CASE REPORT

Solitary Fibrous Tumor Presenting as a Parapharyngeal Mass

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Abstract

Solitary fibrous tumors (SFT) rarely occur in the neck region and can be easily mistaken for more common tumors if the index of suspicion is not high. They are characterized by remarkable histologic variability. However, immunopositivity for CD34 and Bcl2 with immunonegativity for other markers aid in establishing the diagnosis. Distinction from the other common benign entities is essential, as SFT are tumors of low grade aggressiveness and unpredictable biologic behavior. Complete resection is the key feature determining outcome.

Keywords: Parapharyngeal, solitary, fibrous, tumor.

INTRODUCTION

Solitary fibrous tumors are rare neoplasms, originally reported in the pleura.[1] In the recent years, this tumor is being increasingly recognized at extrapleural sites. These have been documented in the trunk in 42% of cases, extremities in 39%, with head and neck involvement reported in 19% cases only.[2-9]

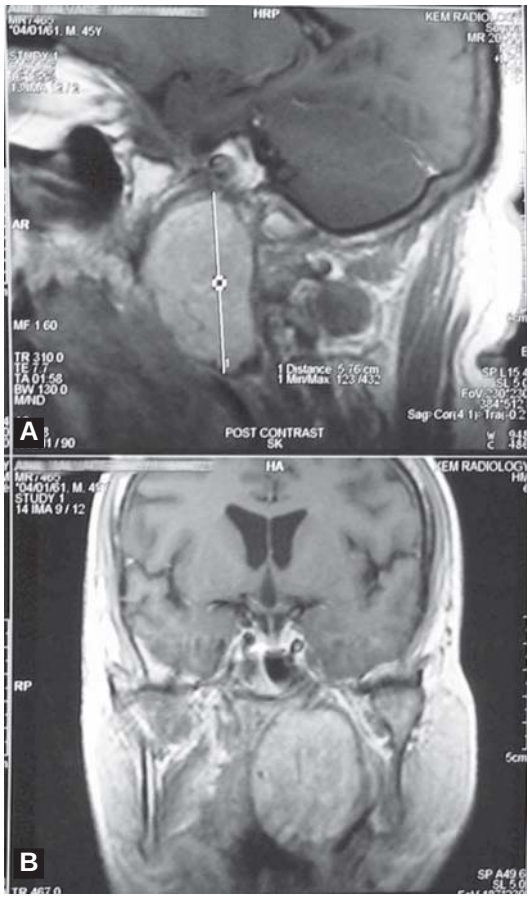
We report a case of parapharyngeal SFT due to its rarity and diagnostic difficulty.

CASE REPORT

A 40-year-man presented with complaints of left nostril blockage and left ear discharge of one month duration. Computed tomography scan (CT) (Figs 1A and B) revealed a well-circumscribed, lobulated mass in the left parapharyngeal space, unrelated to the parotid gland and not infiltrating vascular, neural structures or the overlying mucosa. At surgery the well circumscribed mass was easily shelled out (Fig. 2).

On gross examination, a 6 cm circumscribed, unencapsulated mass showed a smooth lobulated external surface. The cut surface was homogenous, grey white and firm. Formalin fixed, paraffin embedded representative sections were stained by hematoxylin and eosin for light microscopy. Immunohistochemical studies by Streptavidin – biotin complex with appropriate positive and negative controls were carried out. The antibodies included CD34 (monoclonal My10 1:40), vimentin (monoclonal V9 1:40), Bcl2 protein (monoclonal Oncoprotein,), CD31 (Monoclonal JC/70), smooth muscle actin (monoclonal HHF 35), desmin (monoclonal D33, 1:100 1:50), S100 protein, calponin and Ki 67 analog (MiB1).

On light microscopy the tumor had a distinct nodular growth pattern, consisting of alternate hypercellular and hypocellular zones with intervening large ectatic vessels. There was an intimate admixture of mature adipocytes within the tumor (Fig. 3). The hypercellular zones were composed of short spindly cells arranged compactly in a



Figs 1A and B: CT scan showing a well-circumscribed lobulated mass in the left parapharyngeal space

