

Solitary Fibrous Tumours In Otorhinolaryngology : A Case Based Illustration

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Citation

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Abstract

Objective: Solitary fibrous tumours (SFTs) are a rare but distinct mesenchymal spindle cell neoplasm that was initially described in pleural tissue. SFT of the head and neck region were largely under-diagnosed till late 1990s. This article reports 3 head and neck presentations of SFT we encountered and managed. The aim is to highlight SFT and help increase its awareness in the domain of the otolaryngologists.

Method: 3 cases of head and neck solitary fibrous tumour are illustrated. We endeavour to highlight the changing concept on SFTs and hemangiopericytomas based on the updated WHO classification of soft tissue tumours.

Results: The first case is a recurrent naso-orbital SFT. The second case presented with a nasal bleed and was found to have a pedunculated mass arising from the nasal septum. The last patient presented with an oral mass. All the patients underwent excision of the mass and were confirmed to have SFT by histopathology and immunohistochemistry. They are being regularly followed up.

Conclusion: SFTs in the head and neck can have varied presentations. To avoid misdiagnosis judicious use of immunohistochemistry is mandatory for spindle cell tumours. Management involves surgical excision. Long term follow-up is imperative since there are no definite features to predict prognosis.

INTRODUCTION

SFT was first described by Klemperer and Rabin in 1931¹. The aetiology is unknown and usually presents in the middle aged and elderly between the fourth and eighth decades of life² without any clear gender predominance³. Studies reveal that they originate from submesothelial soft tissue⁴. We address three cases of SFTs of the head and neck and detail its management.

CASE REPORT 1

A 33 year old lady presented complaining of progressive painless protrusion of the left eye for the past eight months. She was referred to us by the ophthalmologist. In the past about two years and eight months back she was evaluated for a left nasal bleed. The documents affirmed a haemangiomatic mass in the inferior meatus arising from the lateral nasal wall for which she underwent endoscopic clearance of the mass. Biopsy was recorded as SFT.

Patient was otherwise asymptomatic on post operative follow ups for the past 2 years until she noticed

protrusion of the left eye again. On examination there was a soft mass felt in the lacrimal sac area extending 2.5 cm into the medial aspect of the orbital floor. The space between the globe and orbital rim was free in all other areas. The mass was non-reducible with no pulsations, thrill or bruits. Pupillary reflexes were preserved and visual acuity was 20/20 in both the eyes. Nasal endoscopy showed a mass medial to left inferior and middle turbinates. Middle turbinate was thin with the mass pushing the lateral wall into the middle meatus.

Imaging showed a well enhanced mass lesion involving the left inferior and middle meati, ethmoidal cells and inferomedial orbit with involvement of the lacrimal system on the contrast enhanced CT scan (see Fig 1a). In MRI the mass showed isointense signal with respect to gray matter on T1 weighted images. On T2 weighted images heterogeneous signals were shown with strong enhancement with gadolinium.

The primary diagnosis was recurrent nasal SFT. The other possibilities included Fungal granulomas, Malignant

tumour of nasal cavity, Wegeners granulomatosis and Lymphoma.

She underwent excision of the mass by a combined approach (nasal endoscopic and external infraorbital incision). Peroperatively we noticed that the portion of the tumour in the orbit was well delineated inferiorly and medially. But it was adherent to tissues superiorly, posteriorly and inferolaterally. It was dissected free from the medial and inferior rectus muscles. The nasal portion of the tumour was excised endoscopically in piecemeal. The remnant of the inferior turbinate and the ascending portion of the middle turbinate were trimmed. Histopathology confirmed SFT. The cellularity of this tumour was high in comparison to the blocks of the mass excised two years and eight months back.

Postoperatively her proptosis improved. CT scan taken three months later showed residual/recurrent tumour in the extraconal compartment, extending into the nasal cavity (see Fig 1b). She was strictly followed up and a surgical clearance was planned for a later date. However, on a routine follow up six months later nasal endoscopy showed soft tissue in the left middle meatus though proptosis had resolved. The patient was advised another surgical clearance but she opted to wait and watch before further intervention.

