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儿童肾母细胞瘤合并下腔静脉及心房瘤栓1例并文献复习

孙子莹，马　楠，裴瑜馨，李素萍*

中山大学附属第一医院儿科ICU，广东　广州
Email: *lsping3398@163.com

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摘　要

目的：探讨儿童肾母细胞瘤合并下腔静脉及心房瘤栓的诊断、治疗和预后。方法：回顾分析1例诊断为巨大肾母细胞瘤合并下腔静脉及心房瘤栓患儿的临床资料及诊疗过程，并对相关文献进展进行复习。结果：本例患儿为6岁男童，以腹部膨隆为主要表现，化疗过程中出现心悸、气促，CT提示左肾巨大肿瘤、心房及腔静脉内瘤栓，择期行一期手术，完整切除原发肿瘤后，于体外循环下切开右心房及下腔静脉摘除瘤栓。肿瘤大部分坏死，未能分型，术后病理报告示肾母细胞瘤，术后第2天开始化疗，化疗7个疗程后，放疗12次，随访8个月至今无瘤生存。结论：一期胸腹联合手术并结合化疗及放疗可以有效地治疗儿童肾母细胞瘤合并下腔静脉及心房瘤栓，延长生命，降低死亡率。

关键词

肾母细胞瘤，瘤栓，心房，腔静脉，儿童

Wilms Tumor with Inferior Vena Cava and Atrial Thrombus in Children: A Case Report and Literature Review

Ziying Sun, Nan Ma, Yuxin Pei, Suping Li*

PICU, The First Affiliated Hospital of Sun Yat-sen University, Guangzhou Guangdong
Email: *lsping3398@163.com

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Abstract

Objectives: To investigate the diagnosis, treatment, and follow-up of wilms tumor with cavoatrial

*通讯作者。

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tumor-thrombus in children. Methods: Review of the clinical data of a case diagnosed as giant wilms tumor with inferior vena cava and right heart extension with related literatures. Results: A 6-year-old boy initially presented with distended abdomen, and developed palpitation, short-winded during chemotherapy. A giant tumor was showed in left kidney with atrium and inferior vena cava thrombosis at CT. After the primary tumor complete resected of, the child was removed the inferior vena cava and atrial thrombus through thracic and abdominal approach under cardiopulmonary bypass (CPB) with deep hypothermia and circulatory arrest (DHCA). The tumor with nearly complete necrosis, failed to classify the histological type, was showed Wilms tumor by postoperative pathology. The chemotherapy was started on the next day after operation. Follow-up of 8 months, the boy is still free survival, Combined with Radiotherapy 12 times and 7 courses of chemotherapy after surgery. Conclusions: A thoracoabdominal surgery combined with chemotherapy and radiotherapy can effectively treat Wilms' tumor with inferior vena cava and atrial thrombus in children, prolong life, and reduce mortality.

Keywords

Wilms Tumor, Thrombus, Atrium, Vena Cava, Children

1. 引言

肾母细胞瘤是小儿最常见的恶性肿瘤之一，占小儿所有恶性肿瘤的 6% [1]，容易侵犯血管形成瘤栓[2]，增加治疗难度及风险，国外报道约 1%的肾母细胞瘤患儿其瘤栓可沿血管长入心脏、填塞心腔，但国内少见相关报道。本文针对 1 例存在肿瘤播散、下腔静脉及心房瘤栓的肾母细胞瘤患儿于体外循环下成功实行胸腹联合手术进行个案报道，并通过复习相关文献，以期找出适合此类患儿的围手术期诊断治疗方案，从而降低死亡率，提高无瘤生存率。

2. 临床资料

6 岁男性患儿，腹痛 4 月余、发现左腹部包块 2 月余，CT 示左肾肿物并下腔静脉瘤栓，左肾肿物穿刺活检示：肾母细胞瘤并灶性坏死，因穿刺组织量少、难以行组织学分型。根据 NWTS (National Wilms Tumor Study)分期定为 III 期，术前评估手术困难，先予长春新碱(vincristine, VCR) + 放线菌素 D (actinomycin D, Act-D) + 吡柔比星(theprubicin, THP)化疗 2 疗程，期间患儿肿瘤仍逐渐增长，复查 CT 示肝脏、双肺转移，再次评估 NWTS 分期为 IV 期，予改方案为依托泊苷(Vepeside, VP16) + 异环磷酰胺(Ifosfamide, IFO) + 顺铂(Dichlorodiamineplatinum, DDP)化疗 1 疗程，期间患儿出现心率快、气促、排肉眼血尿，心脏彩超提示右心房肿瘤，考虑瘤栓可能随时脱落堵塞心脏流出道，于 2013 年 10 月转入我院 PICU。查体：恶液质，呼吸 45 次/分，双肺呼吸音粗，无啰音，心尖搏动位于第 3 肋间右胸骨旁线外 0.5 cm，心率 145 次/分，心律齐，心音有力，腹部明显膨隆，左上腹扪及巨大包块，约 20 cm * 24 cm，质中，边界尚清，较为固定，有触痛，因腹部肿物巨大，肝、脾触诊不满意。

