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# 肾外Xp11.2易位/*TFE3*基因融合相关性肾细胞癌1例并文献复习

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## 摘　要

**目的：提高对Xp11.2易位/*TFE3*基因融合相关性肾细胞癌(Xp11.2 RCC)的认识，降低误诊率。方法：回顾性分析我院诊治的1例Xp11.2 RCC患者的临床资料，并查阅相关文献进行复习。结果：患者为19岁女性，以间歇性左下腹痛为首发症状。下腹部增强CT示左侧腹膜后占位性病变，诊断为腹膜后肿瘤，行达芬奇机器人辅助下腹膜后肿瘤切除术，术后病理报告为Xp11.2 RCC。结论：本病临床罕见，影像学上可能不表现为明显的肾脏占位，术前难以明确诊断，唯一可能获得良好预后的方案是手术彻底切除肿瘤。**

## 关键词

**肾细胞癌，Xp11.2易位，*TFE3*基因**

# One Case of Extrarenal Renal Carcinomas Associated with Xp11.2 Translocations/*TFE3* Gene Fusions and Literature Review

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## Abstract

**Objective: To improve the understanding of renal carcinomas associated with Xp11.2 translocations/*TFE3* gene fusions (Xp11.2 RCC) and decrease misdiagnosis rate. Methods: A retrospective analysis was performed for the clinical data of a patient with Xp11.2 RCC who was diagnosed and treated in our hospital, and literature review was also performed. Results: The patient was a 19-year-old woman with an initial symptom of intermittent pain of left lower abdomen. Lower abdomen contrast-enhanced computed tomography showed a left retroperitoneal space-occupying lesion, diagnosed as a retroperitoneal tumor; with the help of Da Vinci robot retroperitoneal tumor resection was performed; postoperative pathological examination suggested Xp11.2 RCC. Conclusion: This disease is rare in clinic, and it may not be manifested as an obvious kidney occupation in imaging. It is difficult to confirm the diagnosis before surgery. The only option with a good prognosis is complete surgical removal of the tumor.**

## Keywords

**Renal Cell Carcinoma, Xp11.2 Translocation, *TFE3* Gene**



## 1. 前言

Xp11.2 易位/*TFE3* 基因融合相关性肾细胞癌(Renal carcinomas associated with Xp11.2 translocations/*TFE3* gene fusions, Xp11.2 RCC)是一种临床罕见的肾脏恶性肿瘤，WHO 于 2004 年将其归类为肾癌独立亚型，该亚型以 Xp11.2 位点上 *TFE3* 基因发生断裂并与 ASPL、PSF 等基因平衡易位形成新的融合基因为特征[1]。本病类型特殊，主要发生于儿童及青少年，在成人中发病率低[2]。现报道我院于 2018 年 8 月收治的 1 例临床表现为腹痛、影像学发现腹膜后实性占位性病变的患者，病理检查确诊为 Xp11.2 RCC (该病例报道已获得患者知情同意)。复习相关文献，提高对 Xp11.2 RCC 的认识，降低误诊率。

## 2. 临床资料

患者女，19 岁，未婚。因间歇性左下腹痛 4 天入院。患者 4 天前无明显诱因出现间歇性左下腹痛，伴发热，体温达 38.6℃，无寒战，无腹胀，无尿频、尿急、尿痛，无肉眼血尿。既往心肌炎病史 10 余年。腹膜后彩色多普勒超声示：脐略偏上腹主动脉左侧探及 5.4 × 4.7 × 4.5 cm 低回声团块，形态尚规则，边界清，内回声欠均匀，见无回声区，CDFI：内可探及较丰富条状血流信号，与腹主动脉分界尚清，外缘紧邻左肾，二者分界尚清。考虑：腹膜后肿物。下腹部增强 CT 示左侧腹膜后脊柱旁见团块状软组织密度影，密度不均，最大截面约 44.8 mm × 46.2 mm，呈明显不均匀强化，病变内部见多发迂曲小血管影。双侧肾脏大小形态可，肾实质内未见明显异常密度影。诊断：左侧腹膜后肿物，神经鞘瘤？副神经节瘤？Castleman 病？肾动态显像：双肾滤过功能正常。诊断为腹膜后肿瘤。积极术前准备，行达芬奇机器人辅助下腹膜后肿瘤切除术。术中见：肿块位于左肾前方，呈实性，质硬，直径约 6 cm，沿肿瘤边缘剥离，保护左侧输尿管及左侧肾脏，完整切除肿瘤。术后病理检查示：灰黄结节一枚，大小 8 × 5 × 3.5 cm，表面包膜较完整，切面灰白灰黄，含多个囊腔，切面质韧，部分区域质软，呈灰红色，似坏死。部分肿瘤

