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CASE REPORT

Solitary fibrous tumour of the parapharyngeal space – a case report

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Summary
We present a case of a female patient with solitary fibrous tumour (SFT) originating in the parapharyngeal space which was successfully resected using an open transoral approach. The patient presented with a worsening airway obstruction, obstructive sleep apnoea, and dysphagia for three years. Rhinolaryngoscopy revealed a right nasopharynx and oropharynx lateral pharyngeal wall submucosal mass. Histopathological analysis was consistent with an SFT. This uncommon site of a rare tumour often presents diagnostic challenges or leads to overt misdiagnosis. We interrogated the literature in respect of the diagnostic and therapeutic approaches for this rare condition.
Keywords: solitary fibrous tumour, parapharyngeal space, transoral approach

Case report

A 73-year-old female was admitted with a three-year history of dysphagia, shortness of breath, loss of weight, and neck masses. Patient had previously presented to a local clinic from where she was referred. There was no stridor, but symptoms of obstructive sleep apnoea and voice changes were present. She was alert and cooperative with a "hot potato voice". There was no fever, and her vital signs were normal. Bilateral ear examination was normal. Nasal examination revealed bilateral discharge. The throat examination revealed a large submucosal swelling obstructing the entire oropharynx, with the uvula deviated to the left. On bimanual examination of the oropharynx and the neck externally, there was right-sided submandibular mass extension from the oropharynx. Rhinolaryngoscopy showed an obstructed right-sided nasal passage. The scope further showed a right nasopharynx and oropharynx lateral pharyngeal wall submucosal mass, normal supraglottic and glottic regions, with mobile true vocal cords.

A contrast-enhanced computed tomography (CT) scan of the head and neck was performed, showing a heterogeneously enhancing mass measuring 73 x 52 x 82 mm (TV x AP x CC) in the pre-styloid compartment of the parapharyngeal space. There was a severe narrowing of the airway at the nasopharynx, as well as obstruction of the nasal passage posteriorly. Superiorly, the mass abutted the hard palate and posteriorly it extended to the prevertebral soft tissues with no clear plane of separation. The tongue was inferiorly and anteriorly displaced. There was, however, no bone erosion of the adjacent vertebrae. A large mass centred in the nasopharynx and oropharynx, initially thought to be a nasopharyngeal carcinoma. The mass extended inferiorly to the level of the hyoid bone resulting in significant airway obstruction. (Figure 1).

Preoperative evaluation of the patient involved a careful review of the cross-sectional CT imaging which facilitated a better understanding of the size, location, extent, and presence of infiltration, if present, of tumour into the surrounding tissue. It further facilitated comprehension of the anatomic relationship of the tumour to vascular structures, as well as demarcations of clear planes between tumours. The patient was fully examined to ascertain the following: a) amount of tumour that was submucosally visible, b) patient's dentition with respect to possible challenges with placement of oropharyngeal retractors, c) presence of

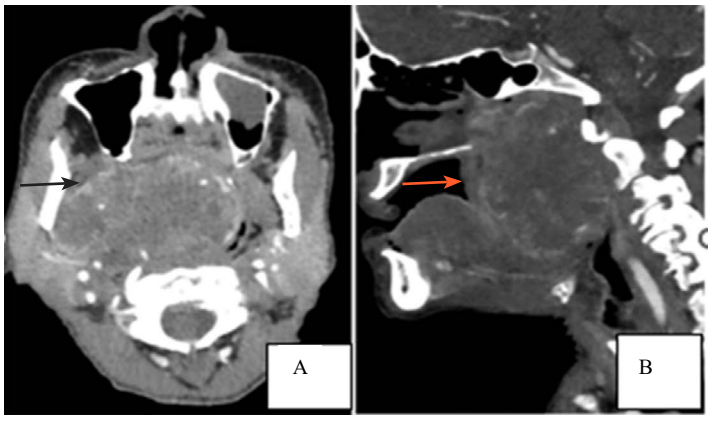

Figure 1: CT scan of the neck (A) axial and (B) sagittal views. A – well circumscribed heterogeneously enhancing mass (thin black arrow) in the right parapharyngeal space at the level of the nasopharynx crossing the midline and compressing the airway; B – thin orange arrow shows posterior extension to the prevertebral space and posterior tongue depression.