Figure 1a

Before second surgery. Heterogenous Mass in Nose Extending into Inferomedial Orbit

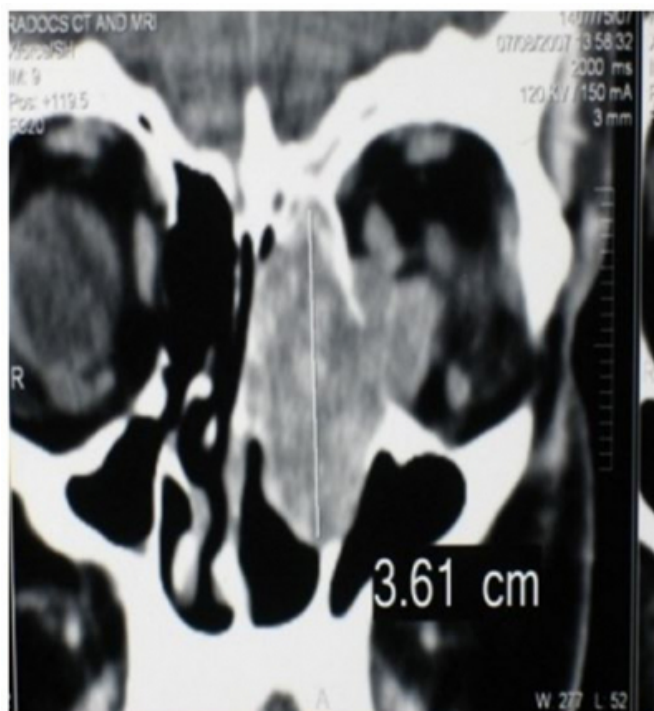


Figure 1b

Small Mass in Inferomedial Orbit three months after second surgery

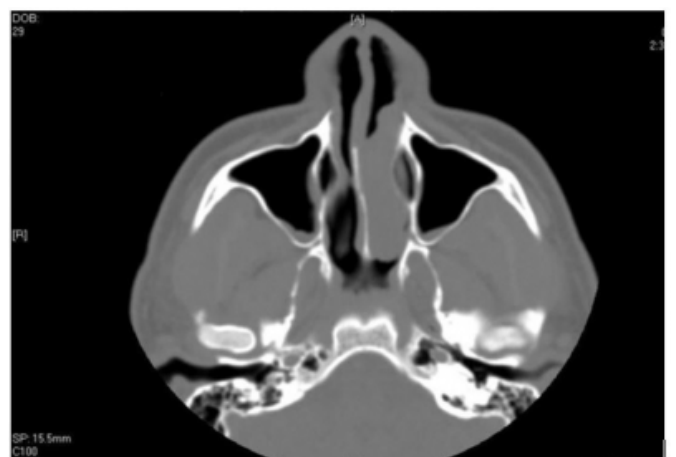


CASE REPORT 2

An otherwise healthy 32 year old gentleman presented with recurrent spontaneous left sided nasal bleed of two weeks duration. Nasal endoscopy revealed a firm, pedunculated, elongated mass with a linear attachment to the lower portion of nasal septum on the same side. Radiological evaluation showed a heterogeneously enhancing mass extending to nasopharynx (see Fig 2).

