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Solitary Fibrous Tumor of the Breast: A Rare Case and its Diagnostic Pitfalls

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ABSTRACT

Background: Solitary fibrous tumors (SFT) are the rare mesenchymal tumors originally described in the pleura. SFT of breast is even rarer and to the best of our knowledge about 35 cases are reported to date, including only six malignant SFT cases.

Case presentation: We report a case of a 52-year-old lady with a large left breast mass involving all the quadrants. The tumor was diagnosed as malignant SFT in a core needle biopsy which was later confirmed on the resection specimen.

Conclusion: Herein, we describe the approach and importance of optimal utilization of immunohistochemistry for diagnosing such rare tumors of the breast, particularly, when clinical presentation, radiology and fine needle aspiration cytology are incongruous.

INTRODUCTION

The solitary fibrous tumor is one of the rare mesenchymal origin tumors with an incidence of <0.1/100,000 people.[1,2] This tumor tends to pose diagnostic challenges due to similarity in clinical, radiological and histological features with several more common soft tissue tumors like synovial sarcoma, leiomyosarcoma, phyllodes, malignant peripheral nerve sheath tumor (MPNST), etc. The tumor was previously reported to primarily occur in pleura which is still the most common site accounting for 70% of the cases. However, now it is known to occur at any anatomical sites with a varying spectrum of histological features. Histologically, it ranges from hypocellular to hypercellular tumors to anaplastic SFT with sarcomatous transformation.[3] Fibroblastic cell is considered as the cell of origin in these tumors. Historically, SFTs are sub-classified into three groups: i) Benign (local disease); ii) not otherwise specified (usually not metastatic); and iii) malignant.[4] The general criteria adopted for malignant SFTs include a large tumor size, mitotic rate of ≥4/10 high-power fields (HPFs), nuclear pleomorphism, and necrosis. Demicco *et al.* proposed a modified four-variable risk stratification model for development of metastasis in solitary fibrous tumors based on age, tumor size, necrosis and mitotic count into three risk classes of low, intermediate, and high risk.[5] The diagnosis as well as risk stratification of SFT at unusual sites like the breast are very demanding and not possible on clinical and radiological examination. Accurate diagnosis requires a combined evaluation of clinical, pathological, immunohistochemical and molecular features together. Herein, we report a case of malignant SFT in a 52 year-old lady with a focus on diagnosis.

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CASE PRESENTATION

A 52-year-old lady presented with a progressively enlarging lump in the left breast over the last 12

months. On examination, a large, painless mass was noted measuring approximately 10cm in the greatest dimension involving all four quadrants. The overlying skin and nipple areola complex were unremarkable. Earlier, the patient underwent a mammographic examination in a private hospital which revealed a well-defined lobulated dense lesion in superior lateral and superior medial part of the left breast measuring 8.2X5.1cm; Breast Imaging-Reporting and Data System (BIRADS) category 4B (Figure 1A, 1B).

