

Paratesticular Solitary Fibrous Tumour Mimicking Cellular Angiofibroma: An Unusual Morphology and Rare Site

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ABSTRACT

Solitary fibrous tumour (SFT) is a ubiquitous benign mesenchymal tumour of fibroblastic origin, which occurs most often in middle-aged adults. It usually presents as lung mass originating from pleura, but extrapleural occurrence is also common. Tumour is characterised by hypo- and hyper-cellular areas of spindle-shaped cells, arranged in haphazard manner with dispersed staghorn-shaped vessels. Surgical excision is the curative treatment. SFTs of the primary testicular or paratesticular region are extremely rare, but they exhibit histologic findings similar to SFTs originating at other body sites. Here, we report the case of a paratesticular SFT in a 37-year male, who presented with a non-tender, firm, mobile, left-sided retrotesticular swelling with unique histological features closely mimicking cellular angiofibroma, a common tumour of paratesticular location.

Key Words: Solitary fibrous tumour, Paratesticular, benign, angiofibroma.

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INTRODUCTION

Solitary fibrous tumour (SFT) is a slow-growing, spindle cell tumour characterised by NAB2-STAT6 gene fusion.¹ It usually arises from the visceral pleura; however, extra-thoracic occurrence at various locations has been reported.² The tumour has tendency to occur at any age, but the most common incidence of pleural SFT is in the sixth to seventh decade. There is no sex predisposition and the causative factor is still unknown.³ SFT is usually benign, but malignancy can be seen in 10% of the cases. Local recurrence rate is 10% and metastatic potential approaches 5-10%. The tumour on histology demonstrates a haphazard spindle cell fibroblastic proliferation with variable cellularity. Stromal and perivascular hyalinisations are typically present along with hemangiopericytoma-like vessels of variable number and size.⁴ SFT shows positive expression of immunomarkers CD34, CD99 and STAT6.⁵ Occurrence of SFT in paratesticular area is very uncommon; moreover, unusual morphology is extremely rare.

We, herein report the case of a 37-year male who presented with left retrotesticular swelling. The patient underwent surgical excision and the tumour histology closely mimicked cellular angiofibroma. SFT was eventually diagnosed, based on positive nuclear staining for STAT6.

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CASE REPORT

A 37-year male presented with the complaint of a painless, firm, mobile, nodular left retrotesticular swelling. Ultrasound showed a 13×9.5×6 cm paratesticular swelling.

Computed tomography (CT) revealed a circumscribed, firm, lobulated extratesticular mass without inguinal lymphadenopathy. Surgical excision was done. The specimen received was coded as "Left retrotesticular swelling". Grossly, it was an encapsulated nodular tissue mass that measured 13×9.5×6 cm. The cut surface was tan-white and rubbery. No normal testicular tissue was identified.

Microscopy showed a lesion composed of intersecting fascicles and haphazardly arranged cells (Figure 1). There were hypo-cellular areas separated by thick bands of hyalinised keloidal collagen. There was perivascular and interstitial hyalinisation. Scattered small to intermediate-sized vessels were present ranging from thin-walled branching vessels to thick-walled vessels exhibiting myocyte proliferation and intimal thickening (Figure 2A). Individual cells were ovoid to spindle-shaped, had indistinct borders, scant to moderate pale cytoplasm, and vesicular nuclei with finely dispersed chromatin. Multinucleated stromal giant cells were also present. The mitotic count was 1-2 mitoses/10HPF. No significant necrosis or atypia was appreciated.

Immunohistochemical stains were performed and tumour cells showed positivity for CD34, BCL-2, STAT6 and ASMA; whereas, desmin was negative. Based on all these features, this tumour was diagnosed as SFT. Later, orchidectomy and removal of the adjacent paratesticular soft tissue and scrotal skin were performed due to close margins and no evidence of

testicular involvement or residual tumour in paratesticular soft tissue was seen.

