

对文献复习及本病例诊断过程,体会到SPCH做为一种独立疾病,在诊断方面影像与病理结合必不可少,病理科医生能通过组织病理学及免疫组化诊断为肺毛细血管瘤,但镜下组织有限,需结合CT检查才能确定肺内病灶为孤立的而非弥漫性病变,只有影像与病理相结合才易于鉴别SPCH与PCH。

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· 短篇报道 ·

盆腔罕见成脂型孤立性纤维性肿瘤一例

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【关键词】盆腔; 磁共振成像; 孤立性纤维瘤

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A Case of Rare Lipogenic Solitary Fibrous Tumor of the Pelvis

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Keywords: Pelvis; Magnetic Resonance Imaging; Solitary Fibrous Tumor

患者, 女, 78岁, 尿频两月余, 8天前于当地卫生院进行彩超检查, 提示: 盆腔内囊性包块。后在我院进行盆腔MRI检查提示: 盆腔偏右侧多房囊性占位, 体积较大, 分隔及实性成分较多, 内可见出血, 考虑右侧卵巢交界性粘液性囊腺瘤或囊腺癌可能。实验室检查: 癌胚抗原、CA199及CA125均未见明显异常。完善相关检查后全麻下行“经腹腹膜后肿瘤切除术+全子宫+双附件切除术+右侧输尿管部分切除术+右侧输尿管再植术+右侧输尿管支架植入术”, 术后病理结果提示盆腔梭形细胞肿瘤, 有异型, 可见核分裂象, 局部伴出血, 倾向间叶源性肿瘤, 建议免疫组化; 右侧输尿管局部镜检外膜可见肿瘤组织, 断端切缘未见明显异常, 后经上海阿克曼医学检验所免疫组化提示: 符合脂肪瘤样(“成脂型”)孤立性纤维性肿瘤。免疫组化结果:ER(-), PR(-), Desmin(部分+), SMA(-), Caldesmon(-), CD(-), P53(野生型表达), P16(斑片状), CD34(血管+), FH(+), S-100(-), Ki-67(+, 约5%), Calretinin(-), Inhibin a(-), CD117(-), DOG1(-),

D2-40(-), CK5/6(-), STAT6(核+), H3K27Me3(+), Bcl-2(+), CD99(+).

讨论: 孤立性纤维瘤(solitary fibrous tumor, SFT)起源于间叶组织^[1], 在临床上较为少见, 好发于中、老年人, 性别上无明显差异^[2], 胸膜是SFT最常见的好发部位^[3-4], 但发生在腹盆腔较为少见, 成脂型孤立性纤维瘤更为罕见。SFT可以含成熟的脂肪细胞, 以前被称为脂肪瘤样孤立性纤维瘤, 但现在被认为是SFT的脂肪形成性变异^[5]。

SFT多为类圆形或分叶状, 边界清晰, 在显微镜下可见大量梭形细胞疏密交替排列, 其内可见散在分布的纤维基质及“鹿角样”薄壁间质血管^[5]。免疫组化(immunohistochemical, IHC)结果肿瘤细胞对广谱角蛋白、CKAE1/AE3、CK5/6、34 beta E12、MNFI16、CAM5.2、CK、CK19、EMA和p63均为阴性, 基本排除了低级别化生性癌。肿瘤细胞对波形蛋白、CD34、STAT-6呈强烈和弥漫性免疫阳性, 这种IHC特征有利于诊断成脂型孤立性

纤维瘤^[6]。
常规影像学检查对其诊断虽然不具有特异性，但仍是目前临床上主要的诊断技术^[7]。CT平扫病灶密度不均匀，以软组织密度为主，囊变坏死区呈低密度。SFT病灶MRI信号上的差异提示了瘤体所含组织成分不同，黏液样变性区在T₂WI序列上表现为高信号，胶原纤维组织成分在T₂WI序列上表现为低信号^[8]，因此病灶延迟强化；由于肿瘤内部血供较为丰富，动脉期病灶明显强化，瘤周可见