入住 PICU 后，密切观察及继续化疗，吸氧、强心、利尿、营养支持等，3 周后复查 CT 示(如图 1 及图 2)：肝、肺转移瘤较前缩小，腹部肿瘤较前无明显改变。在家长的坚决要求下，行胸腹联合手术治疗，术中见瘤体巨大，占据整个腹腔，与周围组织紧密粘连，左肾静脉内可探及瘤栓。将左肾连肿瘤完

整切除(此过程中肿瘤没有穿破)、腹膜后淋巴结清扫，探查发现下腔静脉内可触及坚硬的肿瘤栓子，由左肾血管平面下 3 cm 开始，向上延伸至右心房内，长约 16 cm。建立体外循环，经右心房行心内及下腔静脉瘤栓摘除术，术中见瘤栓完全堵塞下腔静脉口，且与血管内壁紧密粘连，于瘤栓起始处阻断并切开下腔静脉，用子宫探条由下向上反复捅、松解瘤栓，并从心房下腔静脉口取栓至瘤栓取净。术中出血共 600 ml，深低温停循环时间 20 分钟。术后送检肿瘤组织重 2808 克，大小 24 cm * 17 cm * 17 cm，病理检查提示肾母细胞瘤(如图 3 及图 4)。镜下见肿瘤大部分坏死，仅见小灶散在的幼稚胚基细胞，符合肾母细胞瘤化疗后改变。术后患儿曾出现低血压、一过性心酶升高、发热，予血管活性药、强心、利尿、抗感染等治疗后生命体征平稳，术后第 2 天起予 VP16 + IFO 化疗，化疗后当天患儿出现肉眼血尿，考虑出血性膀胱炎，予停用 IFO、水化碱化尿液、更改化疗方案为 VP16 + 卡铂，患儿肉眼血尿缓解。术后第 13 天顺利出院。

3. 结果

术后至今化疗 7 个疗程，放疗 12 次，随访 8 个月，患儿一般情况良好，复查 CT 未见肿瘤复发。

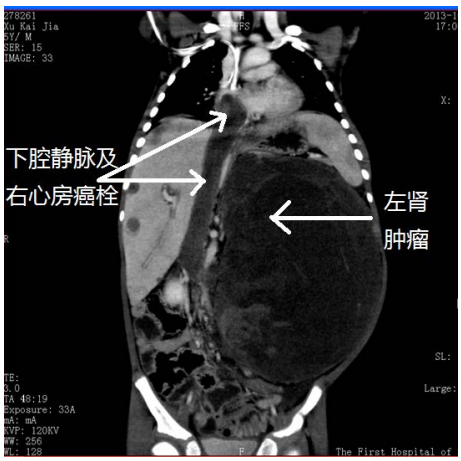

Figure 1. CT: Huge left kidney tumor with inferior vena cava and right atrium tumor embolus
图 1. CT：左肾巨大肿物合并下腔静脉及右心房瘤栓

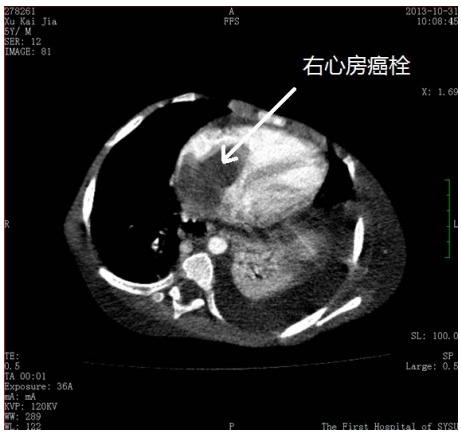

Figure 2. CT: Right atrial tumor embolus
图 2. CT：右心房瘤栓

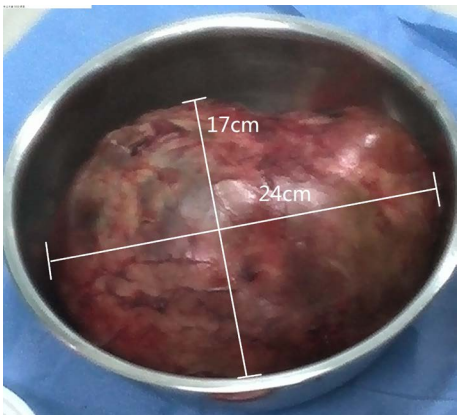

Figure 3. CT: Left kidney and left kidney mass removed during operation (24 cm * 17 cm * 17 cm, weight: 2808 g)
图 3. CT：术中切除的左肾及左肾肿物(24 cm * 17 cm * 17 cm，重 2808 g)

