论 著

儿童肾恶性横纹肌样瘤的CT、MRI表现

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【摘要】 **目的** 探讨儿童肾恶性横纹肌样瘤的CT、MRI表现，以提高该病的影像诊断水平。**方法** 回顾性分析上海市儿童医院2008年5月~2021年3月经病理证实8例肾恶性横纹肌样瘤的患者资料。7例患儿行CT平扫及增强检查，4例患儿行磁共振平扫及增强检查，观察分析肾恶性横纹肌样瘤体的部位、形态、大小、密度/信号特征、强化方式以及转移等。**结果** 6例位于左肾，2例位于右肾，2例病变呈圆形或类圆形，6例为不规则形，6例累及肾窦、肾盂。CT平扫中5例呈稍高密度，2例呈等低密度，肿瘤最大径平均约8.6cm；2例合并出血，3例合并包膜下积液/积血，8例均合并囊变坏死。MRI平扫中4例T_1WI呈低信号为主，3例T_2WI呈稍高信号，1例T_2WI呈混杂信号。2例病变DWI均呈明显高信号、相应ADC为低信号，提示病变弥散受限。增强后呈不均匀轻度强化。5例伴转移。**结论** 儿童肾恶性横纹肌样瘤CT、MRI表现有特征性，儿童肾实质内巨大肿块，合并囊变坏死、出血，集合系统受累，伴有包膜下积液，弥散受限，增强扫描实性成分轻度强化，常合并转移，应高度警惕肾恶性横纹肌样瘤的可能。

【关键词】儿童；计算机断层扫描；磁共振；肾恶性横纹肌样瘤
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CT and MRI Imaging Findings of Malignant Rhabdoid Tumor of the Kidney in Children

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ABSTRACT

Objective To investigate the CT and MR imaging features of Children Malignant rhabdoid tumor of kidney(MRTK)in children. ***Methods*** 8 cases of MRTK confirmed pathologically in Shanghai Children's Hospital from May 2008 to March 2021 were analyzed retrospectively. All 7 cases underwent plain and enhanced CT and 4 cases underwent plain and enhanced MRI. We observed and analyzed the location,shape, size, density /signal, enhancement pattem, and the metastasis of the tumor. ***Results*** 6 cases occurred in the left kidney and 2 cases in the right kidney. 2 cases in MRTK were round or ellipse and 6 cases was irregular shape. Renal pelvis was invaded in 6 cases. 5 cases were iso-or slightly hyper-density and 2 cases such as density on CT scans, The tumor average maximum diameter is about 8.6cm; 2 cases with bleeding. The renal capsule/sub capsule effusion/hemorrhage was in 3 cases, 8 cases were merged cystic necrosis. T_1WI signals of 4 cases were low or equal to the renal cortex on MRI scans, T_2WI signals of 3 cases were high or equal to the renal cortex and 1case was mixed signal. 2cases with high signal on DWI and low signal on ADC. All cases were slightly enhanced. 5 cases with metastasis. ***Conclusion*** MRTK have characteristic imaging findings, especially Children in the renal parenchyma mass, including the lesion with Punctate and patchy calcifications. With merge bleeding, high signal on DWI and low signal on ADC, with mild to moderate enhancement. May need to be mindful of the disease.

Keywords: Children; Tomography Computed; Magnetic Resonance Imaging; Malignant Rhabdoid Tumor of Kidney

肾恶性横纹肌样瘤(malignant rhabdoid tumor of kidney，MRTK)是一种罕见的高度恶性的肿瘤，是来源于肾脏的组织学形态为横纹肌样的恶性肿瘤，其细胞起源及发病机制尚不明确，多数学者认为其起源于肾髓质的某种原始细胞，其遗传学特征为22号染色体长臂上SMA RCBI/INI基因缺失或突变，检出基因产物INI-1蛋白表达丧失有助于诊断。仅发生于儿童，主要发生在婴幼儿及青少年[1]，约80%发生在2岁以下患儿，侵袭性较强，预后差[2]。本文回顾性分析8例经病理证实的MRTK的临床及CT、MRI特点，结合相关文献，探讨其特征性的CT、MRI表现，提高对该病的认识，以助于术前诊断，为临床治疗提供参考。