Figure 2

Mass in Left Nasal Cavity Extending to Nasopharynx

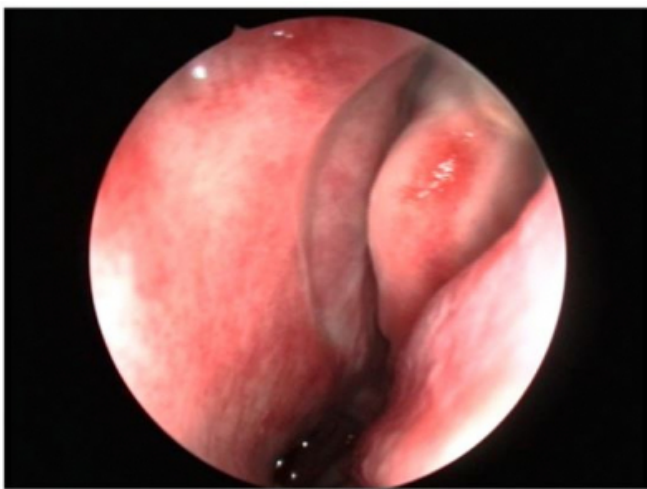


The clinical impression was of a bleeding polypus of the nasal septum or a hemangiopericytoma. He underwent endoscopic excision of the mass. Histopathological examination of the specimen showed a hemangiopericytomatous pattern (see Fig 5). Immunohistochemistry showed strong positivity with CD34, focal positivity with Bcl2 and negative for CD31, Pan CK, S- 100p and CD99 confirming SFT.

Post operative follow ups for the last 2 years have been symptom free and nasal endoscopy showed no recurrence (see Fig 3).

Figure 3

Nasal endoscopy a year later showing no recurrence



CASE REPORT 3

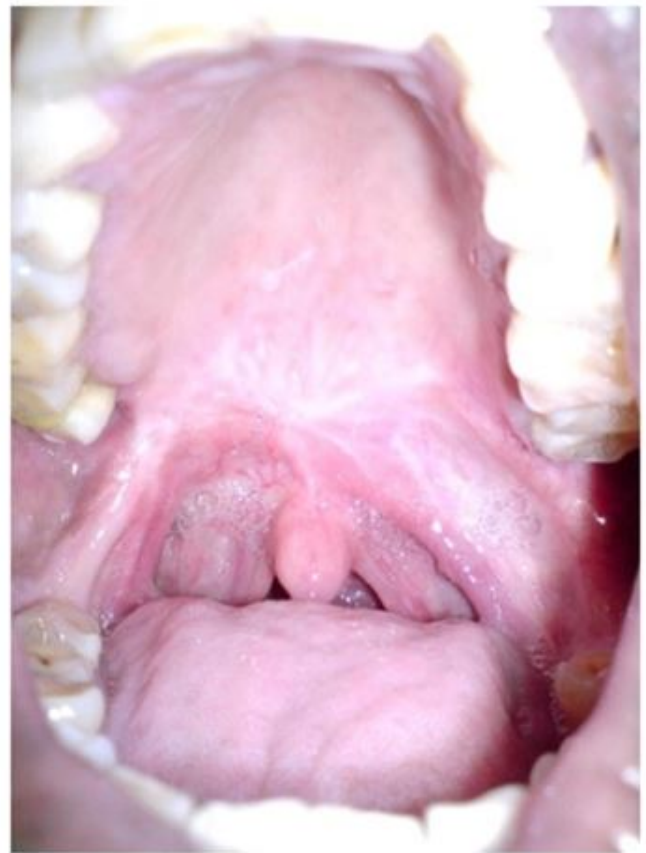
A 40 year old manual labourer came with complaint of difficulty in swallowing due to a slowly growing mass on the palate that was first noticed eight years back. Clinical examination showed a firm, smooth surfaced, mucosa lined, globular mass filling the oral cavity, about 6 cm in diameter, attached to the left side of the soft palate. The free posterior border of the soft palate and the uvula could be viewed by displacing the mass laterally. Rest of the clinical examination was unremarkable. Contrast enhanced CT scan showed a heterogenously enhancing mass hanging into the oral cavity and oropharynx from the palate with no bony erosion of the palate. A primary diagnosis of minor salivary gland tumour was made and biopsied and reported as inflammatory myofibroblastic tumour.

Excision of the well circumscribed mass was done with maximal preservation of palatal mucosa. The histology was confirmed as SFT following immunohistochemical studies. This gentleman has recovered well and has been

symptom free on routine 6 monthly follow ups (see Fig 4).

