

胸膜外孤立性纤维性肿瘤的临床病理特征分析

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[摘要] **目的** 探讨胸膜外孤立性纤维性肿瘤的临床病理特征及鉴别诊断。**方法** 对26例胸膜外孤立性纤维性肿瘤样本进行HE及免疫组织化学染色,并结合临床及影像学资料进行综合分析。**结果** 孤立性纤维性肿瘤的组织学形态多样,典型者可见卵圆形或梭形细胞不规则排列,间质中可见粗大的胶原带、鹿角状血管及管周透明变性;免疫组化染色STAT-6、CD34、Bcl-2、CD99、SMA阳性率分别为96.2%、92.3%、92.3%、76.9%、26.9%。**结论** SFT常见发生于胸膜,亦可发生于其它部位,其组织学特征和免疫组化表型有助于明确诊断,但当其发生于不常见的解剖部位时,应当对重点需要鉴别或排除的疾病加以认真细致的鉴别并综合判断其临床预后。

[关键词] 胸膜外孤立性纤维性肿瘤;组织学形态;生物学行为;临床预后

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Clinical and Pathological Characteristics of Extrapleural Solitary Fibrous Tumor

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[Abstract] **Objective** To investigate the clinical and pathological characteristics of the extrapleural solitary fibrous tumor and the relevant differential diagnosis. **Methods** HE and immunohistochemical staining were performed on 26 samples of isolated fibrous tumors outside the pleura, and a comprehensive analysis was conducted based on the clinical and imaging data. **Results** The histological morphology of isolated fibrous tumors was diverse, with typical cases showing the irregular arrangement of oval or spindle shaped cells. In the stroma, thick collagen bands, antler like blood vessels, and peritubular transparent degeneration could be seen. The immunohistochemical staining demonstrated that the positive rates of STAT-6, CD34, Bcl-2, CD99, SMA were 96.2%, 92.3%, 92.3%, 76.9% and 26.9%, respectively. **Conclusion** SFT commonly occurs in the pleura and can also occur in other areas. Its histological characteristics and immunohistochemical phenotype help to clarify the diagnosis. However, when it occurs in uncommon anatomical sites, it is necessary to carefully and meticulously distinguish and comprehensively judge the clinical prognosis of key diseases that need to be distinguished or excluded.

[Key words] Extrapleural solitary fibrous tumor; Histological morphology; Biological behaviour; Clinical prognosis

孤立性纤维性肿瘤(solitary fibrous tumor, SFT) 1(中间性)和8815/3(恶性孤立性纤维性肿瘤)。
是一种纤维母细胞性肿瘤, ICD-O 编码分别为8815 SFT主要发生于胸膜,称为胸膜孤立性纤维性肿

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瘤(solitary fibrous tumor of the pleura, SFTP), 少数可发生于其他解剖部位, 称为胸膜外孤立性纤维性肿瘤(extrapleural solitary fibrous tumor, ESFT)。其组织学形态千变万化, 且难以通过组织异型性或细胞异型性推测肿瘤的生物行为。当SFT发生于不常见的部位时, 明确性质、鉴别诊断以及预后的判断就显得比较困难。为了更好地认识胸膜外SFT的特征, 收集了26例该类病例并对其临床表现、组织学特点、免疫表型及临床预后进行了综合分析并展开讨论。

1 资料与方法

收集昆明医科大学第三附属医院病理科2000年1月至2021年8月的手术标本中诊断为SFT且发生于胸膜以外解剖部位的病例, 其中不包括曾经诊断过原发于胸膜的SFT而后发生转移的病例。共收集26例, 初诊年龄22~83岁, 平均47.9岁; 其中男性15例, 女性11例(男女比例15:11), 见表1。

表1 病例基本情况
Tab. 1 Basic information of patients

性别	n	年龄(岁)	肿瘤大小(cm)
男	15	>55(n=6)	>5(n=14)
女	11	≤55(n=20)	≤5(n=12)

以上病例的病理切片由3位诊断经验丰富(主治医师及以上职称)的病理医师按照第五版(2020年)WHO软组织肿瘤分类标准^[1]独立阅片, 总结并描述上述病例在光学显微镜下的组织学形态特征和免疫组化表型。