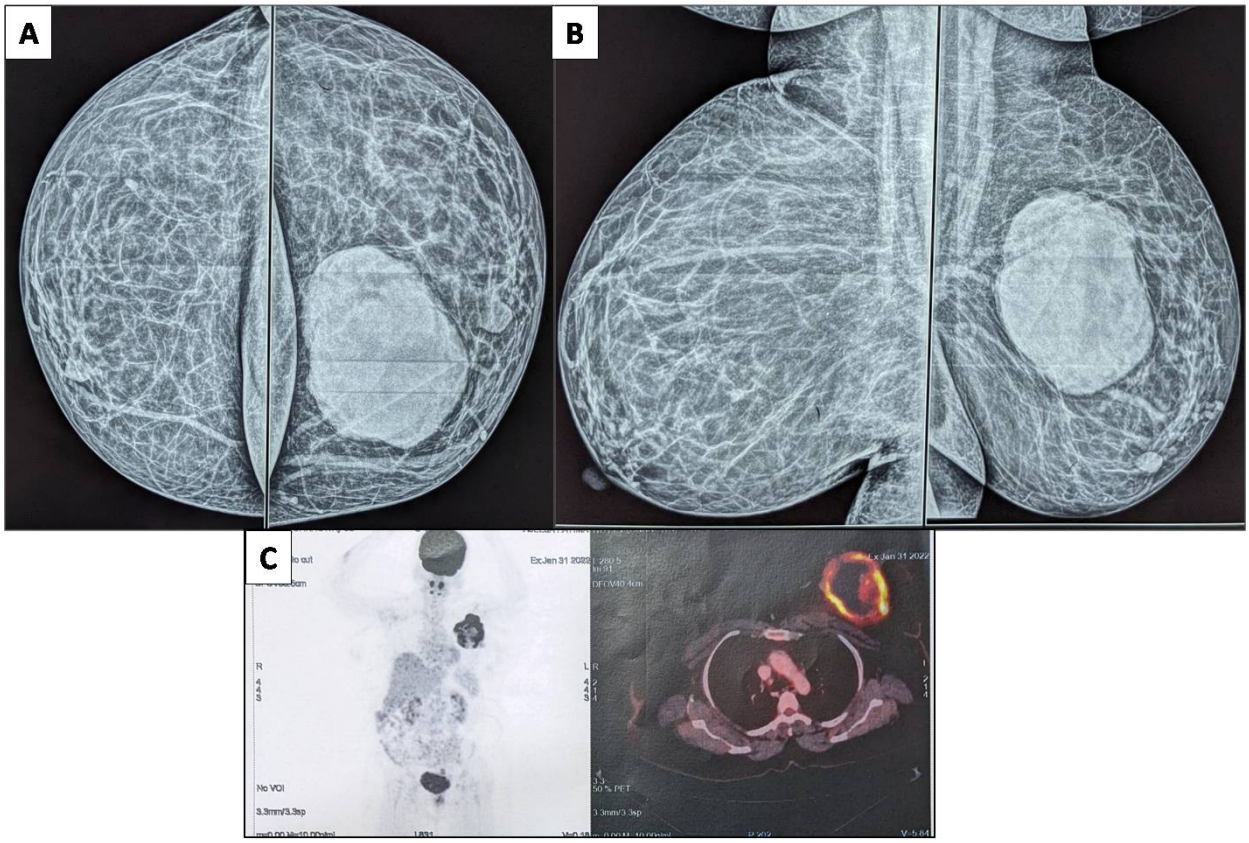

Figure 1. Mammogram showing a well-defined lobulated dense lesion in superior lateral and superior medial part of the left breast measuring 8.2 X 5.1 cm (A, B). PET scan showing a FDG avid soft tissue density mass involving the entire quadrant with hypo metabolite areas suggestive of necrosis (C).

A biopsy was also done outside which confirmed the lesion as a phyllodes tumor. At our center, a biopsy was repeated by the surgeon to confirm the diagnosis. For metastatic work-up, given the rapidly enlarging large mass, a positron emission tomography (PET) scan was also done. The PET scan showed a FDG avid soft tissue density mass measuring 8.4 (Anteroposterior) x 7.6 (mediolateral) x 8.8 (craniocaudal) cm involving the entire quadrant with hypo metabolite areas suggestive of necrosis (Figure 1C). No significant family history was present.

The core needle biopsy showed a cellular tumor comprising spindle to oval cells arranged in long fascicles, and a focally herringbone pattern with intervening capillary channels in a characteristic hemangiopericytomatous pattern. The tumor cells showed moderate nuclear pleomorphism, frequent mitotic activity including atypical forms (5/10 hpf), and an infiltrative growth pattern. Perivascular accentuation by tumor cells and the area of hyalinization were also seen. A focal area of necrosis was noted. No glandular/ductal component was seen. The possibility of a primary mesenchymal tumor with the following differentials was considered: Solitary fibrous tumor (SFT), synovial sarcoma, malignant phyllodes, leiomyosarcoma, metaplastic carcinoma and malignant peripheral nerve sheet tumor (MPNST). An immunohistochemistry (IHC) panel comprising CK, ER, PR, CD34, SMA, STAT6, Caldesmon, SOX10, TLE-1, and MIB-1 was applied to confirm the diagnosis. The tumor cells showed diffuse nuclear positivity for STAT6 with CD34 highlighting blood vessels and were negative for CK, ER, PR, Caldesmon, SMA, SOX10, and TLE-1. The MIB-1 labeling index was approximately 20%. Based on the histomorphology and IHC findings a diagnosis of SFT, the intermediate risk group was suggested with advice for NAB2-STAT6 fusion gene study. Following the histopathology report, surgeons performed a modified radical mastectomy. On gross

examination, a large grey-white fibrous lobulated unencapsulated tumor measuring 8 x 7.5 x 6 cm involving entire quadrants with area of necrosis was seen. A total 16 lymph nodes were retrieved from the attached axillary tail. The tumor showed similar histological features as in the core biopsy. The lymph nodes showed reactive changes, free of tumor.