Figure 4

12 months following Excision of the palatal mass

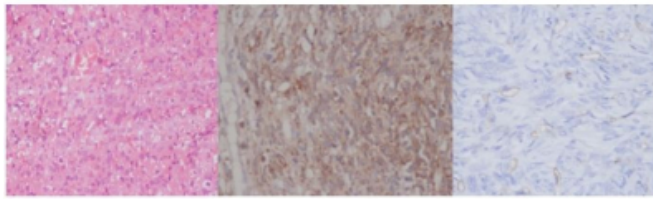


HISTOPATHOLOGICAL FINDINGS:

Representative sections were studied from the three cases. Hematoxylin and Eosin stained sections showed a spindle cell tumor with foci of collagenisation, along with areas where the cells were round to oval. Mitosis were seen, but were sparse. No necrosis was noted. Case 2 showed a hemangiopericytomatous pattern. Immunohistochemistry was done on all three cases and showed strong positivity with CD34 in all. (Biogenex, Prediluted). Focal positivity was noted with Bcl2 (Biogenex, Prediluted). All tumors were uniformly negative for CD31 (Dako, Prediluted), Pan CK (Biogenex, 1:100 diln.), S-100p (Biogenex, Prediluted) and CD99 (Biogenex, 1:100 diln.)

Figure 5

5a: x40 H&E stain - spindle cell tumor with foci 5b: Strong positivity with CD34 (Biogenex, Prediluted) and 5c: Negative for CD31 (Dako, Prediluted)



DISCUSSION

SFT was originally thought to develop exclusively from the pleura. Over the past two decades, an increasing number of extra pleural sites have been reported. These sites include meninges, liver, lung, retro-peritoneum, mediastinum and orbit⁵. In the head and neck region, SFT has been documented in external auditory canal, larynx, thyroid, salivary glands, tongue, parapharyngeal space and infra-temporal fossa. The nose and para-nasal sinuses are rare sites, with around 21 reported cases in global medical literature⁶.

The cause of the tumour is unknown. Besides connection with trauma and a possible component of heredity is under discussion². The exact incidence of SFTs is yet to be determined. This may be due to the fact that the concept of SFTs is evolving and the classification has been changing in the past few years, resulting in difficulties in correct diagnosis. Although still rare, the incidence of SFTs is increasing and it seems to be much more common than previously thought.

Histogenesis of the neoplasm originally thought to be mesothelial in origin, is now considered to be mesenchymal and possibly fibroblastic⁵. The clinical behaviour of SFTs is variable. Mostly they are benign, but local invasion and recurrences have been reported. The predominantly benign nature of extra-pleural SFTs is in contrast with the more aggressive clinical behaviour of pleural tumours which was seen in as many as 23% of such cases⁶. Cases of distant metastasis have been documented in pleural SFTs. About eight cases of recurrence of orbital SFTs have been reported so far; but no cases of metastasis have been documented⁵. Authors have not come across cases of recurrent or metastatic nasal SFTs in journals, though a single case of malignant nasal SFT has been reported⁶.

SFTs are painless and slow growing and can progress to a large size of over 10cm². Rarely some SFTs present with

systemic paraneoplastic syndromes due to secretion of insulin like growth factor. Nevertheless most cases are discovered through an incidental radiologic finding or mass effect².

SFTs are well circumscribed, unencapsulated masses, with nodular surface, often tethered to a pedicle. Cut surface is firm and grayish white in colour⁷. Microscopically spindle cells of various widths are scattered in a collagenous background. Histological arrangements are often described as patternless, storiform, fascicular or herring bone. Strikingly microscopy shows a wide range of morphologic features ranging from fibrous lesions to those with cellular and less fibrous lesions. The fibrous forms are more common than the cellular forms. SFTs display prominent vasculature with branching staghorn vascular channels as a prominent feature, which is similar to vascular channels in hemangiopericytoma⁸. Histological differential diagnosis for SFTs in head and neck include schwannomas, fibrous histiocytomas, fibrosarcomas, meningiomas, nasopharyngeal angiofibromas and hemangiopericytomas⁷.