细胞透明，呈乳头状、腺泡状及巢状结构，部分肿瘤细胞嗜酸性，可见砂砾体，结合形态学及免疫组化结果，考虑为 Xp11.2 RCC。免疫组化：CD10(+)，CK7(−)，CA IX 部分(+)，*TFE3* 弥漫强(+)，E−Cadherin(+)，Pax−8(+)，Ki−67(+)约 15%，CD117(−)，PLAP(−)，SALL4(−)，CD30(−)，OCT4(−)，GPC3(−)，AFP(−)。基因检测结果：肿瘤体细胞变异检测示肿瘤相关基因变异 2 个(AR 和 TAF1)，意义不明变异 2 个(KDM5C 和 MED12)，变异形式均为基因扩增；胚系变异检测未发现明确有害的可遗传胚系变异；肿瘤突变负荷(TMB)为 0 Muts/Mb (百分位：<80%)；微卫星不稳定性(MSI)检测示微卫星稳定(MSS)。基因检测证实了存在 Xp11.2 易位。

## 3. 讨论

Xpl1.2RCC 临床罕见，JONG 等于 1986 年首次对本病进行了报道[3]，其发病率低，对本病的相关研究往往出自病理学家、遗传学家[4] [5] [6] [7]。该肾癌亚型以独特的 Xp11.2 易位产生新的融合基因为特点，融合后的 *TFE3* 基因表达量升高是因为新形成的融合基因启动子的转录活性更高，使得 *TFE3* 蛋白高表达，细胞对转录调节的干扰作用受其影响，促进肿瘤形成[8]。目前较为常见的是以下五个伴侣基因，包括 ASPL-*TFE3*：t(X; 17) (p11.2; q25)，PSF-*TFE3*：t(X; 1) (p11.2; p34)，PRCC-*TFE3*：t(X; 1) (p11.2; q21)，CLTC-*TFE3*：t(X; 17) (p11.2; q23)和 NonO-*TFE3*：inv(X) (p11; q12) [9] [10] [11] [12] [13]。另外还有 FUBP1-*TFE3*、MATR3-*TFE3* 等数十种不同的融合基因[14]。这些易位位于 X 染色体上，说明本病的临床特征可能存在性别差异[15]。多数研究认为女性发病率高于男性[15] [16] [17] [18]，亦有报告显示男性发病率更高或无性别差异[19] [20] [21] [22]。由于本病发病率低，其临床特征的性别差异仍存在争议[16] [17] [19] [23]。本病好发于儿童，占儿童肾癌的 20%~40%，在成人中发病率低，占成人肾癌的 1%~1.6%，且中年以上患者少有报道[2] [22]。

本病常见的临床表现为血尿，腰痛，腹部肿块，少有患者同时具备典型的肾癌三联征，约 1/3 的患者无症状。肾外症状同样不多见，更多的患者是在体检中行腹部影像学检查时偶然发现。此外，尚有部分患者首先表现为转移灶症状[24]。本病单纯根据影像学表现明确诊断十分困难，在超声及 CT 检查时主要表现为肾脏占位，但本例患者在影像学上表现为非肾脏来源。该肿瘤大多起源于肾脏髓质，边界清楚，为乏血管肿瘤，常伴有坏死、钙化和囊变[17]，增强 CT 表现为轻中度强化、延迟强化[25]，在平扫 CT 上密度不均。平扫 MR 上与正常肾皮质相比，T2WI 呈稍低信号，T1WI 呈等信号或稍低信号，DWI 呈稍高信号，ADC 值明显低于正常肾皮质，MR 动态增强扫描肿瘤各期信号强度变化(皮质期、髓质期和延迟期)低于正常肾皮质[26]。该肿瘤影像学强化方式的特异性可用于鉴别其他肾癌亚型[27]，综合临床及影像学表现，有助于术前正确诊断。若发现肾脏占位性病变且年龄小于 45 岁的患者，伴有肉眼血尿，影像学表现与本病相符，术前应考虑到 Xp11.2 RCC 的可能。