1.1 HE染色

按照常规组织学制片方法固定、包埋、切片并进行苏木精-伊红染色。

1.2 免疫组织化学技术

采用EnVision二步法, 二氨基联苯胺(diaminobenzidine, DAB)显色, 苏木素复染; 所用抗体及试剂盒购自福州迈新生物技术开发有限公司。

2 结果

2.1 病理特征

标本大体情况: 肿瘤分别位于骶骨、腹股沟、腹膜后、大脑额叶、肩部、盆腔、胃壁、颈椎、膀胱、皮肤、心包、宫颈、股骨、面部皮下、鼻

前庭、阔韧带、项部、乳腺、腠窝、会阴、肾上腺、睾丸、前列腺等部位。肿瘤最大径0.7~14.9 cm, 平均4.5 cm; 肿瘤边界尚清; 切面灰白灰红, 实性, 质软到质中, 灶区呈多结节状; 多有包膜。

组织学形态: 肿瘤细胞形态为卵圆形或梭形, 无序分布于丰富的、玻璃样变的胶原间质中; 细胞核较小、深染, 核染色质较为细腻、均匀, 核仁不易见, 核分裂象少见; 胞浆稀少, 呈双嗜性, 细胞边界不清; 可见细胞密集区和稀疏区交替分布; 其中, 13例肿瘤间质中可见“鹿角状”的薄壁血管; 6例可见血管周透明变性; 1例肿瘤细胞丰富, 细胞异型性明显, 可见小灶性坏死; 此外, 部分病例可见间质黏液变性(6/26)、微囊性变(3/26)、小出血灶(3/26)、成熟的脂肪细胞(2/26)和淋巴细胞浸润(2/26)等形态学特征, 见图1、表2。

免疫组化表型: 26例Vimentin(+), 25例STAT-6(+), 24例CD34(+), 24例Bcl-2(+), 20例CD99(+), EMA 5例阳性、8例为小灶弱阳, 7例SMA呈现灶阳性或弱阳性, S-100 3例为阳性、3例为散在阳性; 1例Desmin为灶状阳性。综上, Vimentin阳性率为100%(26/26), STAT-6、CD34、Bcl-2、CD99、SMA阳性率分别为96.2%(25/26)、92.3%(24/26)、92.3%(24/26)、76.9%(20/26)、26.9%(7/26), Desmin、S-100、CD117及EMA则为3.8%(1/26)、23.1%(6/26)、0.0%(0/26)及50.0%(13/26), 见图2、表2。

2.2 影像学特征

CT平扫、增强和三维重建, 多表现为单发、孤立性, 类圆形或椭圆形软组织密度肿块影; 常具有明显包膜, 边界清楚, 有浅分叶; 病灶密度欠均匀, 其内可见不规则低密度区。增强扫描呈均匀“地图样”强化或显示絮状强化影, 部分区域呈轻-中等强化; 血管成像示病灶期血管丰富并可见多发畸形血管团; 三维重建显示肿瘤推挤周围结构而非浸润性生长。

2.3 随访资料

26例均经根治性手术切除肿瘤, 术后随访病例25例(1例失访), 其中23例无瘤生存, 1例因恶性心包积液维持治疗, 1例出现远处转移(肝转移)。

3 讨论

SFT是一种相对少见的梭形细胞肿瘤, 其组

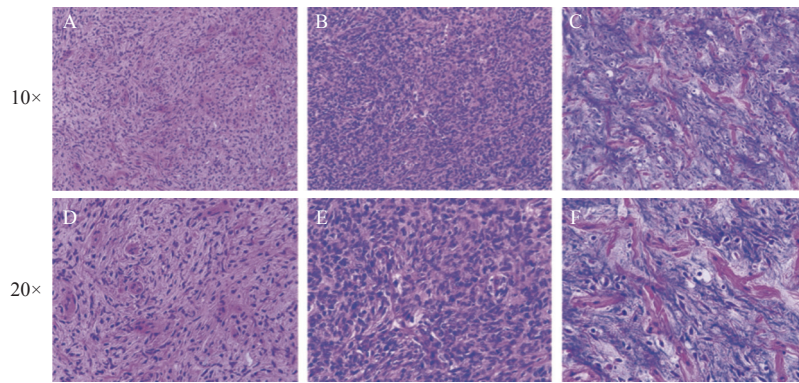


图 1 胸膜外孤立性纤维性肿瘤的 HE 染色(10×/20×)

Fig. 1 HE staining of ESFT (10×/20×)