In addition to histopathological examination, immunohistochemical examination is essential for the correct diagnosis of SFTs. CD34 immunoreactivity was found to be a highly sensitive marker for SFT⁵ (see Fig 3). CD34 is an antigen expressed on the surface on vascular endothelium and hemopoietic progenitor cells. The fibrous forms of SFTs express CD34 and CD99. The cellular forms are less frequently positive for CD34. Bcl2, epithelial membrane antigen and smooth muscle antigens are commonly expressed. Immunohistochemistry helps differentiate SFTs from other soft tissue lesions. CD34 is negative in fibrous histiocytomas and weakly positive in hemangiopericytomas. Schwannomas are CD34 positive. But this is very focal and they are strongly positive for neural markers like S100 protein. Positive staining for CD34, CD99 and Vimentin are common in SFT. Markers for which it is negative are keratin, EMA, S100 protein, carcinoembryonic antigen, desmin, IHHF-35, Factor VIII related antigen and glial fibrillary acidic protein⁹.

The radiological investigations are nonspecific. SFTs illustrate a well defined enhanced mass lesion on CT and MRI. CT specific features include smooth surfaced, well delineated mass, with heterogenous contrast enhancement. Bone remodeling is seen with no frank bone destruction, which shows tumours slow growth pattern and its lack aggressiveness⁵. MRI demonstrates hypointense to isointense signal on T1 weighted images and hypointense

signal with heterogeneity on T2 weighted images with variable gadolinium contrast enhancements. Other spindle cell tumours typically demonstrate hyperintense T2 weighted images. MRI is the most sensitive imaging procedure for excluding invasion of neighbouring structures.

Formerly journals have described malignant extrathoracic SFTs and it usually refers to an atypical histological feature and not to its clinical behaviour. Approximately 10 – 15% SFTs located outside thoracic cavity can recur and metastasize¹⁰. A small percentage of histologically benign cases demonstrated clinically malignant behaviour^{2,5}. Recurrent tumour specimens show higher grade of atypia than the primary tumours. But they retain the same immunohistochemical profiles¹¹ as demonstrated in case 1 of this paper. It has been recognized that SFTs may give rise to metastatic disease or behave aggressively in the absence of any histological signs of malignancy. Thus the relationship between morphology and outcome is poor in SFTs and it is not wise to consider any of the neoplasms as certifiably benign. No reliable guidelines for tumour prognosis have been established².

Presently it has been recognized that a large majority of tumours that were earlier classified as hemangiopericytomas are in fact SFTs. The existence of hemangiopericytomas as a separate entity has been in question for many years. The evolving classification of soft tissue tumours and the changing concept of hemangiopericytomas and SFTs have been recently updated on the basis of the new WHO soft tissue tumour classification². The biggest conceptual change is that SFTs which were regarded as mesothelial in origin are anatomically ubiquitous and can occur very often in somatic soft tissues. The SFTs have a cellular and a fibrous variant. The formerly conventional hemangiopericytomas are cellular variants of SFTs. The conventional SFTs are the fibrous variants of SFTs. Only the hemangiopericytomas of the sinonasal tract seem to fall into two different morphological and prognostic tumour entities. One corresponds to the cellular SFTs and the other larger group has much more in common with the glomus tumours and these might represent true hemangiopericytomas with low malignant potential.