The patient was asymptomatic until the last follow-up four months after the surgery.

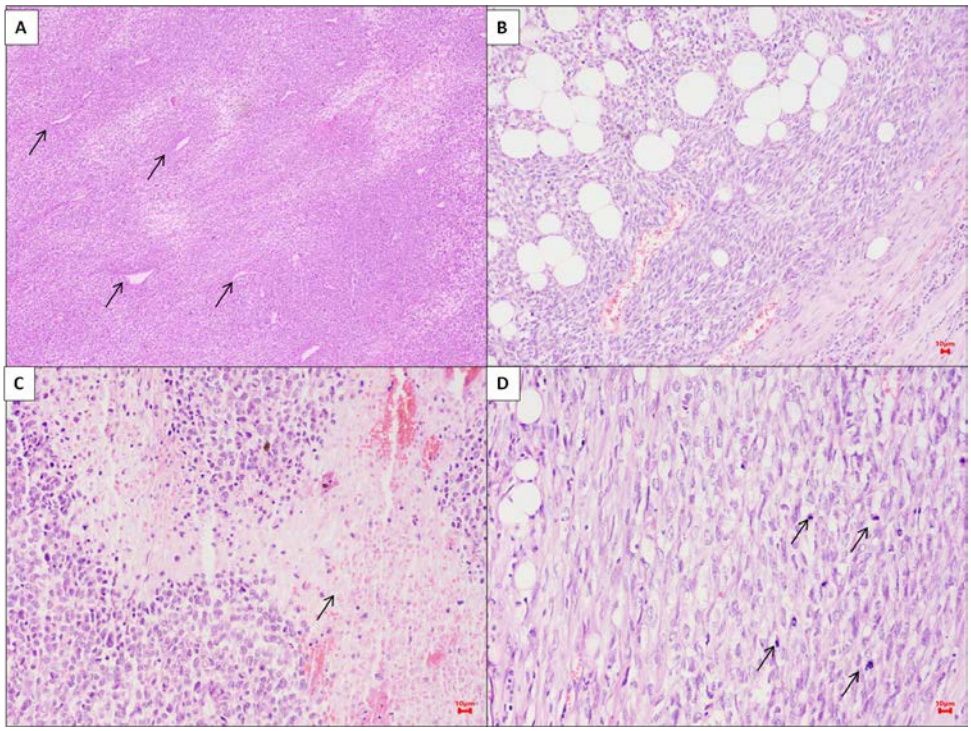

Figure 2. The histological images showing (A) a tumor arranged in long fascicles, and with intervening capillary channels in a characteristic hemangiopericytomatous pattern (arrow) (H&E x40), (B) The tumor cells show an infiltrative growth pattern and perivascular accentuation (H&E x100), (C) Focal area of necrosis (arrow) (H&E x100), and (D) moderate nuclear pleomorphism, frequent mitotic activity including atypical forms (arrow) (H&E x200).