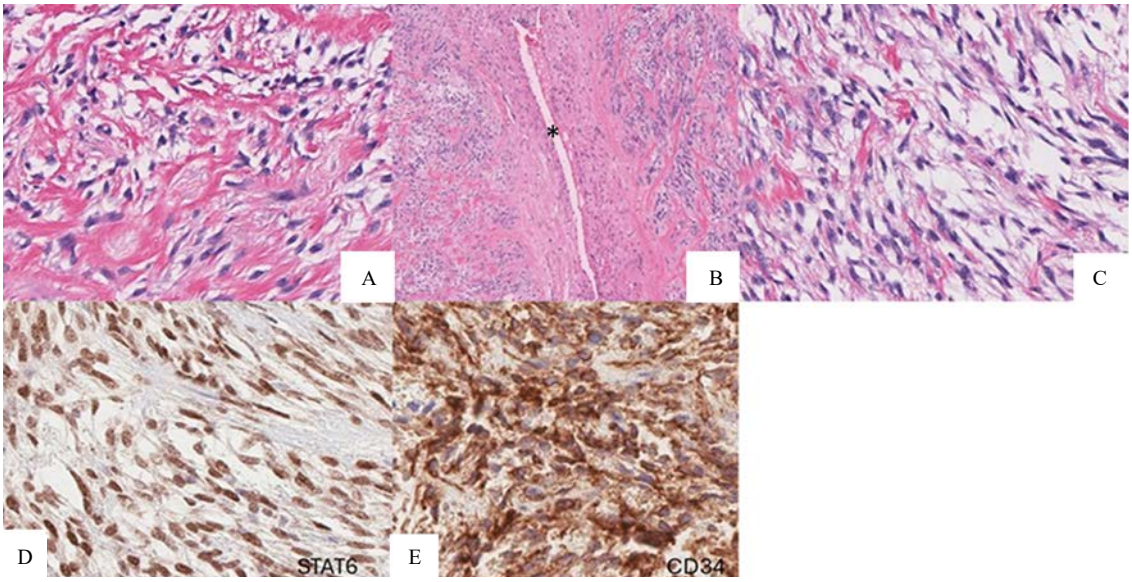

Figure 2: A and C – tumour composed of cells appearing ovoid to spindle-shaped within a predominately hyalinised to myxoid stroma (H&E, x200); B – tumour cells condensing around (*) hemangiopericytomatous vessels (H&E, x100); D and E – tumour cells staining positive with immunohistochemical markers STAT6 (nuclear positivity) and CD34 (cytoplasmic and membranous positivity).

trismus which among other things constitutes difficulty in oropharyngeal exposure, as well as suggestion of possible tumour infiltration into the pterygoid muscles. Mobility of the patient's neck was assessed as limited movement would constitute intraoperative exposure difficulties.

The patient underwent awake fibreoptic nasotracheal intubation with a 5.5 mm endotracheal tube due to severe airway obstruction. A tracheostomy was then performed through a trephine incision, securing the airway with a size 7 tracheostomy tube.

Oral exposure was facilitated using a mouth gag retractor. Headlight illumination was used to enhance visualisation. An open transoral parapharyngeal dissection was carried out. The tumour was palpated prior to incision to guide the extent of the incision required.[1] A vertical curvilinear submucosal incision was made from upper to lower pole of the mass. Subcapsular and blunt finger dissection were performed, leading to the complete excision of the tumour while preserving the neurovascular structures.

Macroscopically, the tumour presented as a lobulated, encapsulated, firm mass with a glistening greyish-white surface. Histopathological examination revealed a well-circumscribed tumour encased by a thin collagenous capsule. The tumour displayed both hypercellular and hypocellular areas, with cells arranged in a haphazard pattern and within poorly defined fascicles. The tumour cells were ovoid to spindle-shaped, featuring plump to attenuated nuclei, small inconspicuous nucleoli, and moderate eosinophilic cytoplasm. The stroma predominantly exhibited hyalinisation, with areas showing myxoid and cystic changes. A hemangiopericytomatous vascular pattern was evident, with tumour cells condensed around these vessels. Mild atypia was noted; however, there was no evidence of pleomorphism, necrosis, or increased mitotic activity. Immunohistochemical analyses showed the tumour to be immune-reactive with STAT6 and CD34 but negative with smooth muscle (Desmin and SMA) and neural markers (S100 and SOX10). The combined morphological and immune profile was consistent with a solitary fibrous tumour (SFT) (Figure 2).

Following progressive recovery, the patient was discharged from hospital nine days postoperatively. As part of the planned surveillance, regular follow-up appointments and serial imaging studies were scheduled to monitor for potential complications or recurrence. Unfortunately, before these surveillance plans could be fully implemented, the patient passed away at home four weeks postoperatively due to unrelated causes.