A: 疏松排列的梭形肿瘤细胞; B: 密集排列的卵圆形肿瘤细胞; C: “鹿角”状血管及间质黏液变性; D-F 分别为 A-C 在 20 倍物镜下的形态学图片。

表 2 胸膜外孤立性纤维性肿瘤的病理学特征

Tab. 2 Pathological characteristics of ESFT

病理特征	n	占比(%)
组织学形态		
“鹿角状”薄壁血管	13	50.0
血管周透明变性	6	23.1
细胞丰富、有异性性	1	3.8
小灶坏死	1	3.8
间质黏液变性	6	23.1
微囊性变	3	11.5
小出血灶	3	11.5
成熟脂肪细胞	2	7.7
淋巴细胞浸润	2	7.7
免疫表型		
STAT-6	25	96.2
CD34	24	92.3
Bcl-2	24	92.3
Vimentin	26	100.0
CD99	20	76.9
SMA	7	26.9
S-100	6	23.1
EMA	13	50.0
Desmin	1	3.8
CD117	0	0

织学起源目前尚无定论。有学者^[2]认为该类疾病起源于未曾分化的原始间叶细胞,且可向成纤维细胞、肌成纤维细胞等分化。SFT好发于胸膜,通常称之为胸膜孤立性纤维性肿瘤(SFTP),也可发生于其它任何解剖部位,包括深部软组织^[2]、

皮肤^[3]、胃肠道^[4]、泌尿生殖系统^[5]、头颈部^[6]、甲状腺^[7]、腮腺^[8]、眼眶^[9]、口腔和涎腺^[10]、喉^[11]、气管^[12]、女性生殖道^[13]、脑脊膜^[14]、肾上腺^[15]、骨盆^[16]、腹膜后、胰腺^[17]、肝^[18]、肾^[15]、肺^[19]和骨^[20]等(其中最常见的发病部位为下肢、腹膜后腔、眼眶^[21]),称之为胸膜外孤立性纤维性肿瘤(ESFT)。

肉眼观察典型的 SFT 是边界清楚、质地较硬的肿块,切面均质、色白,可有微囊腔,有时可有出血。肿瘤大小从 1~20 cm 不等,腹腔内 SFT 可大于 20 cm。组织学上,典型的 SFT 是由形态温和的卵圆形或梭形细胞无序地分布于胶原间质中,间质可能出现不同程度的黏液变性^[22]。间质中几乎均可见具有大分枝的或“鹿角状”的薄壁血管,此外中等大小血管的管周透明变性也是一个常见特征。肿瘤细胞核较小、深染,胞浆稀少、双嗜性,胞界不清。SFT 的变异型是根据其最显著的形态学特征定义的。形成脂肪的 SFT 变异型(脂肪瘤性血管外皮细胞瘤)表现为间质中存在大量成熟的脂肪组织,常位于深部软组织,也可发生于眼眶、颈部、纵隔、心外膜、腹膜后、腹股沟、大腿和腮腺等部位^[23-26]。富于巨细胞的 SFT 变异型(曾被认为是巨细胞血管纤维瘤)其特征为散在的多核巨细胞分布于假血管间隙周围,大多发生于眶周软组织,也可发生于头颈部、背部、腹膜后、臀部、外阴和腹股沟区^[27-29]。恶性 SFT 与 SFT 难以区分,前者切面可不均质,可出现坏死、浸润毗邻组织等。尽管目前尚未达成共识,仍有部分数据提示临床表现具有侵袭性的 ESFT 常有以下病理特征:浸润性生长的边缘,细胞多形性、核异型性,细胞丰富,高增殖活性和坏死等。去分化 SFT 是一种很少见的亚型,它从典型的 SFT 直接转化为高级别肉瘤^[30],组织学上表现