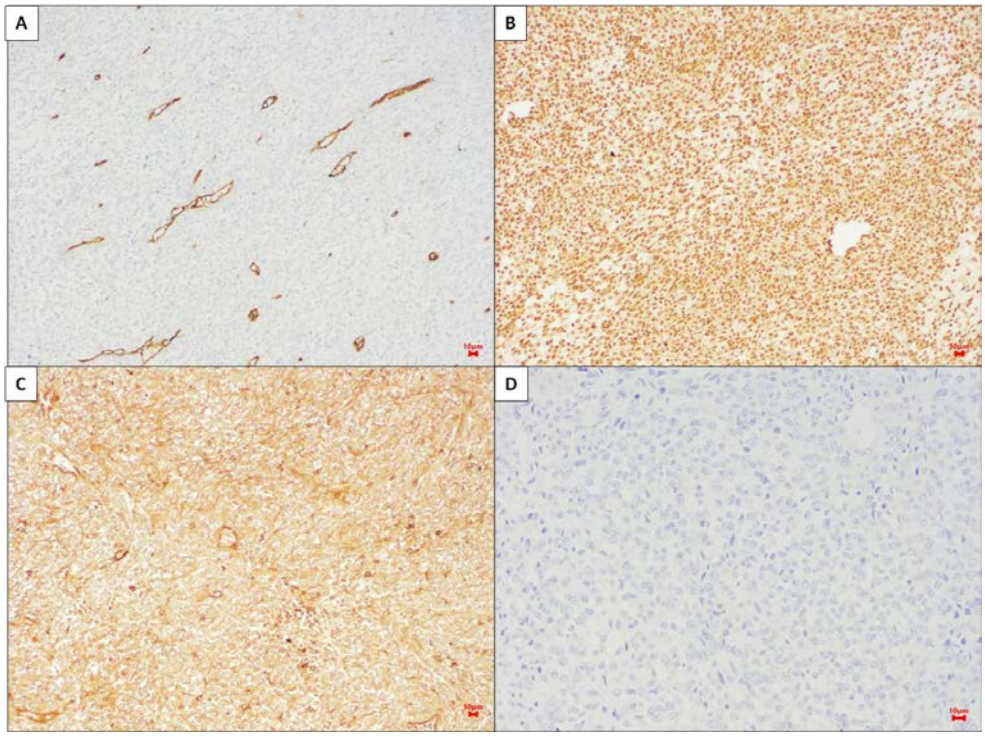

Figure 3. The immunohistochemistry images showing (A) Vascular channels highlighted by CD34, while tumor cells are negative (IHC x100), (B) STAT6 show diffuse and strong nuclear positivity (IHC x100), (C) CD44 diffuse membranous and cytoplasmic positivity (IHC x100), and (D) CK negative (IHC x200).

DISCUSSION

SFTs were formerly known with different terminologies like pleural fibroma, hemangiopericytoma, benign mesothelioma, localized fibrous tumor, giant cell angiofibroma and subserosal fibroma. The peculiar finding in SFT is characteristic staghorn-like blood vessels and perivascular accentuation of the tumor cells. These characteristics are very helpful in making a correct diagnosis of SFT, particularly at unusual sites. Malignant SFTs are usually large, hypercellular tumors displaying at least focal moderate to marked cytological atypia, tumour necrosis, numerous mitoses (≥ 4 mitoses/10 high-power fields), and/or infiltrative margins.[6] In the present case, all the features supportive of malignant SFT described by Vimi *et al.* were present to label it as malignant SFT.[6] In mammography, it was categorized as BI-RADS 4B which is suspicious for malignancy (10-49%). PET scan was suggestive of a malignant lesion.

The modified Demicco Score, which is based on mitotic activity, patient age, tumor size and tumor necrosis to predict the risk of metastasis, is the most commonly used system for the risk stratification of SFTs. Based on this, SFTs can be categorized into three groups: Low, intermediate and high-risk SFTs. However, it is mainly applicable to soft-tissue and pleural SFTs, and valuable in planning the treatment strategy for high-risk SFTs, but its utility for other rare site SFTs as in the breast is not validated. Moreover, it is not an advocated prediction model for the local recurrence in SFTs. The index case, as per the modified Demicco Score fulfilled the intermediate risk category group.

In the present case, the IHC played a critical role in the diagnosis. The minimal panel which helped us comprised CK, ER, PR, CD34, SMA, STAT6, Caldesmon, SOX10, TLE-1, and MIB-1. STAT6 is the most helpful sensitive and specific marker for SFTs.[7] It is even more useful in malignant SFT cases, where the conventional CD34 may be negative. In the current case, CD34 was negative; however, STAT6 showed diffuse nuclear positivity.