Discussion

SFTs are rare[2] spindle-cell neoplasms originating from mesenchymal tissue. Typically found in the pleura, SFTs in the head and neck region are uncommon.[3] Accurate diagnosis requires a multifaceted approach due to the tumour's rarity. Head and neck SFTs often arise in the sinonasal tract or oral cavity,[4] with anatomical location influencing clinical behaviour.

Consistent with other reports of head and neck SFTs, our case reveals that SFTs in this region are typically slow-growing, well-circumscribed masses that can occur in any age group.[5,6] Symptoms associated with SFT of the parapharyngeal space include dysphagia, pharyngitis, dyspnoea, voice changes, palpable mass, snoring, and headache.

The exact prevalence of SFT remains unclear, however, evidence from the Surveillance, Epidemiology and End Results Program (SEER) database reveals that 1 134 cases were diagnosed between 2000 and 2019.[3] While SFTs have been reported to occur at equal frequencies among male and female genders and often diagnosed among patients between their fourth and seventh decades of life, no risk factors have been attributed to the SFTs.[7]

Radiological imaging, particularly contrasted CT scans and MRI, is crucial for diagnosing SFTs and assessing their relationship with surrounding head and neck structures. Usually, SFTs are seen as well-defined soft tissue mass with heterogeneously or homogeneously strong enhancement on CT, while on MRI benign SFTs usually have relatively homogeneous low-to-intermediate T1 and T2 signal intensity relative to skeletal muscle. SFTs have variable

appearances, making differential diagnosis challenging. For parapharyngeal SFTs, differentials include schwannoma, paraganglioma, haemangioma, and pleomorphic adenoma.[7] In our patient, contrasted CT scan showed a heterogeneously enhancing lesion with a hypo-dense centre, consistent with existing reports.[6]

SFTs also exhibit unpredictable behaviour, falling into an intermediate category, and cannot reliably be classified as benign or malignant based solely on their morphological characteristics. Histologically, the SFTs are similar to other spindle cell tumours and are variably cellular, a feature that makes differential diagnosis challenging. The most common immunohistochemical (IHC) feature of SFTs is the consistent expression of CD34, although this marker is not exclusively specific to SFTs.[8] Strong nuclear STAT6 IHC staining is a reliable diagnostic marker for SFTs, distinguishing them from other soft tissue tumours. In an evaluation of 231 tumours, including 60 cases of SFT, 59 (98%) of the 60 SFTs showed nuclear expression of STAT6 while all others were negative, except for four tumours that demonstrated weak staining.[8] In this case, histological report showed no pleomorphism or increased mitotic activity. At molecular level, SFT is characterised by NAB2-STAT6 fusion. The majority of the SFTs are benign with only 10–20% associated with local recurrence, distant metastasis, histological malignancies such as infiltrative margins, nuclear atypia, tumour necrosis, as well as high mitotic count.[7]

Management of this patient involved the multidisciplinary collaboration between surgeons, radiologists, pathologists, and oncologists. For a favourable oncological outcome, the treatment of choice is surgical resection with clear margins.[6,9] The complex anatomy of the head and neck region complicates surgical approaches due to surgical access challenges. Furthermore, tumour location and proximity to critical structures of the head and neck can make complete resection difficult. In a review of the National Cancer Database, of 135 head and neck SFTs, the most common site was sinonasal (33.1%), followed by orbital (25.9%).[10] The study revealed that patients with SFTs located in the skull base, as well as those with government-funded insurance, experienced significantly poorer overall survival rates.[10]

Several surgical approaches have been reported in the literature–: transoral open, transoral robotic surgery, endoscope-assisted transoral, and transcervical combined with the transoral approach.[1,5,11] In this patient, we achieved complete resection using the transoral open approach. A potential drawback of the transoral approach is the risk of tumour capsule disruption as opposed to the transcervical approaches.

SFTs of the parapharyngeal space are rare and often misdiagnosed as schwannoma, paraganglioma, haemangioma, or pleomorphic adenoma. This case highlights the challenges of diagnosing and managing SFTs in this complex region, emphasising the need for a multidisciplinary approach (imaging, histopathology, immunohistochemistry), meticulous preoperative planning, skilled surgical technique, and collaborative care.

Our experience underscores the importance of considering SFT in the differential diagnosis of parapharyngeal space tumours and demonstrates that optimal outcomes can be achieved with a comprehensive and expert approach.

Conflicting of interest

The authors declare no conflict of interest.

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Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.

Written informed consent was obtained from a legally authorised representative for anonymised patient information to be published in this article.

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