

Solitary Fibrous Tumour: An Unusual Nasal Cavity Tumour

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Abstract

Objective: To describe the diagnosis and management of an unusual case of solitary fibrous tumor of nasal cavity.

Methods: The case records of a patient with solitary fibrous tumor of nasal cavity were reviewed.

Results: This patient presented with constant nasal obstruction of one and a half years duration. Nasal endoscopy revealed a firm polypoidal mass filling the left nasal cavity medial to middle turbinate. After imaging, she underwent endoscopic excision of the mass which on histopathological examination was diagnosed as solitary fibrous tumor. At follow up after one year, there was no recurrence of tumour.

Conclusion: Solitary fibrous tumours are infrequent neoplasms of mesenchymal origin presenting as a well circumscribed, avidly enhancing mass that is iso- to hyperintense on T1W and T2W MRI images. Even though most SFTs are benign, some of these tumors can be locally invasive and have the potential to be malignant. Surgical excision with long term follow up is the treatment of choice.

Keywords: Solitary fibrous tumours; Nasal cavity; Mesenchymal tumour

Introduction

Solitary fibrous tumours (SFTs) are rare neoplasms of mesenchymal origin. They arise mostly from serous membranes and primarily involve the pleura [1]. However SFTs have been described in a wide variety of extrapleural locations. Although about 12%-15% of them occur in the head and neck area, SFT of the nasal cavity and paranasal sinuses are extremely rare. In the absence of definite clinical features, the definitive diagnosis is established by a combination of histologic characteristics, including architectural, cytomorphologic, and immunophenotypic features. Total excision of tumor remains the gold standard of treatment. Even though most SFTs are benign, some of these tumors can be locally invasive and have the potential to be malignant. Considering the slow growing pattern of the tumor, a long term follow up is necessary to rule out local recurrence.

Case Report

49 year old female presented with bilateral hearing loss and continuous nasal block of one and half years duration. There was no history of any itching, pain or discharge from the ears. She complained of mucopurulent nasal discharge and mouth breathing. Examination revealed intact tympanic membrane on both sides. Nasal septum was deviated to the right. There was no lymph node enlargement. Rigid nasal endoscopy revealed a firm polypoidal mass filling the left nasal

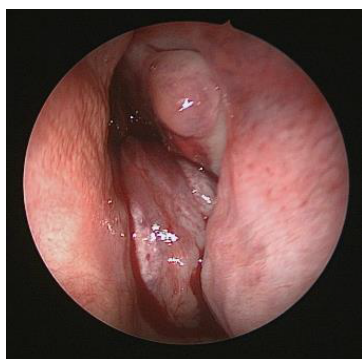


Figure 1: Pink polypoidal mass medial to the middle turbinate in the left nasal cavity.



Figure 2: Computed tomography showing an intensely enhancing polypoidal lesion in the left nasal cavity extending posteriorly into the nasopharynx.

cavity medial to middle turbinate, pushing the septum to the right and filling the right choana (Figure 1). Computed tomography (CT) showed an intensely enhancing polypoidal lesion in the left nasal cavity extending posteriorly into the nasopharynx through choana suggestive of a vascular polyp (Figure 2). Hence an excision biopsy was planned under general anaesthesia. A smooth, pink, firm, fleshy mass was seen in the posterior part of left nasal cavity attached to the posterior most part of left side of septum, anterior face of sphenoid, inferior third of the superior turbinate and just extending into the sphenopalatine foramen. The mass was excised in to by endoscopic approach and sent for biopsy.

Histopathological examination revealed an unencapsulated

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tumour in the submucosa composed of a patternless array of bland spindle shaped cells displaying spindled to ovoid nuclei with uniformly dispersed chromatin, inconspicuous to occasionally visible nucleoli and scant amounts of eosinophilic cytoplasm (Figures 3 and 4) set in a collagenized stroma. Many thin walled branching vessels and occasional thick walled blood vessels with perivascular hyalinisation were also seen. There was no mitotic activity or necrosis. On immunohistochemistry, the tumour cells were focally positive for CD34 and showed weak diffuse nuclear positivity for TLE-1. S-100 protein, SMA and MSA were negative (Figures 5-8). The immunomorphological features were

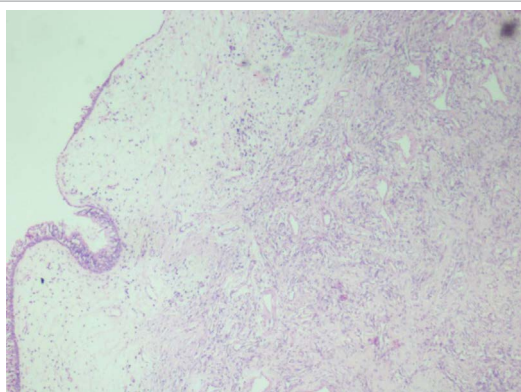


Figure 3: Respiratory mucosa with an unencapsulated tumour in the submucosa composed of cellular proliferation of bland spindle shaped cells and interspersed thin walled vascular channels (H&E stain at 40x magnification).