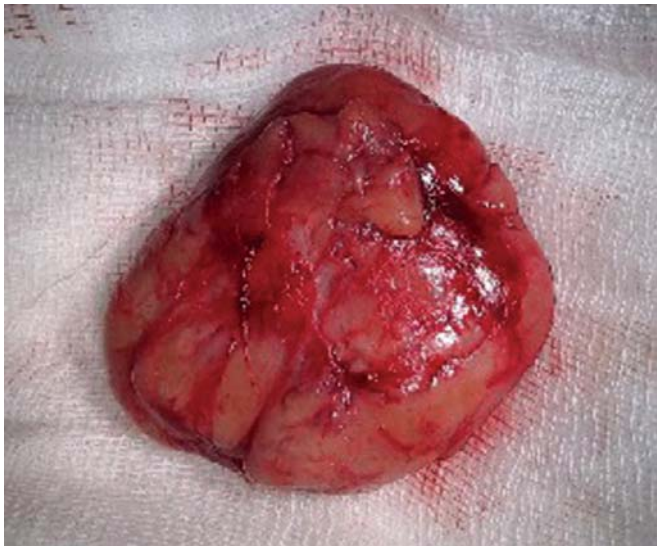


Fig. 2: Gross photograph – well-circumscribed lobulated mass–easily shelled out

vague storiform pattern around a prominent hemangiopericytomatous vasculature. The cells showed bland nuclei and indistinct cytoplasm (Fig. 4). Nuclear pallisading was

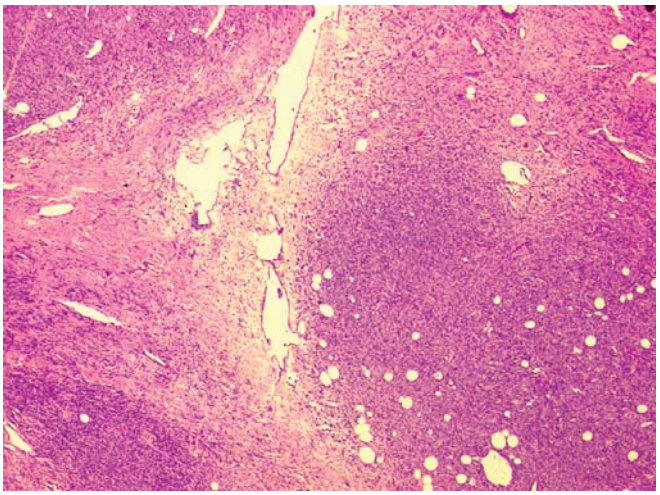


Fig. 3: Nodular growth pattern of the tumor consisting of hypercellular and hypocellular areas separated by large ectatic vessels. Mature adipocytes intimately admixed within the tumor (H and E X 40)

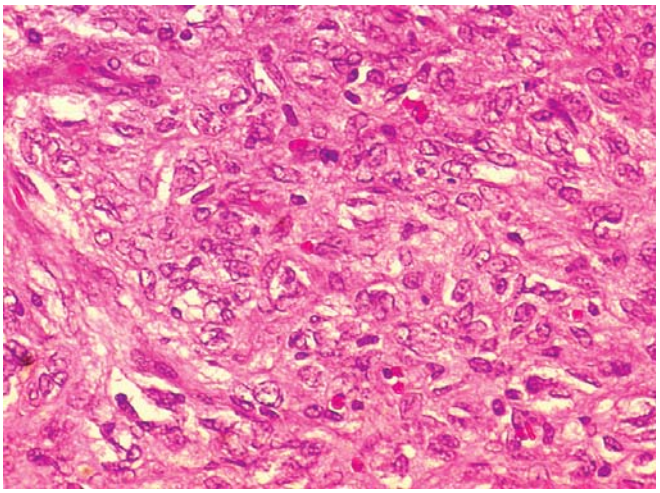


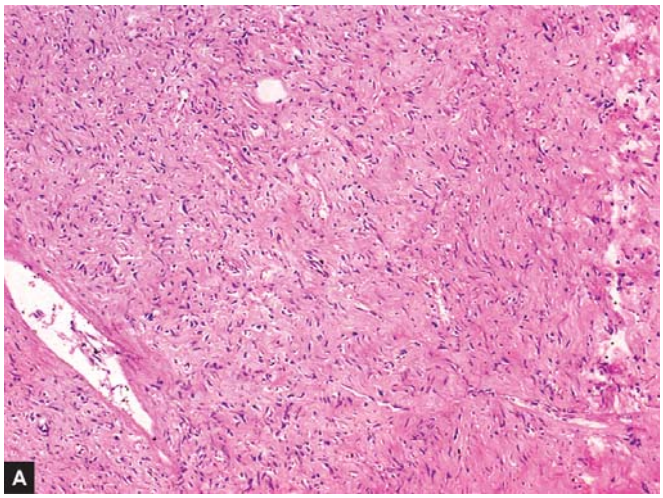
Fig. 4: Higher magnification of hypercellular areas. Spindle to ovoid cells with scanty cytoplasm and bland nuclei with low mitotic activity (H and E X 400)

