Solitary Fibrous Tumor of Parapharyngeal Space: A Rare Case Report

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Abstract

Solitary fibrous tumors are rare vascular tumours, earlier referred to as hemangiopericytoma. Their occurrence in parapharyngeal space is very rare. Hence, they should also be considered in the differential diagnosis of parapharyngeal soft tissue tumours. This case is being reported to bring out an extremely rare vascular tumour at rare site and represents a surgical challenge because of difficult access in parapharyngeal space with difficult planes between tumour and rest of parapharyngeal space, approach to it is also difficult

Key words

Solitary fibrous tumor, Parapharyngeal space, Vascular tumor, Spindle cell tumor, Embolisation

Introduction

Solitary fibrous tumors (SFT) are rare spindle cell neoplasms [1]. SFTs are generally associated with serosal surfaces, but in recent years it has been reported in extra pleural sites such as the liver, adrenal gland. SFT has an incidence of 2.8/100000 population, with similar rates between men and women, and it is more frequent during the 5–6th decade. Head and neck SFTs accounts for 6% of all SFT in human body [2]. It is often misdiagnosed due to its microscopic resemblance to several other spindle cell tumors. Their occurrence in parapharyngeal space is extremely rare, difficult site to reach when tumour is reaching up to skull base and far medially. Herein, we present a rare case of SFT located in the left parapharyngeal space that was successfully excised by combined approach. This case proposes that, although rare, SFT should be considered in the differential diagnosis of soft-tissue tumours in the parapharyngeal space.

Case report

25 years old male, presented with progressively increasing painless swelling in left parotid region of three years duration. On examination, a 9x9cm swelling was seen in left parotid region which was non-tender, smooth surface, soft in consistency, not adherent to overlying skin. Oropharyngeal examination revealed a smooth bulge on left side of soft palate. (Fig 1) Rest of ENT sites were normal and no constitutional symptoms. Neck examination did not reveal any palpable nodes.

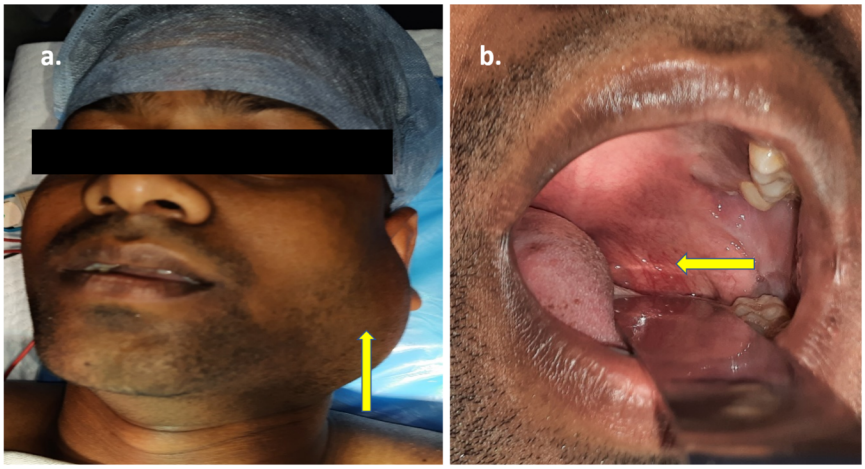

Fig 1. (a) Swelling of size 9x9cm in left parotid region which was non-tender, soft in consistency, smooth surface, (b) Smooth bulge seen over soft palate left side

Patient had reported with CECT scan of neck which showed heterogeneously enhancing lesion 35x44x47mm in size epicentered in left parapharyngeal space, pushing the parotid laterally with large number of arteries/ veins seen in and around the lesion. Lesion was in contact with left styloid and carotid space, superiorly abutting base of skull and medially pterygoid plates and sphenoid.