1 资料与方法

1.1 研究对象 收集上海市儿童医院2008年5月至2021年3月经病理证实8例MRTK患者资料，其中女4例，男4例，年龄4个月~14岁，平均年龄2.8岁。临床表现包括4例血尿、2例血尿伴发热、2例腹部包块就诊。术前患者影像学检查为7例行CT平扫及增强检查，4例行MRI平扫及增强检查。8例患者住院期间均接受根治性手术，完整切除病灶后病理诊断为MRTK。

1.2 检查方法 患儿行CT及MRI检查，对于3岁以下儿童检查前禁食3-4h，患儿口服水合氯醛，按照0.5mL/kg体质量镇静，待患儿安静入睡后开始检查。CT采用GE Light Speed VCT及TOSHIBA Aquilion 64CT检查。扫描参数及范围：管电流10mA，管电压800kV；肝下缘至盆腔。增强扫描采用非离子型造影剂碘佛醇，剂量1.5mL/kg/bw。MRI采用Philips Ingenia3.0 T MRI仪器扫描，行轴位、矢状位、冠状位肾脏扫描。扫描包括轴位T_1加权像(T_1 weighted imaging)、T_2加权像(T_2 weighted imaging)、脂肪抑制T_2WI序列及DWI(diffusion-weighted imaging)序列，增强扫描采用钆喷替酸葡甲胺(gadopentetate dimeglumine, Gd-DTPA)，剂量0.2mL/kg体质量，行T_1WI增强轴位、矢状位、冠状位扫描。

1.3 图像分析 由两位高年资的影像科医师阅片，采用双盲法进行观察，并记录肿瘤的位置、数目、最大径、形态、实性成分密度/信号(实性成分与正常肾实质对比)、病灶成分(是否伴钙化、出血)、包膜下积液/积血、强化程度、肾周组织侵犯、有无血管侵犯、转移等。意见不一致时协商达成一致。

2 结 果

2.1 影像学表现 8例MRTK均为单侧病灶，发生于右肾2例及左肾6例；全部起源于肾实质，均靠近肾门，6例累及肾窦、肾盂。CT平扫病变中5例呈稍高密度，2例呈等低密度，肿瘤最大横断面直径约3.1~17.4cm，平均约8.6cm；2例病变呈圆形或类圆形，6例病变呈不规则形，并可见分叶。3例可见包膜下积液/积血，呈新月形。8例均有囊变坏

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死，均无钙化。1例病灶内含有少许脂肪组织；2例肿瘤内合并出血。增强扫描可见肿块呈不均匀轻度强化，实性成分强化程度低于肾实质，瘤内坏死、囊壁及血肿区无强化。5例合并转移，1例下腔静脉合并癌栓，2例伴有腹膜后淋巴结转移，1例肺转移且肾静脉合并癌栓，表现为脑或肺内多个结节灶，1例随访1年后出现脑及肺转移。

4例MRI表现：与CT比较，显示肿块轮廓更清楚，T_2WI呈稍高信号3例，T_2WI混杂信号1例，T_1WI均呈低信号为主，2例DWI呈高信号、ADC呈低信号，提示病灶弥散受限。增强扫描肿瘤呈不均匀轻度强化，与CT相比，肿瘤实性部分强化更显著，但仍显著低于正常肾实质。

