



Solitary Fibrous Tumor of Head & Neck Region: Problems in Diagnosis Due to “Borderline” Features

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Authors' contributions

This work was carried out in collaboration between all authors. Author SVA diagnosed the case, designed the study and wrote first draft and rectified the manuscript draft to its perfection. Author VCA helped to write first draft. Author AJ managed the literature searches with help of authors AKT and AGV reviewed the manuscript. All authors have read and approved the final manuscript.

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Case Study

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ABSTRACT

Aims: To report SFT in a 45 years male who had a lobulated tumor in neck for longest duration of 18 years with borderline histomorphology.

Presentation of Case: The case discussed is of a forty-five year old Indian man who presented with painless mass in right side of neck for the duration of 18 years with rapid enlargement in last one year.

Discussion: Solitary fibrous tumor is an unusual spindle cell neoplasm arising from primitive mesenchymal cell with distinct “patternless pattern” on histomorphology. The commonest site for this tumor is pleura but in recent years it has been described in various anatomic sites and is known to involve any region of the body. The extra- pleural sites include extremities, pelvis, head and neck and urogenital region. SFT of Head and Neck region is rare and usually involves deep soft tissues. Morphologically SFT resembles many benign and malignant soft tissue tumors.

Conclusion: SFT is a rare tumor in neck region and has to be differentiated meticulously from other common and rare soft tissue tumors. Close follow up is essential after complete surgical excision in cases with “borderline” histomorphology.

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ABBREVIATIONS

SFT- Solitary fibrous tumor.

H &E- Hematoxylin and eosin.

1. INTRODUCTION

Solitary fibrous tumor is a rare, mesenchymal tumor affecting mainly middle aged adults with no sex predominance however can occur in younger patients [1-5].

This spindle cell neoplasm usually arises in the pleura but is known to involve extra-pleural sites in 50-70% of cases [1,2]. These extra-pleural sites include extremities, pelvis, head and neck and urogenital region [1-4]. About 40% of extrapleural SFTs are located in subcutaneous tissue while 60% are located in the deep soft tissues of extremities, abdominal cavity, retro peritoneum and head neck region [6].

Recently Vogel et al. [7] have reported 28 cases of SFTs at different sites (both thoracic and extra- thoracic) in which they have described only 2 cases of SFTs in head and neck region with one case in the neck region. In head and neck the commonest site is oral cavity followed by orbit, nose and paranasal sinuses [4]. Subcutaneous tissue of neck is an unusual site for this tumor.

Estimated 5-20% of thoracic solitary fibrous tumors may have malignant features but malignant extrathoracic tumors are rare. However the clinical behavior of extra-thoracic SFT is unpredictable [3,5].

According to Wilky et al. [4], extra-thoracic solitary fibrous tumors are more likely to have atypical histological features. We report SFT in a 45 year male who had a tumor in neck for longest duration of 18 years with borderline histomorphology

2. CASE PRESENTATION

A 45-year-old non-smoker, non-alcoholic male presented to surgical OPD with painless swelling involving right side of neck. A small swelling was present in right side of neck since 18 years; however there was rapid enlargement with lobulation over a period of one year. There was no past history of trauma, surgery or any major medical illness. There was no history of fever, dyspnea, night sweats or weight loss.

Local examination revealed large, ovoid, lobulated swelling of 10.5X8.5 cm (Fig. 1a). The overlying skin was stretched with focal ulceration.

CT scan revealed well defined lobulated lesion showing enhancement in subcutaneous tissue on right side of neck (Fig.1b and 1c). USG neck revealed a large well defined, smoothly marginated, heteroechoic lobulated mass in the right side of neck.

Due to sudden enlargement of the lesion which was existing for 18 years, the clinical suspicion of malignancy arising in benign tumor was strong. Thus the mass lesion was excised and sent for histopathological examination.

Pathological examination: Gross examination revealed 10.5X8.5X7 cm well circumscribed lobulated tumor. Cut surface of the tumor was solid, grayish white, glistening without areas of hemorrhage and necrosis (Fig. 2a).

Microscopic examination revealed a well circumscribed tumor located in the dermis. Tumor showed spindle cell proliferation with varying cellularity and hemangio-pericytoma like vessels. The hyper-cellular areas showed spindle cells arranged in fascicles whereas hypo-cellular areas showed pattern-less pattern with extracellular collagen deposition. The cells had eosinophilic cytoplasm, oval bland looking nuclei with minimal mitoses (<2/10 HPF). The nuclear pleomorphism was minimal and necrosis was absent (Fig. 3a, 3b and 3c). Immunohistochemically tumor cells showed diffuse positivity for CD34 and CD99 (Fig. 3d and 3e), but were negative for S100 protein and desmin. Based on histomorphology and immunohistochemistry the diagnosis of solitary fibrous tumor was rendered.