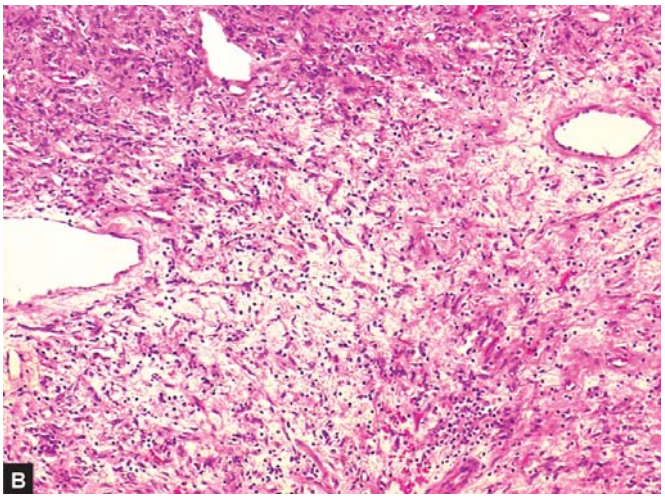
not seen. The hypocellular zone consisted of haphazard proliferation of cells admixed with collagenous and myxoid areas. Lymphocytes were sprinkled through out the tumor (Figs 5 A and B).

The mitotic count in the hypercellular areas was 1-2/10 hpf. Occasional cellular nodule showed up to 4-5 mitoses/10 hpf.

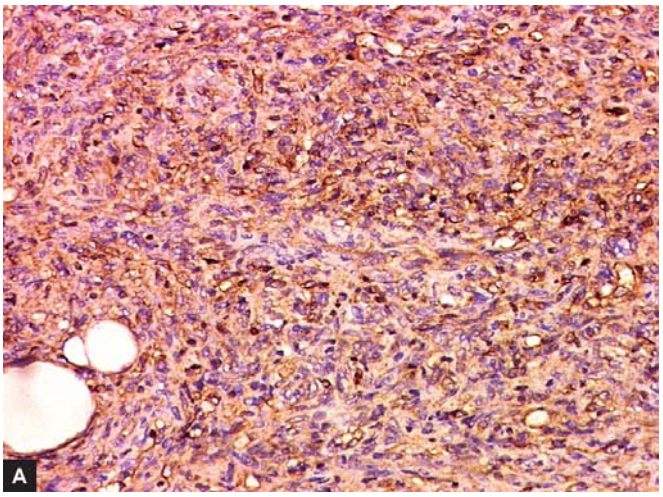
The differential diagnoses included benign peripheral nerve sheath tumor, meningioma and follicular dendritic cell tumor.

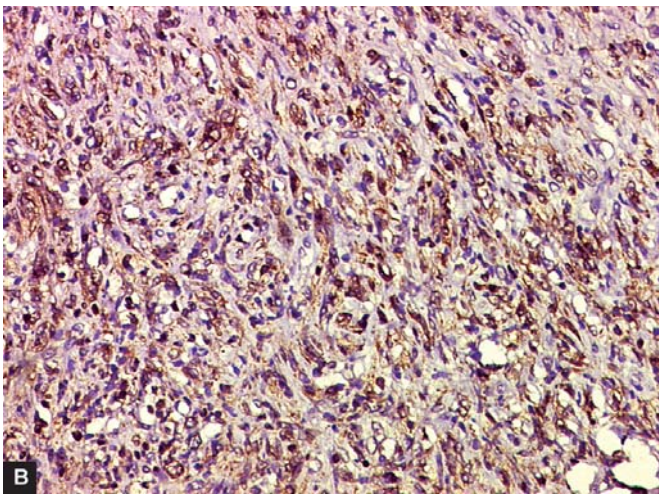
The tumor cells were strongly and diffusely immunopositive for vimentin, CD34 and Bcl2 (Figs 6 A and B) and





Figs 5A and B: Higher magnification of the hypocellular areas. Haphazard arrangement of cells within collagen and myxoid areas with lymphocytes (H and E X 100)





Figs 6A and B: The tumor cells stain strongly and diffusely for CD34 (A) and Bcl2 (B) (ABC X 200)

were immunonegative for S100 protein, Desmin, SMA, EMA, CK, CD21, CD35 and CD31.

A diagnosis of SFT was confidently made in view of the characteristic morphology and immunohistochemistry.