蛇纹石血管^[9]。T₂WI的低信号特征对SFT有一定的诊断价值。
成脂型孤立性纤维瘤影像学诊断困难，发生于盆腔时需要与交界性卵巢粘液性囊腺瘤/癌、神经鞘瘤、脂肪肉瘤或其他间叶组织源性肿瘤进行鉴别，虽然影像学是目前临床常用诊断手段，但其诊断金标准仍是穿刺或术后病理免疫组化^[10]。目前临床上SFT常见的治疗方案是手术根治性切除，术后预后良好。

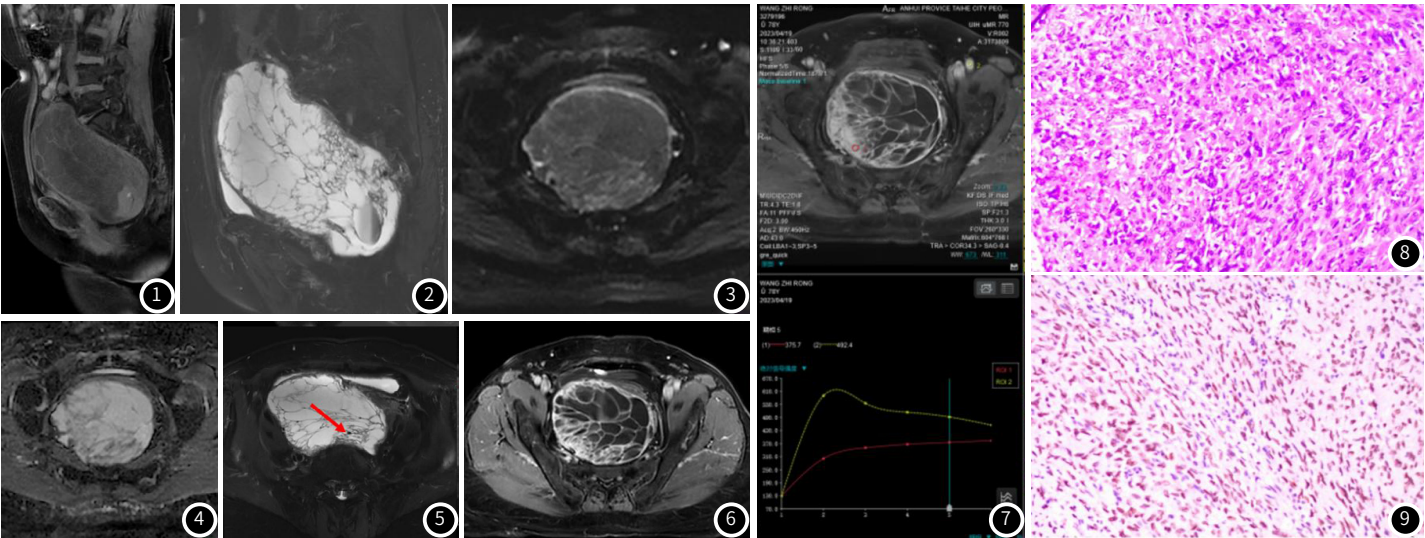


图1 T₁WI示盆腔内见团块状高、低混杂信号；
图2 T₂WI示病灶以囊性为主，多发分隔，并可见液-液平面（考虑合并出血）；
图3 DWI序列病灶实性成分及分隔呈高信号；
图4 病灶相应部位ADC图呈低信号；
图5 T₂WI轴位像病灶内及边缘可见多发流空血管影（红箭头所示）；
图6 增强扫描病灶实性成分及囊壁可见明显强化，囊液无明显强化；
图7 选取实性成分作为感兴趣区域，时间-信号曲线呈缓慢流入型；
图8 HE染色，镜下可见大量梭形细胞交替排列；图9 免疫组化染色可见STAT6（核+）。

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