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CASE REPORT

Decoding a Soft Tissue Enigma: Malignant Solitary Fibrous Tumour of Thigh

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Abstract

Solitary fibrous tumour (SFT) is a rare mesenchymal neoplasm, most commonly arising from the pleura, with infrequent extra-pleural involvement. We report a malignant extra-pleural SFT in a 64-year-old woman presenting with a longstanding thigh mass. Wide local excision demonstrated a spindle cell neoplasm with hemangiopericytoma-like vasculature, necrosis and high mitotic activity. Differential diagnoses included malignant SFT and synovial sarcoma. Immunohistochemistry revealed diffuse CD34 expression and strong nuclear STAT6 positivity, confirming malignant SFT. This case illustrates the diagnostic challenges of spindle cell tumours and emphasizes the importance of STAT6 staining. Owing to malignant potentialof extra-pleural SFTs, long-term surveillance is recommended.

Key Words

Extremities, Immunohisto chemistry, Solitary Fibrous Tumors

Introduction

Solitary fibrous tumour (SFT) is a rare mesenchymal neoplasm originally described in pleura,with recognized extra-pleural occurrences. These tumours arise from primitive fibroblast-like CD34 positive mesenchymal cells and are typically well circumscribed and slow growing.[1] While most SFTs are benign, approximately 10–15% show aggressive features, including recurrence or metastasis, leading to worse prognosis.[2] Tumours in extremities particularly thigh, have a higher risk of malignant transformation.[3] We present a case of an extra-pleural malignant SFT of thigh that clinically and histologically mimicked synovial sarcoma. Immunohistochemistry (IHC) was instrumental in establishing correct diagnosis.

Case Report

A sixty four-year-old female presented to surgical outpatient clinicwith a longstanding swelling on medial aspect of her right thigh, first noticed 15 years ago. Examination revealed a firm, non-tender mass approximately 12 × 10 cm. Magnetic Resonance Imaging (MRI) of thigh showed a well circumscribed soft tissue lesion measuring 11 × 10.5 × 9 cms, isointense on T1 and hyperintense on T2 sequences [Figure 1a]. Computed tomography (CT) of thorax revealed semi-solid and ground-glass pulmonary nodules, suspicious for metastasis.

Wide local excision of mass revealed a well encapsulated lesion with areas of hemorrhage, necrosis and myxoid change [Figure 1b]. Microscopy showed malignant spindle cells arranged in a vague storiform andhemangiopericytoma-like vascular pattern [Figure 2a, 2b]. Few of the tumor cells were seen rosetting around hyalinised blood vessels. Areas of myxoid change, necrosis, hyalinization and fibrosis were present with 8–10 mitoses/10 HPF. A preliminary diagnosis of malignant

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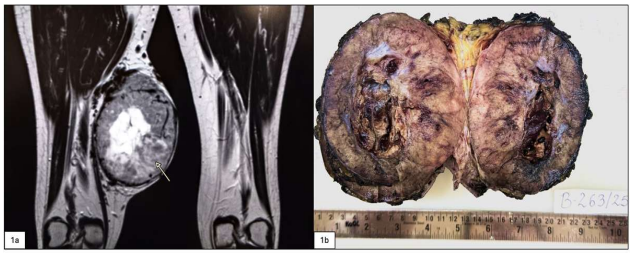

Figure 1a: MRI of thigh showing a Well Circumscribed Soft Tissue Lesion Measuring 11×10.5×9 cms (arrow); 1b: Cut Surface of the Mass Revealing a Well Encapsulated Lesion with Areas of Hemorrhage, Necrosis and Myxoid Change.

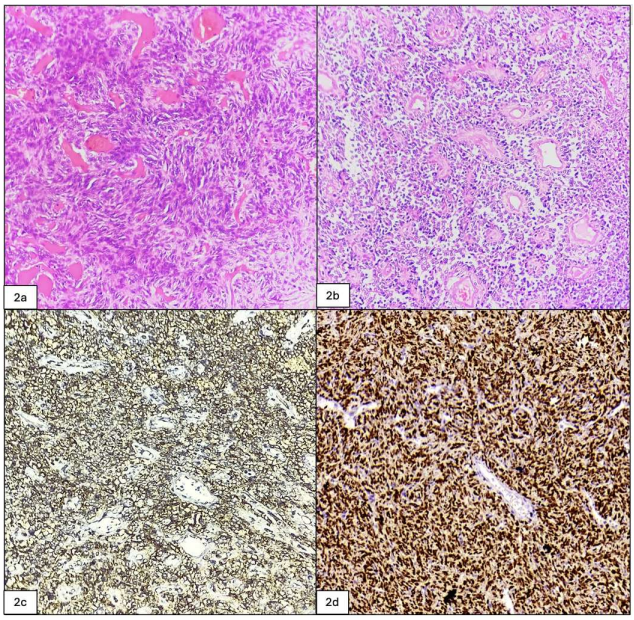

Figure 2a: Microscopy showing Atypical Spindle Cells Arranged in Hemangiopericytoma-like Vascular Pattern (H&E,x400), 2b: Hyalinised Vasculature with Rosetting of Tumor Cells (H&E,x200), 2c: CD34 IHC showing Diffuse Membranous Positivity (x100); 2d: STAT6 IHC showing Strong Nuclear Positivity (X200).

spindle cell tumour was made with differential diagnoses including malignant SFT and synovial sarcoma.