Management of SFTs involves complete surgical excision. The possibility of profuse bleeding during the procedure should be considered even during initial biopsy. Proximity to vital structures in the head and neck region limits exposure and necessitates two or more attempts towards complete and total clearance. Long term follow-up is a must because

recurrences can occur late. The efficacy of radiotherapy and chemotherapy in the treatment of residual or recurrent diseases is unclear. SFTs with atypical histological features like nuclear atypia, areas of increased cellularity, necrosis, four or more mitosis per 10 HPF, tumour size more than 10cm and incomplete resections are positively correlated with local recurrence and metastatic disease. These require stringent follow-up for the first two years and then on a close follow-up for a period of eight years¹¹. However, the absence of the above mentioned atypical histological features does not guarantee a benign clinical course following surgical excision⁶

CONCLUSION

It is now recognized that a large majority of lesions formerly classified as hemangiopericytomas are in fact variants of SFTs. Although still rare in occurrence, SFTs are being increasingly recognized and the clinician should be aware of its presentations. Immunohistochemistry plays a key role in diagnosis. Proper diagnosis of these tumours is essential to avoid confusing diagnosis with a variety of benign and malignant neoplasms like hemangiopericytoma, schwannoma, mesothelioma, fibrosarcoma, synoviosarcoma, leiomyosarcoma and leiomyoma, all based on the location of the tumour. The treatment of choice is conservative surgery. Resectability is the most important prognostic factor and a careful and long term follow up is required, due to lack of availability of reliable guidelines for tumour prognosis.

References

1. Klemperer P, Rabin C.B. Primary Neoplasms of the Pleura - A Report of Five Cases. *Arch Pathol*: 1931;11:385-412
2. Gerd Jurgen Ridder, Gian Kayser, Christian Barna Teszler, Jens Pfeiffer. Solitary Fibrous Tumours in the Head and Neck: New Insights and Implications for Diagnosis and Treatment. *Ann Otol Rhinol Laryngol*: 2007;116(4):265-70
3. Shnayder Y, Greenfield BJ, Oweity T, DeLacure MD. Malignant Solitary Fibrous Tumor of the Tongue. *Am J Otolaryngol*: 2003;24:246-9
4. Cordillo GC, Facciolo F. Localized Solitary Fibrous Tumour of Pleura- An Analysis of 55 Patients. *Ann. Thorac Surg*: 2000;70:1808-1812
5. Subramaniam Krishnakumar MD, Nirmala Subramaniam, Ravindra Mohan, Lakshmi Mahesh, Jyotirmay Biswas, Narsing A Rao. Solitary Fibrous Tumour of Orbit: A Clinicopathologic Study of Six Cases with Review of Literature. *Survey of Ophthalmol*: 2003;48 No. 5:544-554
6. Daniel M. Zeitler, M.D., Seth J. Kanowitz, M.D., and Gady Har-El, M.D. Malignant Solitary Fibrous Tumour of the Nasal Cavity. *Skull Base*: July 2007; 17(4): 239-246
7. Witkin G.B, Rosai J. Solitary Fibrous Tumour of the Upper Respiratory Tract - A Report of 6 Cases. *Am J Surg Pathol*: 1999;15:842-848
8. Mentzel T, Bainbridge T.C, Katern Kamp D. Solitary Fibrous Tumour – Clinicopathological, Immunohistochemical and Ultrastructural Analysis of 12

Cases Arising in Soft Tissues, Nasal Cavity and Nasopharynx, Urinary Bladder and Prostate. Virchows Arch: 1997;430:445-453

9. Peraz-Ordonez B, Kautlas I.G, Strich E, Glibert R.W, Jordan R.C. Solitary Fibrous Tumour of Oral Cavity – An Uncommon Location for a Ubiquitous Neoplasm. Oral Surg Oral Med Oral Pathol Oral Radiol Endod: 1999;87:589-593

10. E Romer M, Bode B, Schuknecht B, Schimid S,

Holzmann D. Solitary Fibrous Tumour of Orbit – 2 Cases and Review of Literature. Eur Arch Otolaryngol: 2005;262:81-85

11. Vallat Decouvelaere A.V, Dry S.M, Fletcher C. Atypical Malignant Solitary Fibrous Tumour in Extrathoracic Locations – Evidence of their Comparability to Intra-thoracic Tumours. Am J Surg Pathol: 1998;22:1501-1511

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