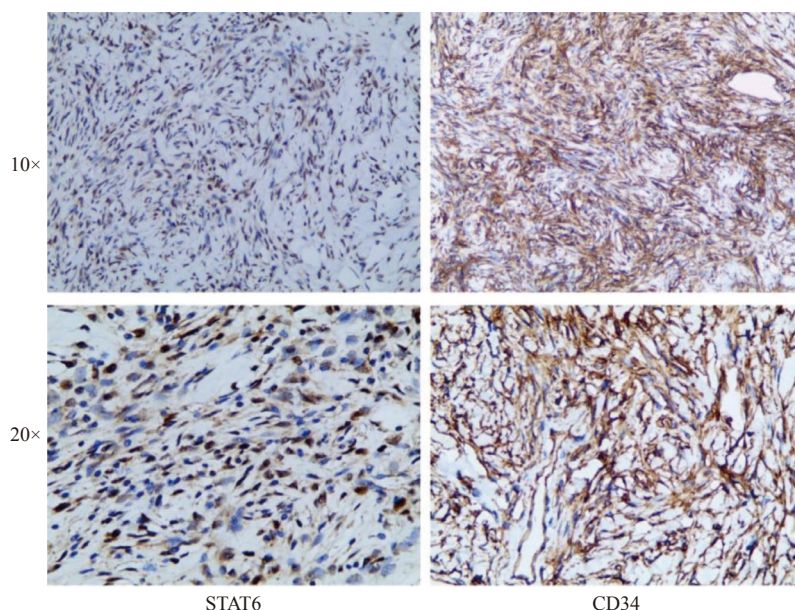


图2 STAT6和CD34的免疫组化染色(10×/20×)

Fig. 2 IHC staining of STAT6 and CD34 (10×/20×)

为梭形细胞肉瘤,而非其它具有特定分化的肉瘤或未分化多形性肉瘤^[31]。

ESFT具有与SFTP相似的形态学特征,如胶原间质中无序分布的椭圆形或梭形的细胞。此外,还可能具有混合细胞成分的、成脂肪的、含有多量巨细胞的形态。从细胞形态、肿瘤边界、细胞极性和侵袭性来说,该类肿瘤有很大的变异性,且有报道ESFT几乎在每个解剖部位都可发生。

免疫表型方面,SFT常弥漫强阳地表达CD34、Bcl-2和CD99,然而,约有5%~10%的典型SFT和绝大部分恶性SFT中CD34表达缺失。众所周知,有几种梭形细胞肿瘤都可表达CD34,使得它在SFT鉴别诊断中的意义有所弱化^[32]。Bcl-2是SFT的1个敏感指标,但特异性不佳^[33]。CD99的敏感性不如CD34和Bcl-2,且几乎不具特异性^[32]。NAB2-STAT6融合基因的发现使笔者得知STAT6在SFT中几乎恒定的核表达,此后STAT6成为了典型和恶性SFT诊断的最敏感(91%)且特异(75%)的指标^[33]。近年还有研究发现SFT中高表达GRIA2基因,继而导致GRIA2蛋白的异常表达^[34]。GRIA2蛋白是1种谷氨酸受体,通常不在中枢神经系统以外出现,但在SFT和隆突性皮肤纤维肉瘤中表达^[35]。已有研究报道80%的SFT、86%的恶性SFT和100%的去分化SFT表达GRIA2蛋白^[36]。在1个鉴别脑膜SFT、脑膜瘤和滑膜肉瘤的基因表达谱研究中,研究者发现ALDH1在SFT中稳定表达。在另1个研究中发现,76%的SFT中存在ALDH1的胞浆表达^[37]。此外,SFT中还

有EMA、SMA、 β -catenin、NSE和GFAP等蛋白的局灶或少许表达,而CD31、Desmin、h-caldesmon和S-100通常都不表达。有的腹部恶性SFT病例可有多灶CK阳性,可能会被误诊为肉瘤样癌或间皮瘤^[38]。STAT6和ALDH1的共表达对于SFT的诊断有很高的敏感性和接近100%的特异性^[39]。当肿瘤位于胃肠道时,必须考虑到与胃肠间质瘤(gastrointestinal stromal tumors, GIST)的鉴别。GIST的组织学形态多种多样,当与SFT相似时,借助CD117和DOG1加以鉴别,因大多数GIST都表达这2个蛋白且呈弥漫强阳性,而SFT通常不表达。

ESFT可发生于5~92岁,50~70岁为高峰发病年龄,儿童或青少年患者少见,性别差异不明显,也有报道称男性患者略多于女性(约3:2)^[40-41]。ESFT的临床表现取决于其解剖部位,通常表现为缓慢生长的肿块且为偶然发现,有时可因压迫邻近器官组织而被发现。此外还有罕见情况是因患者低血糖而被发现,可能是由于肿瘤细胞分泌胰岛素样生长因子(insulin-like growth factor, IGF)引起^[42-43]。腹腔内的SFT往往比肢端、躯干或头颈部的肿瘤体积更大^[44],但其原因取决于腹腔内有更大的生长空间,而非肿瘤具有更强的生长能力。ESFT的总体中位生存期为5~94个月,5a和10a生存率分别为89%、73%^[44]。大范围的外科手术切除是局限性病灶的标准治疗方案,临床上不常规应用放射治疗或辅助化学治疗^[45]。如在切除原发肿瘤时已有转移,同时切除转移瘤可使患者获益^[46]。然而尽管外科手术切缘阴性,局部复