DISCUSSION

SFT were first described in the pleura by Klemperer and Rabin in 1931.[10] These tumors were originally thought to arise from mesothelial cells or submesothelial fibroblasts.[11] Recent studies however propose SFTs to be the neoplastic counterparts of ubiquitous interstitial dendritic cells based on their universal strong diffuse immunoreactivity for CD34.[3,12] Therefore, it is well-accepted that SFT may arise at any location not related to serosal surfaces.[3,13-17] The various sites for the occurrence of SFT in the head and neck region are as shown in Table 1. Orbit is reported as the most common site with the neck structures being rarely involved.

Table 1: Solitary fibrous tumor documented in the head and neck region.[13,14]

Site	*Number of cases*
Meninges, infratemporal fossa	8
Orbit, lacrimal gland	42
Nose, nasopharynx and paranasal sinus	20
Pharynx and parapharyngeal region	5
Epiglottis	1
Oral cavity	10
Salivary gland	9
Thyroid and perithyroid tissue	5

A unifying concept has been proposed that classifies benign or low grade spindle cell lesions in various organs previously designated as hemangiopericytoma, myofibroblastoma and angiofibroma as SFT.[13] This is based on the premises that SFTs show remarkably specific and distinctive

clinicopathologic features which are consistently noted irrespective of the site of involvement.[13] The concept of SFT is still evolving, the terminology is undergoing a change as proposed by Gengler et al.[18] Under the recent nomenclature the present case can be classified as a fibrous variant of SFT.[18]

Clinically most patients are asymptomatic or present with obstructive and compressive symptoms as seen in our case. SFT have been reported more commonly in males, in any age from 17 to 76 years with 53 years as the median age of occurrence.[3,11,13,19,20] Our patient conformed to these demographic details.

SFT is distinguished by its variable histological appearance not only from case to case but also within a given case.[13, 21] Chan et al[13] have proposed diagnostic criteria that incorporate the most consistent features and aim at introducing objectivity in the diagnosis. The criteria include features such as circumscription, alternating hyper and hypocellular areas, bland spindly cells arranged in patternless manner with occasional storiform or fascicular arrangement, low mitotic activity (< 4/10 hpf) and intimate association of the cells with thick collagen.[11,13,20] All these features were well seen in the present case. However, lack of awareness, its histological variability and occurrence at rarely reported sites may lead to SFT being mistaken for more common tumors such as nerve sheath tumors, fibromatosis, salivary gland neoplasms and FDC sarcoma.

Immunopositivity for CD34 and Bcl2 help confirm the diagnosis of SFT and are essential diagnostic criteria for SFTs occurring at extrapleural sites.[3,13,22] The rate of immunopositivity for CD34 varies from 80 to 100% in various studies.[3,13]

Schwannoma is the closest differential diagnosis to SFT. Both tumors are circumscribed, however, schwannomas are encapsulated and related to a nerve. This feature mandates good clinicoradiologic correlation as well as detailed gross examination. Though there are many overlapping light microscopic features, immunopositivity for CD34 is focal and weak in schwannoma while S100 protein is strongly and consistently expressed clinching the diagnosis.

The importance of distinguishing this entity from other more common lesions especially at unusual sites lies in its unpredictable behavior which mandates a wide excision and a close and long-term follow-up. The reported incidence of aggressive behavior varies from 13 to 23% and includes local aggression, recurrence and an occasional metastasis.[1,23,24] Recurrence has been noted in literature as late as 31 years after surgery, thus making a long-term follow-up essential.[11] None of the usual histological criteria of aggressive disease such as tumor size, necrosis, cellularity, pleomorphism or mitotic activity correlate with worse prognosis in the case of SFT.[1,25] Resectability of the tumor is the key prognostic feature. Pedunculated or circumscribed tumors without infiltration into the adjacent tissues, even with mild to moderate atypia tend to have a better prognosis.[1,22,25] Radiotherapy has been advocated for incompletely resected tumors. Chemotherapy is reserved for histologically aggressive tumors.

CONCLUSION

The ubiquity of the interstitial cells and the 'discovery' of SFT at unusual sites have pushed it into limelight. A high index of suspicion and a constellation of morphologic features is essential for diagnosis. Diffuse immunostaining with CD34 is specific, though it may not be seen in 20% of cases. A panel of immunostains therefore is essential for an accurate diagnosis.

One of the most important reasons that SFT has remained in the limelight, is the uncertainty associated with the outcome and the dearth of specific prognostic factors. Though the tumors generally have a favorable outcome, aggressive behavior is known. More studies are required to demystify the ambiguity associated with SFT.

SUMMARY

1. A high index of suspicion is essential for diagnosis of SFT, especially at unusual sites.
2. The entire spectrum of findings including gross, histology and immunohistochemistry must be considered while ruling out the more common entities.
3. Variability of histological appearance is characteristic.
4. Completeness of excision determines outcome.

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