We have also evaluated CD44 IHC expression in the present case. CD44 is a cell adhesion molecule that also represents a biomarker of cancer stem cells, and plays an important role in epithelial-mesenchymal transition. Demirag *et al.* evaluated the expression of CD44 and matrix metalloproteinases (MMP-2) in pleuro-pulmonary SFTs.[8] They reported MMP-2 positivity in two malignant SFT cases; however, CD44 expression was observed in benign cases only. In contrast, we found diffuse positivity of CD44 in the present case. The role of CD44 in SFTs pathogenesis and malignant potential needs to be further validated.

To the best of our knowledge, only six cases of malignant SFTs in the breast are reported in the literature. The recurrence rate of malignant SFT is high, and a complete removal is suggested for better outcomes.[9] However, metastasis is rarely documented in any case. In the present case, axillary lymph nodes were free of tumour. PET CT was also not suggestive of any distant metastasis. As a richly vascular tumor treatment options may have angiogenic pathway blockers but due to a limited number of cases, they are still on trial.[10]

CONCLUSION

Due to the infrequency of breast SFT, it diagnosis is a matter of exclusion. Clinical and radiological features are not specific to make a certain diagnosis and the usual differentials considered are phyllodes tumor or carcinoma. A careful histological examination and IHC interpretation are very important to make an accurate diagnosis.

ETHICAL CONSIDERATIONS

Informed consent from the patient was obtained.

CONFLICT OF INTERESTS

None to declare.

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REFERENCES

1. Klemperer P, Coleman BR. Primary neoplasms of the pleura. A report of five cases. *Am J Ind Med*. 1992;22(1):1-31. doi: 10.1002/ajim.4700220103.
2. Kinslow CJ, Wang TJC. Incidence of extrameningeal solitary fibrous tumors. *Cancer*. 2020;126(17):4067. doi: 10.1002/cncr.33057.
3. Kazazian K, Demicco EG, de Perrot M, Strauss D, Swallow CJ. Toward Better Understanding and Management of Solitary Fibrous Tumor. *Surg Oncol Clin N Am*. 2022;31(3):459-483. doi: 10.1016/j.soc.2022.03.009.
4. Wang Y, Wang L, Huang J, Liu X, Sun H, Sui X, et al. A giant breast malignant solitary fibrous tumor: A rare case report and brief review. *Oncol Lett*. 2023;25(6):249. doi: 10.3892/ol.2023.13835.
5. Demicco EG, Wagner MJ, Maki RG, Gupta V, Iofin I, Lazar AJ, et al. Risk assessment in solitary fibrous tumors: validation and refinement of a risk

stratification model. *Mod Pathol*. 2017;30(10):1433-1442. doi: 10.1038/modpathol.2017.54.
6. Vimi S, Punnya VA, Kaveri H, Rekha K. An aggressive solitary fibrous tumor with evidence of malignancy: a rare case report. *Head Neck Pathol*. 2008;2(3):236-41. doi: 10.1007/s12105-008-0073-2.
7. Schweizer L, Koelsche C, Sahm F, Piro RM, Capper D, Reuss DE, et al. Meningeal hemangiopericytoma and solitary fibrous tumors carry the NAB2-STAT6 fusion and can be diagnosed by nuclear expression of STAT6 protein. *Acta Neuropathol*. 2013;125(5):651-8. doi: 10.1007/s00401-013-1117-6.
8. Demırağ F, Cakir E, Alpar S, Taştepe I, Kaya S. Expression of CD44 and MMP-2: possible association with histopathological features of pleuro-pulmonary solitary fibrous tumors. *Turk Patoloji Derg*. 2011;27(2):127-33. doi: 10.5146/tjpath.2011.01060.
9. Vallat-Decouvelaere AV, Dry SM, Fletcher CD. Atypical and malignant solitary fibrous tumors in extrathoracic locations: evidence of their comparability to intra-thoracic tumors. *Am J Surg Pathol*. 1998;22(12):1501-11. doi: 10.1097/00000478-199812000-00007.
10. Park MS, Ravi V, Araujo DM. Inhibiting the VEGF-VEGFR pathway in angiosarcoma, epithelioid hemangioendothelioma, and hemangiopericytoma/solitary fibrous tumor. *Curr Opin Oncol*. 2010;22(4):351-5. doi: 10.1097/CCO.0b013e32833aaad4.

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