Immunohistochemistry (IHC) revealed negativity for cytokeratin (CK), CD99 and TLE1. The tumour showed diffuse CD34 expression and strong nuclear STAT6 positivity, confirming malignant SFT [Figure 2c, 2d]. The metastatic risk score was given as per three variable model as 5 (high metastasis risk) [Age – 1; Mitosis- 2; Tumor size -2]. Postoperative period was uneventful and the patient was referred to higher center for adjuvant therapy.

Discussion

Solitary fibrous tumors (SFTs) represent a spectrum of mesenchymal tumors encompassing tumorsformerly known as hemangiopericytomas, which are categorized by 2013 WHO classification as having intermediate biological potential, with rare tendency to metastasize.[1] SFT was first described in 1931 by Klemperer and Rabin in a patient with distinctive pleural lesion and were thought to arise exclusively within thoracic cavity.[4] However, SFTs have also been rarely identified at extrapleural locations like extremities, abdominal cavity, pelvis, retroperitoneum, head and neck (sinonasal, orbit) and trunk.[1]

SFT can occur at any age but are most commonly diagnosed in people between 50 and 70 years old with no strong gender predilection. Though some female predominance has been reported, similar to our case.[4] SFTs are generally slow-growing neoplasms with afavourable prognosis, but approximately 10% of SFTs can show aggressive behaviour with local recurrence and distant metastasis observed on long-term follow up.[5] In our case, pulmonary nodules observed on CT scan were suspicious of metastases; however, this was not confirmed histopathologically. The exact origin of malignant SFT is still under debate. Some experts suggest malignant SFT may arise from pre-existing benign tumors, supported by cases showing both types in same patient, while others believe malignant SFT can develop de novo. Diagnosing Malignant SFT is often challenging, as clinical and imaging findings are usually non-specific. Hence, they are frequently missed or misdiagnosed. Also they demonstrate wide histologic variability and can mimic other spindle cell neoplasms. However, recent advancements in diagnostic techniques have significantly enhanced our understanding of this disease.[4]

Wide resection is the treatment of choice for SFTs. Poor prognostic factors include positive surgical margins and tumour size >10 cm. Combination therapy with temozolomide and bevacizumab has recently emerged as a potentially promising regimen for malignant SFTs.[6] These clinically aggressive tumors with local recurrence and distant metastasis exhibit features such as hypercellularity, cellular atypia, tumor necrosis, >4 mitoses/10 high-power fields and infiltrative margins.[1] Our case presented with a tumour >10 cm and histopathology revealed high-grade malignant features, including increased mitotic activity, atypia and necrosis.

A major breakthrough in understanding SFT came with the discovery of **NAB2–STAT6 gene fusion**, which leads to formation of a chimeric protein that promotes tumor cell growth by activating **EGR1**, a gene involved in cell proliferation. This fusion is considered a **driver mutation** and a useful and highly specific diagnostic marker, especially when detected via nuclear STAT6 expression on IHC.[4] Additionally, bFGF and Ki67 labeling indexes are useful in assessing malignancy and may have prognostic value.[3] TERT promoter mutations represent another important marker, associated with increased telomerase activity, older age, larger tumours, higher mitotic rates and poorer prognosis.[5]

Differential diagnosis includes fibrosarcoma, malignant peripheral nerve sheath tumor, malignant fibrous histiocytoma and dermatofibrosarcoma protuberans.The closest mimic is synovial sarcoma, considered in our case due to overlapping histologic features.[6] Synovial sarcoma is a highly cellular spindle cell neoplasm with a rich vascular network including ectatic and sinusoidal vessels. Distinguishing these entities is clinically important, as synovial sarcoma is relatively chemosensitive, whereas malignant SFT responds poorly to chemotherapy and radiotherapy. IHC and molecular studies are helpful in making the distinction. TLE1 positive tumor cellsand SSXrearrangement by FISH, helps in confirming the diagnosis of synovial sarcoma, whereas SFT is immunoreactive to CD34 (90%–95%), EMA, SMA, CD99 and bcl2 with more specific staining demonstrated by nuclear STAT6.[7]

A correct diagnosis is mandatory for proper therapy and management of patients with extra pleural SFT, as it is more aggressive than pleural form, particularlycases occurring in mediastinum, retroperitoneum, pelvis and meninges. Although SFT is usually considered a clinically indolent neoplasm, prognosis is unpredictable and only partially related to morphological features.[4]

Conclusion

This case emphasizes the importance of considering SFT in differential diagnosis of large soft tissue tumours in extremities. Accurate diagnosis relies onhistologic features and IHC, particularly STAT6. Given the potential for recurrence or metastasis, even in tumours with deceptively indolent features, long-term follow-up is crucial. This report adds to literature on malignant extrapleural SFTs and supports the need for awareness and vigilance in diagnosis and management.

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