

· 短篇论著 ·

胸壁侵袭性纤维瘤病1例*

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【摘要】目的 通过分析1例胸壁侵袭性纤维瘤病(AF)的临床诊治过程,降低误诊及漏诊。方法 结合文献分析1例胸壁AF女性患者的临床资料,术前及术中误诊的原因及解决措施。结果 胸壁AF患者临床表现及影像学无特异性,术前诊断困难;术中切缘不干净易导致残留复发。结论 对于乳腺既往纤维瘤手术史以及影像学提示胸壁肿物侵犯胸肌,术前病理穿刺提示梭形细胞瘤患者术中需根据冰冻结果采取根治性手术,且术后须长期随访。

【关键词】胸壁侵袭性纤维瘤病;复发;手术

【中图分类号】R738.6

【文献标识码】D

【基金项目】深圳市第二人民医院临床科研项目(20193357028)

DOI:10.3969/j.issn.1009-3257.2023.02.005

Aggressive Fibromatosis of Chest Wall: A Case Report*

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Abstract: Objective By analyzing the clinical diagnosis and treatment of a case of aggressive fibromatosis of chest wall (AF), we can reduce misdiagnosis and missed diagnosis. **Methods** Combined with the literature, the clinical data of a female patient with chest wall AF, the causes of preoperative and intraoperative misdiagnosis and solutions were analyzed. **Results** The clinical manifestations and imaging of patients with chest wall AF are nonspecific, and the preoperative diagnosis is difficult; Unclean incisional margin during operation is easy to cause residual recurrence. **Conclusion** For the previous history of breast fibroma surgery and imaging findings that chest wall tumors invade the chest muscle, preoperative pathological puncture suggests that patients with spindle cell tumors need to take radical surgery according to the frozen results during operation, and long-term follow-up after operation.

Keywords: Aggressive Fibromatosis Of Chest Wall; Recrudescence; Operation

1 临床资料

患者,女,27岁。发现左乳肿物4月余,4年前于我院行左乳纤维瘤切除术,入院查体见左乳5-7点连接胸壁可扪及一大小约8x3cm肿物,质中,边界欠佳,无触痛。检验指标无明显异常。B超示左乳5-7点方向低回声区,上近乳晕区,下超出乳房范围,浅层紧邻皮肤层,边界至胸壁肌层,局部肌肉纹理紊乱消失,范围约88x30x17mm(图1)。术前穿刺活检诊断梭形细胞肿瘤(图2)。核磁结果为左乳片状的不规则低回声,左区皮下脂肪层模糊,T₁等信号,T₂高信号,内部强化均匀,TIC曲线平台型,ADC值混杂高信号,DWI信号高(图3)。术中探查见左胸壁肿物与左胸壁筋膜、胸大肌、肋间肌融为一体,用电刀锐性游离,肌纤维为核心完整切除肿物,术后病理:1.左乳病变组织符合纤维瘤病,2.左胸壁病变组织考虑为侵袭性纤维瘤病,免疫组化结果:SMA(+),S100(-),CD34(-),Desmin(+),B-catenin(细胞浆及细胞核+),AE1/AE3(-),P63(-),Ki-67(3%+)(图4-1至4-4)。术后随访至今1年,患者恢复良好,复查核磁未见复发及明显异常。

2 讨论

AF也叫做肌肉腱膜纤维瘤病、韧带样型纤维瘤病。Farlanes医生在1832年首先描述了该疾病,该病发生率在所有软组织肿瘤中占比小于3%,局部复发率在25%~77%^[1]。本病胸壁类型少见,它的发病原因尚不清楚,或许和创伤、遗传或激素等有关^[2]。临床表现为深在、界限不明的质硬无痛肿物。

术前由于胸壁AF影像学及临床表现无特异性,在年轻女性患者中多发,患者多因乳腺肿物就诊于甲乳外科,在术前临床易误

诊为乳腺来源的纤维瘤,侵及胸肌或胸壁;显微镜下,其边界不清且具有浸润性,由致密的胶原基质和长束带、均匀的成纤维细胞组成,细胞密度低^[3],无多形性和坏死,有丝分裂很少见^[4](每10个高倍视野最多5个)。在肿瘤的外围,有可能导致萎缩的横纹肌残留物,因此放射科医生也可能将其误认为是恶性疾病的证据。本病例术前考虑为复发性乳腺纤维瘤,术前穿刺病理考虑梭形细胞瘤,未能明确诊断,有资料显示在AF初始检查中误诊病例约为30~40%^[5,6],穿刺活检在间质细胞来源肿瘤的诊断,存在固有的取材不足问题。

受术前诊断信息不充分,术中手术医生常按乳腺来源的肿瘤标准来切除肿物,导致肿瘤非腺体部分纤维缘切除不完整,术中残余高概率发生,此外,如果肿物侵犯超过乳腺组织且在纤维轴延伸,即使冰冻回报恶性,术者扩大切除范围仍按环形切除,而非延纤维轴扩大,切缘仍可能阳性。在没有认识到AF多来源胸壁肌肉时,乳腺来源累及肌肉或肌肉来源肿瘤累及乳腺,术后肿瘤基底部的肿瘤残余率会有差异。手术切缘阴性的最佳切除应该是手术的主要目标,从而获得最佳的肿瘤学预后^[7-9]。