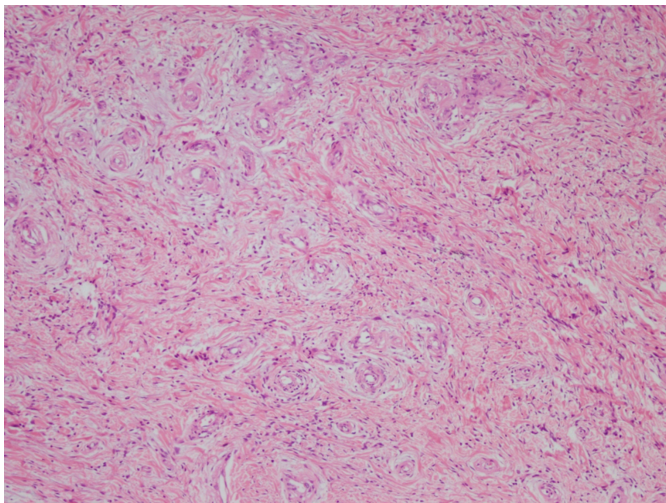


Figure 1: Haphazard arrangement of spindle cells imparting hypocellular appearance with scattered numerous small to intermediate sized, round, hyalinised blood vessels in a collagenised background (H&E, ×200).

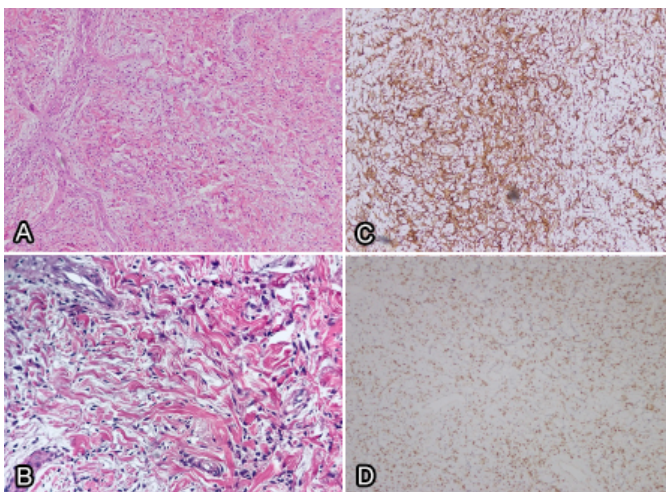


Figure 2: (A) Spindle cells arranged in random manner with hyalinised branching vessels. (H&E, ×100). (B) Another view showing spindle cells with hyperchromatic, relatively plump, wavy serpentine nuclei (H&E, ×400). (C) Positive membranous CD34 stain (CD34, ×100). (D) Positive nuclear STAT6 stain (STAT6, ×100).

DISCUSSION

SFTs are benign tumours composed of fibroblast-like cells. They are common elsewhere in the body including pleura, upper respiratory tract, lung, mediastinum, breast, meninges, liver and pelvic cavity; however, only few cases of paratesticular origin have been reported.⁴ Similar to their behaviour at other anatomic sites, paratesticular SFTs with low proliferative activity have little potential for local recurrence or metastatic spread.⁶

SFTs are histologically characterised by hyper- and hypo-cellular areas of spindle-shaped cells with pattern-less architecture. The cells have oval nuclei with scant pale cytoplasm. Interspersed thin-walled staghorn-shaped blood vessels, hyalinisation and keloidal collagen are present.⁷ Mitotic activity is

usually low. This tumour was different from classic histology of SFT due to hypocellular spindle appearance and the presence of florid proliferation of round-hyalinised medium-sized blood vessels, features characteristic of cellular angiofibroma, a common superficial soft tissue tumour of the genital region.⁸ In areas, tumour cells showed serpentine nuclei with thin collagen demonstrating neural-like appearance (Figure 2B).

Immunohistochemistry-depicted positive CD34 expression, feature common to both tumours (Figure 2C). Immunomarker ASMA was also positive, raising possibility of smooth muscle neoplasm in differential; but desmin was negative and S-100 was also negative to exclude neurofibroma, which typically shows positive CD34 expression. Nuclear STAT6 expression confirmed the diagnosis of SFT (Figure 2D).⁹ Another common tumour at this location is well-differentiated liposarcoma, sclerosing type, which shows well differentiated adipocytic component, fibrous septae, atypical stromal cells and lipoblasts, features absent in this case.

Therefore, SFT should be considered in the differential diagnosis whenever a spindle cell tumour is seen at paratesticular area.¹⁰ Positive CD34 staining with nuclear STAT6 expression can be extremely helpful in establishing diagnosis in histologically challenging cases.

CONFLICT OF INTEREST:

The authors declared no conflict of interest.

PATIENT'S CONSENT:

Informed consent has been taken from the patient.

AUTHORS' CONTRIBUTION:

MBQ: Drafted the case summary and discussion, and edited the manuscript.

MU, QC, NU: Established and confirmed the diagnosis, drafted the manuscript and references.

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