Xp11.2 RCC 大体形态与普通肾细胞癌无明显差异，界限清，常见假包膜，剖面多为灰白色或棕黄色，可见坏死、钙化、囊变或出血[1]。镜下肿瘤组织根据融合基因的不同呈现不同的形态，常见以下两种形态：一种以立方或柱状的癌细胞排列成腺管状、乳头状、巢状为特征，胞界清楚，有大量的透明胞质、嗜酸性胞质，核大、核仁明显，常含有沙砾体，多见于 APSL-*TFE3* 融合性肾癌；另一种形态结构更加紧密，多见实性巢状结构，胞质含量少，核仁不明显，沙砾体少见，多见于 PRCC-*TFE3* 融合性肾癌[28] [29]。由于基因易位导致 *TFE3* 蛋白高表达，免疫组化被广泛应用于本病的诊断，Argani 等研究表明 *TFE3* 蛋白可作为 Xp11.2 RCC 诊断性的免疫标记物[30]。但 Wang 等提出仅仅依靠免疫组化诊断本病特异性低，该亚型的诊断不能只通过免疫组化，还应通过荧光原位杂交技术和其他严格的分子生物学标准明确诊断[31]。

由于 Xp11.2 RCC 发病率低，误诊率较高，目前尚无指南性的治疗方案[1]。唯一可能获得良好预后

的方案是手术彻底切除肿瘤，根治性肾切除术是主要的治疗方法，部分患者行肾肿瘤剜除术后获得良好疗效[32]。保留肾单位的手术方式在处理成人肾脏肿瘤方面日趋成熟，也有研究表明该手术方式在儿童肾癌中与根治性肾切除术相比并发症发生率无统计学差异[33]。Xp11.2 RCC 中有一半以上具有假包膜，对于拥有完整假包膜的小型 Xp11.2 异位性肾癌患者，可以考虑肾部分切除术或肾肿瘤剜除术，保存肾单位，降低长期肾功能不全或终末期肾病的发生率，并提高长期存活率，应谨慎把握保留肾单位手术的适应症，因为其主要缺点是术后局灶性复发，盲目追求肾单位保留可能会导致肿瘤组织特别是难以用肉眼观察到的组织残留，从而增加了复发的风险[34]。当肿瘤直径小于 4~7 cm 且肿瘤位于肾脏一极或外生性生长，可以考虑该手术方式[34] [35]。随着机器人手术系统的不断完善，机器人辅助下微创手术开展日益广泛，考虑本例患者为青年未婚女性，达芬奇系统辅助下微创手术在完整切除肿瘤的前提下，有利于保障患者日后的生活质量。与其他肾细胞癌一样，Xp11.2 易位 RCC 对放化疗不敏感[34] [36]，且细胞毒性化学疗法可能促进肾脏易位癌的发展[37]。而分子靶向治疗对本病具有一定疗效，三类具有抗血管生成活性的药物可供选择：血管生长内皮因子抑制剂、酪氨酸激酶抑制剂、mTOR 受体抑制剂[31]。索拉非尼、舒尼替尼等药物对延长患者生存期部分有效[38] [39]。

Xp11.2 RCC 具有高度的侵袭性，在成年患者中病程进展迅速，与其他 RCC 亚型相比预后较差。在明确诊断时，约有 1/3 的患者已出现转移[40]，多为年龄较大的男性患者[23]。Xp11.2 RCC 是儿童 RCC 的主要病理类型，但在儿童中具有生物学惰性，因此预后比成人更好[34]。儿童和成人之间可能存在临床和病理上的异质性[34]。较高的中性粒细胞－淋巴细胞比值(NLR)，C 反应蛋白/白蛋白比值(CRP/Alb)和血小板－淋巴细胞比值(PLR)与 Xp11.2 RCC 的不良预后相关[41]。Klaassen 等建议患者定期行体格检查和胸腹部 CT，积极随访[42]。

## 4. 结论

本例患者没有家族史，入院后影像学检查提示肿瘤与左肾分界尚清，左肾肾实质内未见异常密度影，且双肾功能无明显异常，术前难以明确肿瘤来源，术后通过病理检查、基因检测最终明确诊断。目前已有的报道中术前均可发现肿瘤与肾脏关系明显，异位肾脏组织来源的肿瘤未见报道，本例研究说明 Xp11.2 RCC 在影像学上可能不表现为肾脏占位性病变，可对诊断以及手术方案的制定造成困难。该肿瘤预后差，本例患者的随访仍在进行。

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