3. DISCUSSION

SFT of soft tissues first described in 1931 by Klemperer and Rabin is a rare entity [1-10]. Most tumors present as a slow growing painless mass [5], but some of the tumors may cause localized pain due to mass effect [8]. The duration of extrathoracic solitary fibrous tumor ranges from 1 month to 6 years [1,2,3,9]. The longest duration for SFT is 20 years which has been reported in paratesticular region by Lee et al. [11]. To our knowledge present case study has longest duration of solitary fibrous tumor for 18 years in

the neck region which has not been described in any case report till date.

SFT occurring in the soft tissue presents as a well circumscribed mass ranging in size from 1-6 cm. Cut surface is usually solid, grey white to pinkish brown with lack of necrosis or hemorrhage [1-3].

Microscopically this tumor has characteristic appearance of "patternless pattern" where the cells are haphazardly embedded in the collagenous stroma [3-7]. Immunohistochemically these cells show strong and diffuse positivity for CD 34, CD 99 and variable positivity for Bcl-2 [9,10]. The other patterns which can be observed are short fascicles, storiform pattern,

hemangio-pericytomatous pattern and leiomyoma-like pattern [3]. The vasculature varies from slit like and dilated vessels to branching vessels with typical 'staghorn' appearance. The nuclei of spindle cells usually have bland appearance with minimal pleomorphism and absent mitotic activity. This appearance of solitary fibrous tumor has to be mainly differentiated from the benign and malignant neurogenic tumors, 'desmoid' fibromatosis, and myofibroblastic sarcoma. The diagnostic problems arise as this tumor is usually not considered in the differential diagnosis clinically and pathologically and also due to the fact that morphologically it may mimic other tumors.



Fig. 1a. Clinical photograph showing large lobulated mass in the neck with stretched out overlying skin



Fig.1b.



Fig.1c.

Fig. 1b and 1c. CT scan of Head Neck region showed well defined lobulated lesion on the right side of neck with enhancement in subcutaneous tissue

Solitary neurofibroma presents as a nodular lesion and histomorphologically is characterized by shredded carrot appearance with spindle cells having wavy buckled nuclei. Immunohistochemically they are positive for S-100 antigen and are weakly positive for CD 34.

MPNST morphologically can be differentiated from SFT by presence of sweeping fascicles of spindle cells with cellular and nuclear pleomorphism and brisk mitotic activity. Tumor cells express S-100 antigen but are negative for CD34 [6].

Low grade myofibroblastic sarcoma differs from SFT by presence of diffuse infiltrative growth pattern with fascicles or storiform pattern. The nuclei are fusiform with indentation and small nucleoli [12]. Immunophenotypically the tumor cells are actin and desmin positive. They may express CD34, CD99 focally which is in contrast to diffuse expression seen in SFT [12].

Desmoid type fibromatosis is distinctly characterized by ill circumscribed mass comprised of sweeping fascicles of spindle cells in collagenous stroma [13]. Like SFT they have low mitotic activity, prominent vasculature and no necrosis but immunohistochemically the cells are strongly positive for vimentin and are negative for CD34 and CD99.

According to recent WHO classification, extrapleural SFT is classified into intermediate

(rarely metastasizing) category [6]. Solitary fibrous tumor usually has a benign course but malignant transformation is known to occur. The criteria for malignancy in SFT are a) Large size (>10 cm) b) High cellularity c) Mitotic activity (>4/10HPF) d) Necrosis [6].

In the studies by Vallat-Decouveleare et al. [8] and Ganly et al. [5], lack of circumscription, nuclear atypia, areas of increased cellularity, necrosis and >4/10 HPF mitoses were suggested as atypical features and predictive factors for malignant behavior and high chances of relapse.

In our case the tumor was large in size with lobulation, had high cellularity, low mitotic activity (<4/10 HPF) and no necrosis. Also clinically there was sudden increase in the size of a long standing tumor, which indicates that this tumor may have high chances of recurrence and thus needs regular follow up.

A study by Wilky et al. [4], supports that extra-thoracic and "Borderline" SFTs with any of England's criteria should be considered high risk tumors. As these SFTs have high recurrence rate and behave aggressively, surveillance imaging for upto 10 years, increasing the screening interval after 2 years is recommended [4].

Treatment for SFT is conservative surgery and resectability is the most important risk factor.