MRI neck showed hyperintense lesion in T1 and T2 of size 48x68x45mm with heterogenous post contrast enhancement. It was anteriorly closely abutting masseter with loss of fat planes, anteromedially involving prestyloid parapharyngeal space and masticator space displacing temporalis, medially eroding pterygoid plates displacing lateral and medial pterygoid muscle and narrowing oropharyngeal airway. It was closely abutting carotid vessels which were narrowed. Superiorly it was reaching base of skull without intracranial extension. Mandibular ramus was thinned out. (Fig 2a, 2b)

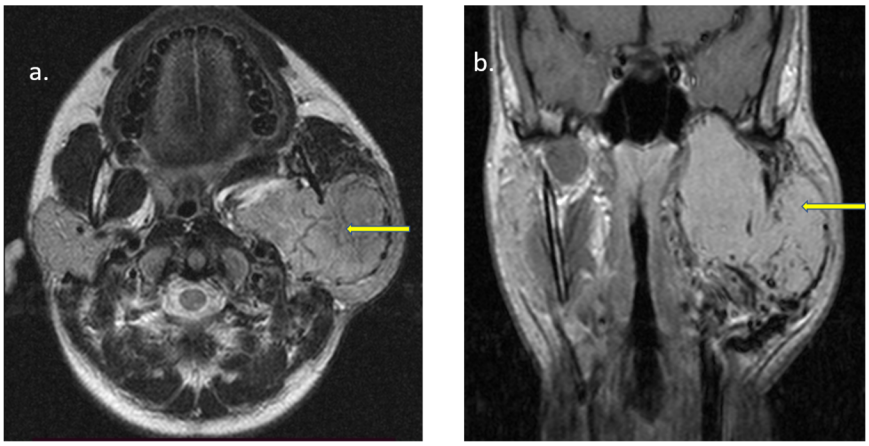

Fig 2. MRI Base of skull to root of neck (a)T2 WI showing a lobulated heterogeneously hyperintense lesion in left parotid gland with internal flow voids suggestive of internal vessels, (b)T1WI post contrast, showing significant post contrast enhancement

CT angiography of neck revealed possibilities of a neoplasm with high vascularity- high flow vascular malformation. It was diagnosed as a case of left parapharyngeal arterio- venous malformation, a plan of excision

of tumor with prior embolization was made. Digital subtraction angiography (DSA) showed hyper vascular tumor pattern in left parotid region with predominant supply from hypertrophied right ECA branch (posterior auricular artery) with minor supply from few twigs of facial artery and no supply from right vertebral artery noted.

Patient underwent pre surgical embolization by contralateral approach-right femoral artery. Particle embolization done with 300- 500 micro and 500-700 micro PVA particles and posterior auricular artery was blocked. Check angiography revealed >90% reduction in tumor blush. (Fig 3a, 3b)

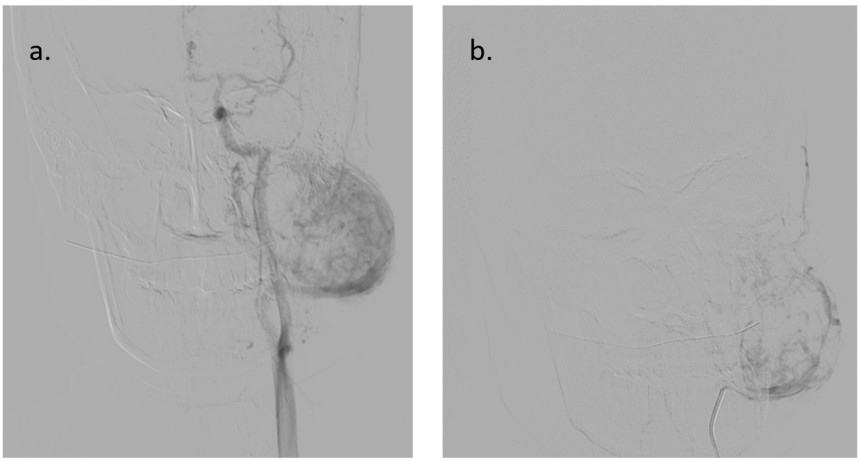

Fig 3. Digital Subtraction Angiography: (a)**Pre embolization:** Coronal- showing feeding vessels in the tumour mass originating from the ECA with a significant tumour blush, (b)**Post embolization:** post coil embolization showing significant reduction in the feeding vessels and the tumour blush

The tumor excision required combined approach (Transparotid, Transcervical and Mandibular swing approach) (Fig 4a, 4b, 4c) with intra op findings of large vascular tumor of size 8.0x7.5cm (Fig 5a, 5b) in left parapharyngeal space infiltrating into deep lobe of parotid laterally, reaching pterygoid plate medially extending up to skull base superiorly and up to angle of mandible inferiorly. Facial nerve was identified and through transparotid approach tumor was reached preserving all the branches of facial nerve. Transcervical and mandible swing were also required for adequate access and excision of the tumor as it was adherent with lot of vascularity. Paramedian mandibulotomy was employed for mandible swing. Brisk bleeding encountered during surgery from dilated tumor vessels.