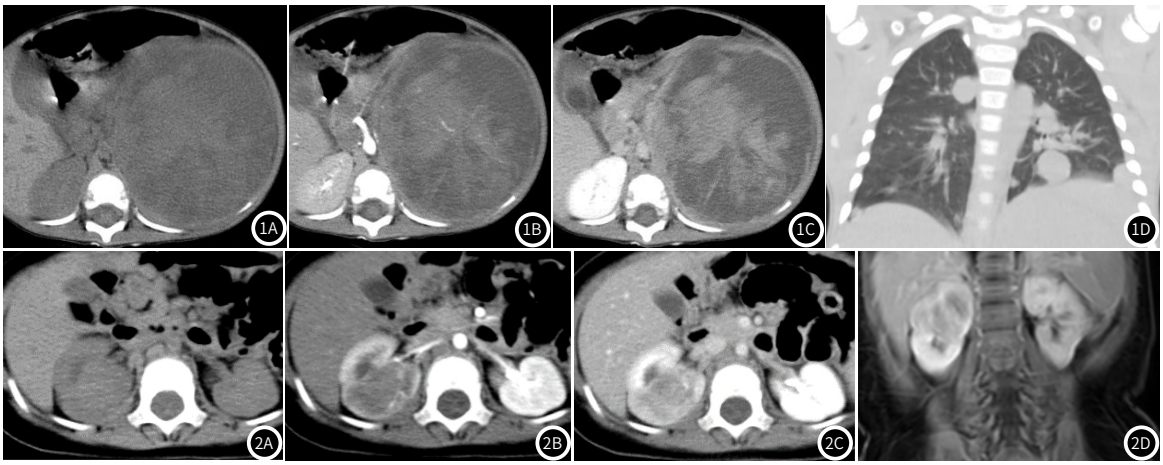

图1A-图1D 病例1，男，2岁，发现腹部包块6天。图1A示CT平扫病灶中心呈稍高密度且边缘可见低密度影；图1B、1C示，CT增强扫描可见病灶内实性成分逐渐强化，强化程度低于肾实质强化，呈轻度强化；图1D示双肺结节；病理示INI1缺失性肿瘤，肾恶性横纹肌样瘤。

图2A-图2D 病例2，女，9个月，血尿5天，图2A示CT平扫呈等密度；图2B、2C示，CT增强扫描可见病灶内实性成分逐渐强化，强化程度低于肾实质强化，呈轻度强化；图2D示增强扫描可见轻度强化；病理示INI1缺失性肿瘤，肾恶性横纹肌样瘤。

2.2 病理学检查 镜下：见纤维组织增生，其内见片状分布的肿瘤细胞，异型性明显，胞浆丰富嗜酸性、可见包涵体，核大、核仁明显。伴见大片坏死。肾盂及肾窦见肿瘤侵犯，肾窦脉管内见瘤栓。输尿管切端未见肿瘤侵犯。本组病例免疫组织化学INI-1阴性，提示INI-1基因突变或缺失。诊断：INI1缺失性肿瘤，肾恶性横纹肌样瘤。

3 讨　论

3.1 临床表现 MRTK好发于儿童，平均发病年龄13个月，男多于女[3]。本组病例中患者均为儿童，4个月~14岁，平均年龄2.8岁，中位年龄14个月，符合文献报道。本组病例中男女比例为1：1，与文献报道不符，可能与样本量较少有关[4]。临床表现缺少特征性表现，主要表现为腹部包块、血尿或腹痛伴发热就诊[5]，与本组病例基本相符。本组病例中4例出现肉眼血尿。儿童期MRTK呈高侵袭性、进展迅速、预后差、死亡率高、易转移。MRTK常伴发脑及其他部位转移瘤等[6]。本组病例中，出现脑、肺、淋巴结转移及静脉癌栓。

3.2 影像表现 MRTK多为单侧发病，偶可累及双肾，肿瘤体积通常较大，瘤体最大径常大于5cm，呈浸润性生长，易侵犯肾盂肾盏，可推移或包绕肾周血管，可形成静脉瘤栓，甚至延伸入右心房[7]。本组8例均为单侧发病，CT显示瘤体多位于肾脏中央，并紧邻肾门，本组病例中有6例邻近肾门并累及肾集合系统。本组8例肿瘤内均含有低密度液化坏死囊变区，部分肿瘤可见高密度出血区、脂肪变等，均无钙化。出血区往往在肿瘤周围区域，考虑肿瘤出血主要于肿瘤外周分布可能与其周围小血管瘤栓形成有关[8]，并发生坏死。增强扫描后肿瘤呈轻度不均匀强化。