Fig. 2a



Fig. 2b

Fig. 2. 2a and 2b. Grossly tumor measured 10.5X8.5X7 cm with lobulated appearance. Cut surface was solid greyish white glistening with no areas of hemorrhage or necrosis

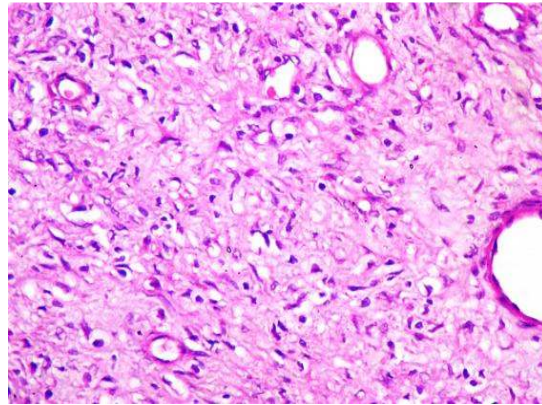


Fig. 3a

Fig. 3a. Microscopic examination showed spindle cell tumor with “patternless pattern” without necrosis (H&E, X100)

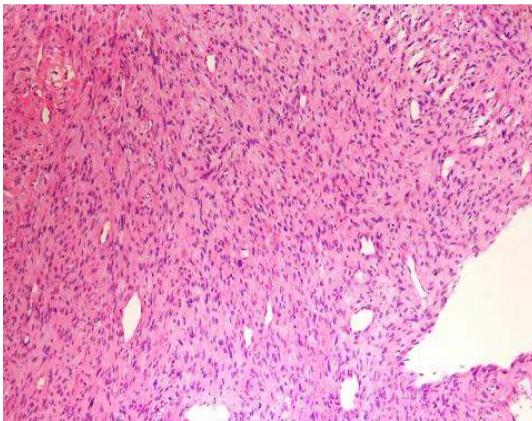


Fig. 3b

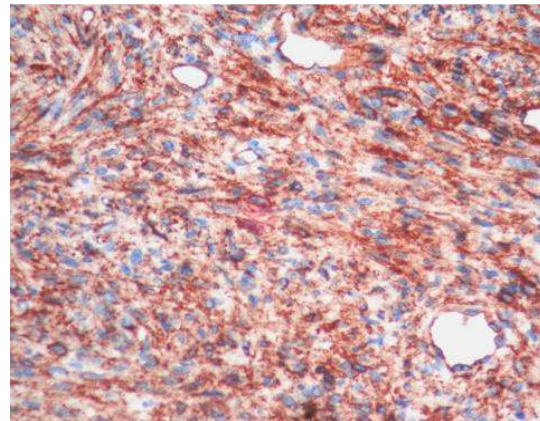


Fig. 3c

Fig. 3b and 3c. Focally tumor showed high cellularity with spindle cells arranged in fascicles (H&E, X100); spindle cells had indistinct cytoplasmic margins and were embedded in collagenous matrix, nuclei were plump to spindle with low mitotic activity (H&E, X40)

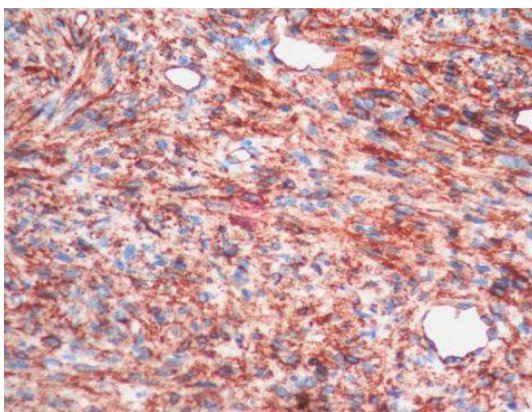


Fig. 3d. CD 34 positivity of tumor cells

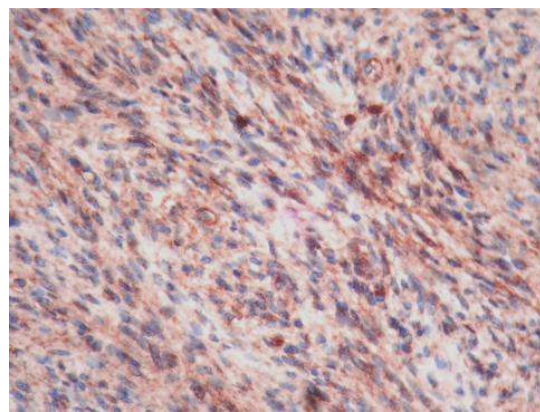


Fig. 3e. CD 99 positivity of tumor cells

4. CONCLUSION

SFT is a rare tumor in neck region and has to be differentiated meticulously from other common and rare soft tissue tumors. Close follow up is essential after complete surgical excision in cases with “borderline” histomorphology.

CONSENT

All authors declare that written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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