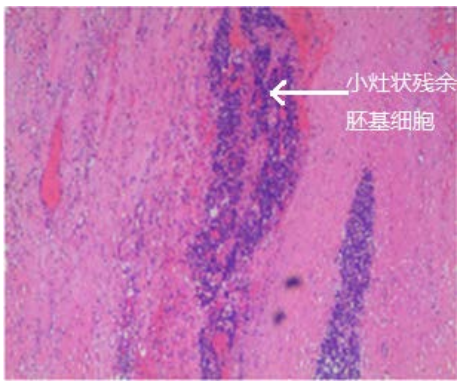

Figure 4. Postoperative pathology of abdominal tumor. Microscopically, most of the tumors were necrotic, and only small scattered immature embryonic cells were seen
图 4. 腹部肿瘤术后病理镜下见肿瘤大部分坏死，仅见小灶散在的幼稚胚基细胞

4. 讨论

随着综合治疗的发展，肾母细胞瘤的预后得到了极大的改善，其整体治愈率高达 90%以上[2]，是肿瘤治疗取得巨大成功的实践之一，具有较高的治疗价值。肾母细胞瘤具有沿血管内生长的倾向[3] [4]，其瘤栓可沿肾静脉长入下腔静脉，甚至延伸至右心房，这虽然并不影响疾病预后[5]，但可能需要体外循环，甚至深低温停循环下进行手术以去除瘤栓[6] [7]，大大增加手术难度和风险，其治疗仍然极具挑战性，需要小儿外科、小儿心脏外科、儿童肿瘤科和儿童重症监护室等多学科的密切合作。

目前有多种儿童肾母细胞瘤合并静脉瘤栓的分级方法，近年来应用较广泛的是美国 Mayo Clinic 五级分类法[8]：0 级：瘤栓局限在肾静脉内；I 级：瘤栓位于下腔静脉内，瘤栓顶端距肾静脉开口处 ≤2 cm；II 级：瘤栓位于肝静脉水平以下的下腔静脉内，瘤栓顶端距肾静脉开口处 >2 cm；III 级：瘤栓在肝内下腔静脉，膈肌以下；IV 级：瘤栓位于膈肌以上下腔静脉内。合并 III 级以上静脉瘤栓的病例，由于瘤栓较长、手术取栓出血量大，且存在瘤栓破裂、血行播散，引起肺转移、甚至肺栓塞的风险。Chiappini 等[8]研究显示，在体外循环、甚至深低温停循环下手术取栓出血较少、术后短期及长期预后良好，可能为

此类患者的最佳选择。本例患儿因合并 IV 级瘤栓，术前已存在心功能不全，在体外循环 + 深低温停循环下顺利取栓后心功能逐渐恢复，提示心功能不全的患儿亦可尝试体外循环下手术，但这是否能提高手术耐受性和成功率，尚需多种心、大样本的研究进一步证实。

肾母细胞瘤首选手术治疗，对于是否应行术前化疗，目前尚无定论[9]。但针对合并下腔静脉瘤栓的病例，由于术前化疗可缩小瘤栓范围，部分病例因此无需体外循环下手术[10]，大大降低手术风险及难度，故推荐对此类患儿常规进行术前化疗。本例患儿有术前化疗指征，但化疗过程中出现肿瘤播散，临床少见。SIOP (the International Society of Pediatric Oncology)研究认为[11]，化疗后肿瘤增大的病例仅占肾母细胞瘤患儿的 5%，且其无瘤生存率和总生存率较化疗有效者明显降低。Cristofani 等[12]报道则提示，即使化疗后肿瘤播散，在患儿可耐受的前提下仍可尝试胸腹联合手术治疗，其五年无瘤生存率高达 100%。

本例患儿经充分术前评估后，在多学科合作下成功施行体外循环下胸腹联合手术，术后顺利撤机，无严重并发症，心功能恢复良好，术后病理示肿瘤细胞大量坏死，无法行组织学分期，但此类患儿往往提示预后较好[13]，继续辅以放化疗，随访至今依然存活，提示在合并心房瘤栓的肾母细胞瘤患儿，若化疗效果不理想，亦可尝试手术切除、减轻肿瘤负荷后，再予放化疗等辅助治疗。

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