction, dense infiltrate of eosinophils and fungal structures). Primary resection of mass was impossible, at first she was treated by Itraconazole, KI and Amphotericin-B for about 3 months and then surgical resection performed (Whipple's procedure and right hemicolectomy).

Discussion:

Gastrointestinal basidiobolomycosis is often misdiagnosed as cancer (lymphoma or carcinoma), tuberculosis or inflammatory bowel disease. Its recognition needs high index of suspicion and increased awareness especially in patients with chest, abdominal or neck masses and eosinophilia. The diagnosis of basidiobolomycosis can be established on histological basis in most cases. The fungal morphology and the Splendore – Hoeppli phenomenon are characteristic histological features of this condition. There are no prominent risk factors. Usually, surgery and prolonged antifungal therapy are required.