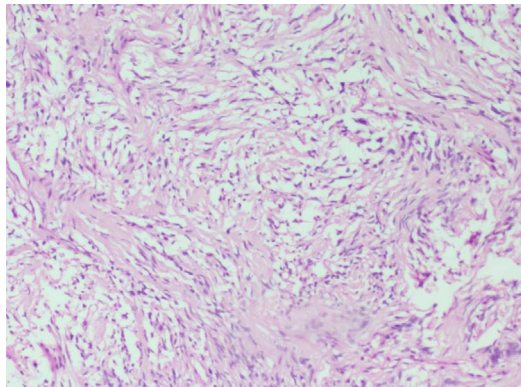


Figure 4: Patternless array of proliferated bland spindle shaped cells set in a collagenous stroma (H&E stain at 100x magnification).

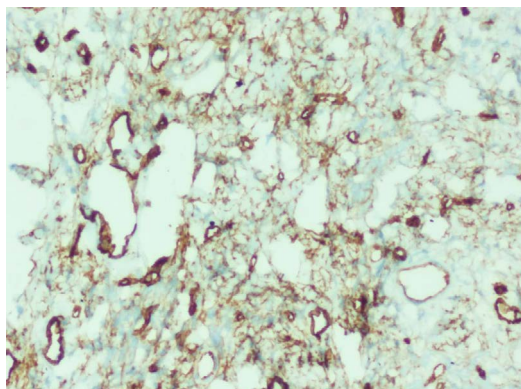


Figure 5: Immunohistochemistry for CD34 stains tumour cells and highlights tumour vasculature (200x magnification).

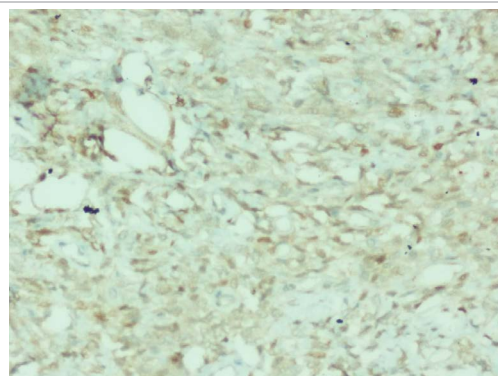


Figure 6: Immunohistochemistry for TLE-1 shows weak diffuse nuclear staining within tumour cells (200x magnification).

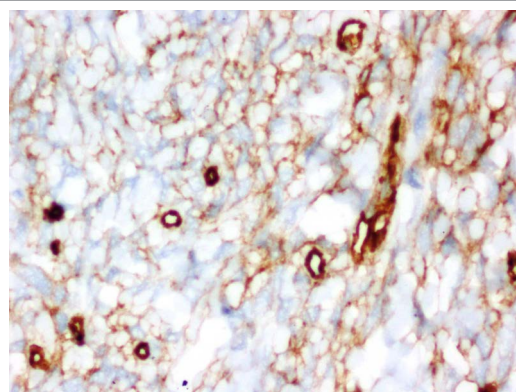


Figure 7: Tumour cells displaying CD 34 immunostain (400x magnification).

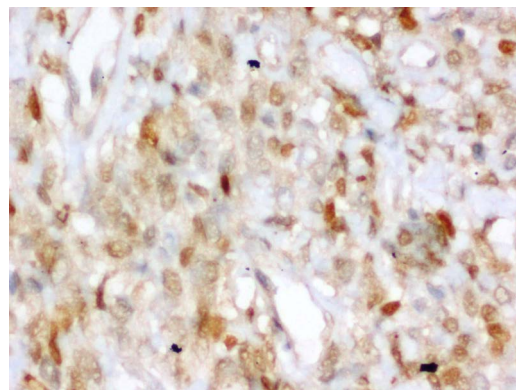


Figure 8: Tumour cells displaying nuclear positivity of weak intensity for TLE1 immunostain (400x magnification).

consistent with solitary fibrous tumour. The patient is on regular follow up. At 1 year post op visit she was asymptomatic and nasal endoscopy did not show any recurrence of tumor.