T_1WI均呈低信号，2例未做DWI、ADC检查，2例DWI呈高信号、ADC呈低信号，提示病灶弥散受限。增强扫描肿瘤呈轻度不均匀强化，与CT相比，肿瘤实性部分强化更显著，但仍显著低于正常肾实质。

MRTK在MRI平扫上T_1WI均呈低信号为主，3例T_2WI呈稍高信号，1例T_2WI呈混杂信号。本组病例中DWI均呈高信号，提示该肿瘤恶性程度较高，亦是该肿瘤的一个重要征象[9]。本组病例中3例(占37.5%)合并包膜下积液/积血，可能为肿瘤侵犯肾周间隙、间接提示肿瘤浸润肾包膜下，或肿瘤破裂所致，局部肿瘤细胞出血并积聚于肾包膜下,这与文献报道一致[10]。文献报道，肾包膜下积液/积血见于的MRTK 病例约占70%，是其特征性表现，而在其他儿童肾肿瘤中仅为12%[11]。本组病例中此征象检出率为37.5%，较低于文献报道，可能与病例数较少有关。

肾MRTK转移及术后复发率高。有文献认为约80%的患者会发生转移，最常见于肺，也见于肝、脑、淋巴结和骨骼等；局部可转移至肾周、腹膜后淋巴结[12]。本组中5例(占62.5%)病例合并转移，符合文献报道。肾MRTK转移早且发生率高，有助于与儿童其他肾肿瘤鉴别[13]。另外，有文献报道肾MRT可能合并颅内原发肿瘤，本组病例中未见。肾MRTK短时间内易复发及合并转移，本组8例患儿中共5例发现转移，其中肾门区、腹膜后淋巴结转移3例、下腔静脉内癌栓形成2例，其中1例随访1年后出现转移，说明该类肿瘤具有较强侵袭性。在行肾肿瘤检查时，同时须行胸部及全腹部CT检查，及颅脑CT或MRI检查[14]。

3.3 鉴别诊断 肾恶性横纹肌样瘤需注意与肾脏恶性肿瘤相鉴别，特别需要与儿童好发肿瘤相鉴别，尤其是肾母细胞瘤、肾透明细胞肉瘤、Xp11.2易位性肾癌等[15-18]。

(1)肾母细胞瘤：发生率高，文献报道发病率占90%(儿童肾脏肿瘤中)，常发生于2-5y儿童，临床表现为腹部包块，CT平扫可见肾区软组织肿块影，肿块内可见坏死囊变及出血，偶可见钙化，增强后实性成分多呈明显轻度不均匀强化，肿块与受累肾脏之间呈抱球征，巨大肿块时可跨越中线生长。

(2)肾透明细胞肉瘤：常发生于1-4y儿童，CT表现肿瘤出血、坏死较前者更为明显，钙化较为常见，强化程度较肾母明显，DWI有弥散受限，具有较高侵袭性，颅骨转移较为多见。

(3)Xp11.2易位性肾癌：好发于儿童，发病率达20%~33.3%，女性多见，可见钙化，易见坏死、出血、囊变，短T2信号假包膜提示为该病，DWI呈明显高信号；增强扫描轻-中度强化；易出现淋巴结转移或静脉癌。

综上，若发生在较小儿童中，肾脏肿瘤表现为肿块较大，期内合并囊变、坏死、出血等，伴有肾被膜增厚及肾周积液/出血，

累及肾盂，以及弥散受限，合并肺、肝、脑转移，静脉癌栓形成，提示本病的可能。

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