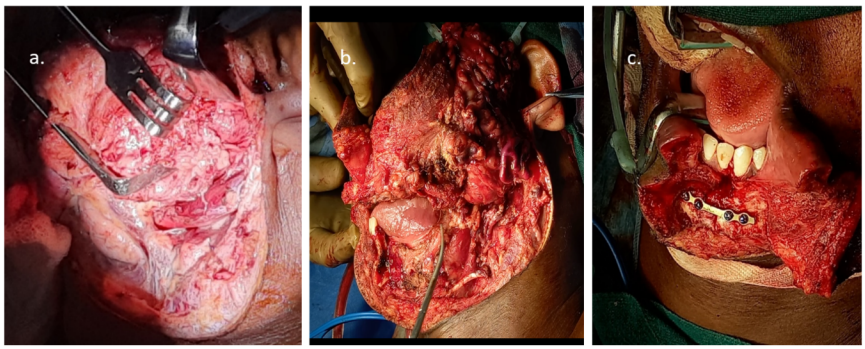

Fig 4. Excision of tumor via combined (a)Transcervical, (b) Trans parotid and (c) Mandibular swing approach

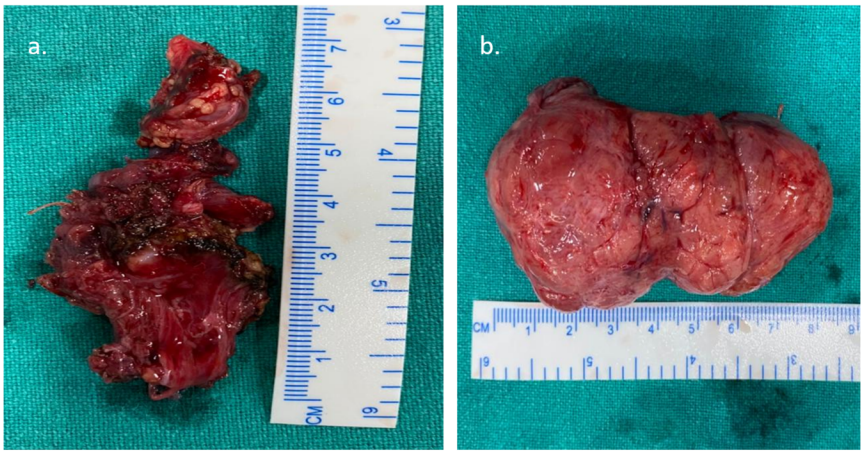

Fig 5a,b . Large vascular tumor of size 8.0x6.5cm in left parapharyngeal space infiltrating into deep lobe of parotid laterally, reaching pterygoid plate medially extending up to skull base superiorly and up to angle of mandible inferiorly

Histopathology of excised specimen revealed a well circumscribed tumor composed of haphazardly arranged spindle shaped cells with attenuated nuclei in a collagenized background with hyper and hypo cellular areas were seen. The stroma showed presence of hemangiopericytomatous pattern of blood vessels. Mitosis was <3/hpf. On IHC, the cells were positive for CD34, negative for SMA, Chromogranin, synaptophysin, CD31, Vimentin and Ki index-2%. Therefore, it was diagnosed as Solitary fibrous tumor. (Fig 6a, 6b)