发或远处转移仍然可能发生,因此近年来术后放疗也被部分学者推荐用于改善肿瘤的局部控制效果^[47]。据报道,ESFT的侵袭性生物学行为发生率为6%~23%^[48]。转移常常是血源性的,靶器官可为肺、肝、肾上腺、骨、脑、肌肉和胃肠道等,发生转移的患者预后差,约75%仅有22~46个月的中位生存期^[48]。实际上,ESFT的预后是难以准确判断的,因其并非严格依赖组织学特征,即便是不具非典型组织学特点的肿瘤,也有可能具有侵袭性,具有非典型组织学特征的肿瘤亦可表现为惰性生物学行为^[49]。值得一提的是,初诊时组织学低级别的肿瘤在复发或转移后往往出现恶性组织学特征^[19]。大多数复发发生在术后2 a,但也有报道可长达17 a^[50-51]。因此,术后2 a内需要每半年复查影像学,之后则保证每年复查的随访至关重要。

SFT的发病机制可能与染色体12q13臂内倒位导致的NAB2-STAT6融合基因有关。NAB2-STAT6融合基因几乎存在于所有SFT病例中,因此目前认为这个突变是该类疾病发生和进展的重要原因,并且与原发的解剖部位和形态学亚型无关^[34]。NAB2-STAT6融合基因编码一个嵌合体蛋白,其内NAB2的C端抑制结构域被STAT6的高度可变部分所取代,导致STAT6蛋白C端部分的核表达^[34]。这个基因融合最显著的后果是导致NAB2的功能受干扰,而非STAT6的失调。SFT中早期生长反应基因1(early growth response 1, EGR1)靶基因的失调就是由于NAB2的突变^[34]。NAB2-STAT6融合的断裂点是可变的,因此可以产生多种变异体,其中NAB2_{ex4}-STAT6_{ex2/3}是最常见的融合变体,可见于70%~90%的病例,通常为SFTP且具有经典的组织学形态和良性生物学行为;而第二常见的变体NAB2_{ex6}-STAT6_{ex16/17}往往在ESFT中出现,且更具侵袭性和异型性。然而关于不同融合变异体与预后相关性的研究目前报道较少,因此还需要笔者的长期随访和深入研究。基于大量文献检索及笔者的临床、病理诊断经验,多数ESFT具有与SFTP相似的信号通路,即STAT家族成员被受体相关激酶磷酸化后,可激活炎症反应通路,继而引发多种免疫细胞和组织细胞的激活和增殖。

此外,核磁共振成像在ESFT的诊断中也是至关重要的。肿瘤中混合存在的组织学成分如纤维组织、细胞成分以及高度血管化的区域均可影响其影像学表现。SFT为边界清楚、边缘光滑的肿块,大多数在T1相对于肌肉表现为低信号或

等信号,T2加权像上为局灶或弥漫的低信号(此为肿瘤的纤维成分所致,且为重要的诊断和鉴别诊断要点),而在高度血管化的区域可有强烈的局灶或弥漫性对比增强表现,有的病灶内可有钙化。肿瘤内组织学特征相对恶性的区域倾向于在T2加权像上表现出中-高度的信号强度。此外,SFT的临床和影像学表现都与纤维组织细胞瘤、纤维肉瘤、滑膜肉瘤和转移性硬癌等有相似之处,在临床工作中应加以鉴别。

综上所述,ESFT的诊断比较具有挑战性,由于该类疾病比较少见,尤其在发病部位罕见时,需要整合临床表现、影像学、病理学、基因检测等多方面的资料来综合分析。并且由于ESFT往往比SFTP的侵袭性更强、复发率更高,因此准确诊断ESFT显得尤为重要。在今后的临床工作中,需要更加关注并总结ESFT的诊断原则、鉴别诊断、分子病理学方面的进展和预后相关因素,从而为未来更加精准地诊断和治疗ESFT提供科学严谨的理论依据。

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