另一些研究报告^[10-11],手术切缘阳性与阴性患者的5年无进展生存率没有显著差异。尽管文献报告对切缘阳性扩大切除和保守治疗间选择对比,常出现相互矛盾的结果,但我们对于手术切缘阳性可手术患者仍推荐扩大切除。考虑到该患者既往乳腺纤维瘤手术史,此次肿块深层累及胸壁肌层,便采用以肌纤维为核心广泛切除病灶,术后完整病理的免疫组化提示胸壁AF。文献报道^[12-13]AF灶里细胞波纹蛋白染色呈强阳性,B-catenin蛋白与平滑肌肌动蛋白(SMA)也呈不一样的阳性程度(图4A、4C),但通常不表达CD-34和S-100蛋白,本病例病理学表现与文献报道相符。

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此外,女性病人对乳房的美容要求较高,顾及到术后乳房的完整性及美观性,手术医生在切除范围权衡时,侧重点偏向了外形,这一点也是术后复发的重要因素。我们建议采取根治性手术,辅以整形修复技术,以最大的可能争取切缘阴性,避免再次手术及早期复发。

综上所述,胸壁AF目前的临床及影像学表现均无特异性,

术前易误诊,术中易残留,整块切除术后病理诊断仍是“金标准”。目前手术仍是一线的治疗手段,初始切除一定要彻底,再次手术以纤维轴广泛切除,对于复发的乳腺纤维瘤病及性质未明的浸润胸肌的肿物,在日常工作中要求临床、放射科及病理科医师考虑到此病的可能性,避免漏诊或误诊。

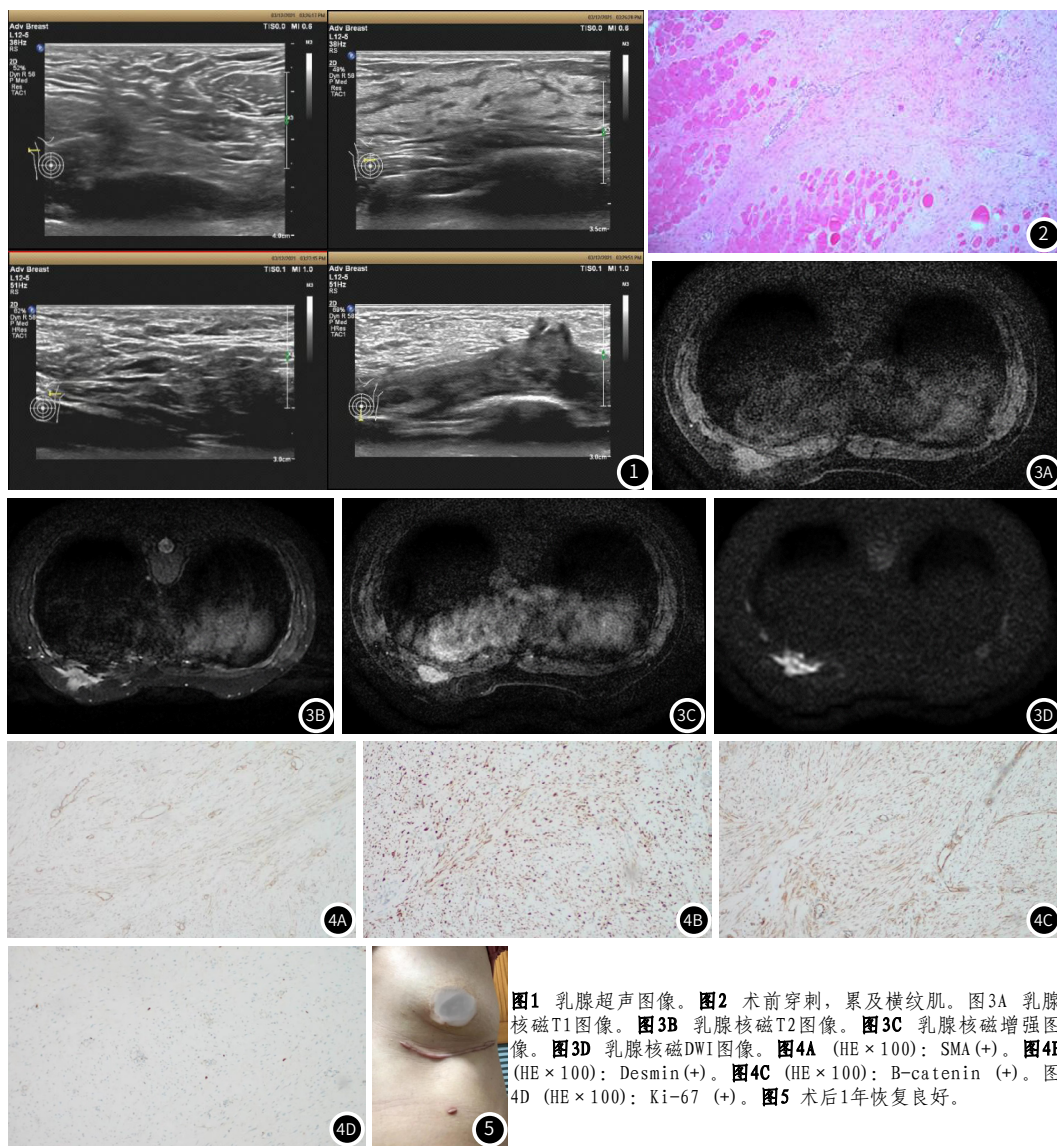


图1 乳腺超声图像。图2 术前穿刺,累及横纹肌。图3A 乳腺核磁T1图像。图3B 乳腺核磁T2图像。图3C 乳腺核磁增强图像。图3D 乳腺核磁DWI图像。图4A (HE×100): SMA(+).图4B (HE×100): Desmin(+).图4C (HE×100): B-catenin (+).图4D (HE×100): Ki-67 (+).图5 术后1年恢复良好。

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(收稿日期: 2022-08-08)

(校对编辑: 姚丽娜)