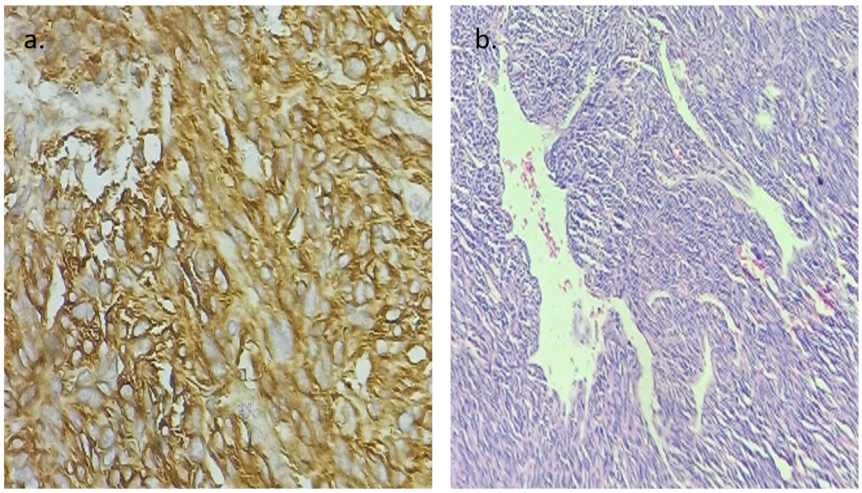

Fig 6. (a)(100X) IHC for CD34- Tumor cells showing membranous immunopositivity, (b)(40X) Photomicrograph showing a spindle cell tumor in a **hemangiopericytomatous** pattern. Blood vessels are dilated and thin walled

Discussion

Solitary fibrous tumors (SFT) were first described in pleura by Lietaud in 1767, followed by Wagner in 1870.[3] Klemperer and Rabin, in 1931, classified pleural tumors into two types: diffuse mesotheliomas and localized mesotheliomas or solitary fibrous tumor [SFT]. [4] In 1991, Witkin and Rosai were the first to report a case in the head and neck region.

SFTs were classified as benign and malignant based on histopathological features. Also been referred to by numerous other names, most of which are now outdated[5]. Subsequent immunohistochemical and ultrastructural studies have suggested that SFTs are most likely derived from adult mesenchymal stem cells rather than mesothelium, 'Solitary fibrous tumor' is the currently preferred term.

Pleural SFTs account for about 30% of all cases followed by meninges of about 27%. The most common extra-pleural and extra meningeal locations include abdominal cavity in 20 %, trunks in 10 %, extremities in 8%, and neck in 5 %. In the head and neck, the most common site affected is the oral cavity [6]. SFT in pleura have a unique gross and microscopic appearance, wherein situation is different when it affects extra pleural sites.

Clinically extra pleural SFT may present with symptoms related to the site of origin of tumor or with systemic symptoms such as hypoglycaemia, arthralgia, osteoarthropathy and clubbing of fingers due to the production of an insulin like growth factor. The systemic symptoms usually resolve on removal of tumors. These tumors usually present as a well-defined, palpable, painless, and slowly growing mass. Extension to the parapharyngeal space to be suspected, if the patient develops compression symptoms. Sleep apnoea is reported, as a result of parapharyngeal extension of the parotid tumour [7]. Radiographic findings are non-specific. Hence, a diagnosis of SFT is based on histological and immunohistochemical findings. On IHC, SFTs are reactive to CD34, Bcl2, vimentin, CD99.

England et al. [8] defined the histological criteria, still valid today, that characterize malignant SFTs (Table 01)

To determine the risk of malignancy of each tumor, Demico in 2017[09] classified SFT into three groups according to the risks of metastasis, which is depending on the malignant potential determined for each tumor. (Table 02)

Follow-up is based on guidelines for soft tissue sarcomas of the National Comprehensive Cancer Network: Low-risk tumours, an imaging test every 6 months for 3 years, and then annually until 5 years, Medium or high-risk tests, every 3–4 months during the first 2 years and then every 6 months until the fifth year.

Surgery in these tumors should be planned with the aim of achieving complete surgical resection with optimal margins as SFT can have a potential malignant behaviour, and to avoid local recurrence.

Parapharyngeal space SFTs are very rare. Hence, they should also be considered in the differential diagnosis of parapharyngeal soft tissue tumors. This case is being reported to bring out an extremely rare vascular tumor at rare site and also represents a surgical challenge because of difficult access in parapharyngeal space with difficult planes between tumor and rest of PPS, approach to PPS is also difficult

Conclusion

Solitary fibrous tumors are rare vascular tumours, earlier referred to as hemangiopericytoma. Surgical removal is the treatment of choice to address cosmesis and functional issues. They need to be removed as they have malignant potential too. Prior embolization may be considered in large and hyper vascular tumours. [10]

Compliance with Ethical standards

Conflict of interest: The authors declare that they have no conflict of interest**.**

Ethical Approval: Not applicable as it involves one case report

Informed Consent: Informed consent was obtained from the individual participating in the study

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