Discussion

Solitary fibrous tumours (SFTs) are rare spindle cell neoplasms of mesenchymal origin. They were first described in 1931 by Klemperer and Rabin [2]. According to the fourth edition of the WHO Classification of Tumours of Soft Tissue, they are included under the fibroblastic/myofibroblastic tumours of intermediate (rarely metastasizing) variety [3]. Solitary fibrous tumours represent less than 2% of all soft tissue tumours. 12-15% SFTs occur in the head and neck and includes most

of the lesions formerly classified as haemangiopericytoma in this region. These are mostly commonly encountered as pleural or serosal tumours. However they have also been described in other regions such as: the urogenital system, mediastinal space, lungs, vulva, orbit, thyroid, nasopharyngeal region, larynx, salivary glands, parapharyngeal space, tongue and infratemporal fossa [4]. Paranasal sinuses and nasal cavity are very rare sites of origin of SFT.

SFTs tend to occur in adults aged 20-70 [3]. They show equal incidence in males and females [5]. These tumors are very rare in children. The presenting symptoms depend on the site of involvement. Initially nasal SFT presents as an asymptomatic, slowly enlarging, well circumscribed soft tissue mass. Later due to the tumour growth, the patient develops symptoms like nasal obstruction, rhinorrhoea, epistaxis, hyposmia, sinusitis, headache and facial pain. Larger tumours can cause resorption of the surrounding bony structures and extend to the orbit causing exophthalmos or can occupy pterygomaxillary and infratemporal regions and even grow intracranially through the cribriform plate and ethmoid roof [1]. Two paraneoplastic syndromes have rarely been reported with the lesion. They are hypoglycemia due to production of an insulin-like growth factor (Doege-Potter syndrome) and hypertrophic osteoarthropathy [3].

Clinically, SFT is seen as pinkish, reddish or white, circular or oval, well encapsulated fibrous mass with rich vascularisation [6]. On CT imaging, soft tissue SFTs are seen as well-defined hyperdense to isodense, intensely enhancing masses. Almost 10% cases may have associated calcification. Larger lesions may show haemorrhage, central necrosis and associated bone erosion [3]. On MRI, SFTs can be isointense or slightly hyperintense on T1W and T2W images. The mass shows intense post-contrast enhancement [7]. Enhancement may be homogenous, heterogeneous or patchy with often intralesional cystic areas and cystic degeneration. SFTs do not typically show restricted diffusion. SFT shows increased metabolic activity on positron emission tomography (PET). Therefore, PET can be useful in ruling out multiple lesions.

On histology a typical SFT will show a cellular proliferation of bland spindle shaped cells in a patternless array. The background stroma shows thick ropey collagen and thin walled, branching vascular channels [8]. Histologic mimics of SFT such as sinonasal haemangiopericytoma, monophasic synovial sarcoma, benign nerve sheath tumours, smooth muscle tumours and fibrohistiocytic tumours can be differentiated with the help of a panel of immunohistochemical markers. Positive immunostaining for CD34 usually favours a diagnosis of SFT coupled with absence of staining for S-100 protein, SMA, Desmin and Actin [9]. Absence of immunostain for a myoid marker such as SMA and characteristic histological features usually helps distinguish this tumour from a close differential diagnosis of sinonasal haemangiopericytoma. Although weak TLE immunopositivity was seen in the present case, CD34 immunopositivity in the tumour cells excluded a diagnosis of synovial sarcoma. SFTs especially the extrapleural lesions typically

follow a benign clinical course. However pleural SFTs may be malignant in 13%-23% cases [3]. Features like the presence of necrosis, 4 or more mitotic figures per 10 high power microscopic fields, increased cellularity and cellular polymorphism increases the malignancy potential. Tumor size more than 10 cm, presence of haemorrhage, infiltrative margins, and presence of anaplastic foci also point to malignancy [3]. For SFTs, the treatment of choice is total surgical excision with clear margins. In around 6% cases, local recurrence can occur after complete resection. In malignant disease, 23% cases may present with a recurrence. Some lesions may show metastasis several years after an apparently total removal [3]. Common sites of metastases are lung, followed by the bone and liver. Since 10%-15% tend to behave in a locally aggressive fashion long-term follow-up is mandatory for all cases of SFT. Radiotherapy can be considered as a treatment option if tumor displays evidence of malignancy and if the surgical margins are positive [10].

Conclusion

In summary, solitary fibrous tumours are infrequent neoplasms of mesenchymal origin presenting as a well circumscribed, avidly enhancing mass that is iso- to hyperintense on T1W and T2W MRI images. Even though most SFTs are benign, some of these tumors can be locally invasive and have the potential to be malignant. Surgical excision with long term follow up is the treatment of choice.

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