

Solitary fibrous tumor of the masticator space – Report of a rare case

ABSTRACT

The solitary fibrous tumor (SFT) is a potentially malignant spindle cell neoplasm of the mesenchymal origin that was originally described as a thoracic lesion originating from the pleural tissue. Recently, numerous extrapleural sites of origin have been described, also affecting the head and neck region. SFTs are benign in most cases, but 10%–15% of extrapleural SFTs show malignant behavior in the form of recurrent or metastatic disease. We present the case of a 25-year-old female who presented with an asymptomatic left-sided facial swelling of over three years. She had a diffuse swelling in the left preauricular region, extending to the temporal region deep to the zygomatic arch. On magnetic resonance imaging, the vascular lobulated mass occupied the masticator space, infratemporal fossa, and parapharyngeal space, eroding the mandible. An ultrasound-guided fine-needle aspiration cytology was suggestive of SFT, positive for signal transducer and activator of transcription 6 and negative for TLE1. After preoperative embolization, the tumor was excised through a midline lip split approach with posterior segmental mandibulectomy and reconstruction with a titanium plate. Histopathological report was consistent with SFT. Due to high-risk features, she was advised adjuvant radiation therapy. SFTs of the head and neck are exceedingly rare and those with aggressive behavior even more so. To our knowledge, this is the only case of SFT arising in the masticator space. Diagnosis is often difficult and not definitive without immunohistochemistry. In most cases, complete surgical excision is the only treatment necessary. Regardless, all patients require close clinical follow-up for several years.

Keywords: Extrapleural solitary fibrous tumor, head-and-neck soft tissue tumors, malignant solitary fibrous tumor, tumors of masticator space

INTRODUCTION

Solitary fibrous tumors (SFTs) are tumors of intermediate biological potential with a low risk of metastasis and a relatively indolent course, as per the 2002 WHO classification.^[1] They were first described as a distinct entity of submesothelial origin in the pleura (Klemperer and Rabin).^[2] Extrapleural SFTs have been reported at various sites across the body but are unusual in the head and neck, comprising about 6% of all SFTs.^[2] These slow-growing tumors are often difficult to differentiate from other soft tissue tumors. Although considered benign, extrapleural SFTs can exhibit malignant characteristics.

In this case report, we discuss the clinical and histological features of SFT of the masticator space, along with the surgical approach and a review of literature.

CASE REPORT

A 25-year-old female presented with left-sided facial swelling for over 3 years. She had intermittent, mild throbbing pain in the swelling and was otherwise asymptomatic. On

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examination, there was a nontender, diffuse, ill-defined swelling measuring 8 cm × 6 cm in the left preauricular region, extending beneath the zygomatic arch to the left temporal region. The swelling was soft, lobulated, noncompressible, and nonpulsatile, with demonstrable cross-fluctuation across the zygomatic arch.

On contrast-enhanced magnetic resonance imaging scan, a T2-hyperintense, homogeneously enhancing lobulated mass was found involving the left masticator space, extending to the infratemporal-temporal fossa and parapharyngeal spaces, with multiple dilated and tortuous vessels within. The vertical ramus of the mandible and the coronoid process were eroded, with extension of the soft tissue on both sides of the sigmoid notch. Left zygomatic arch was scalloped and thinned out [Figure 1]. An ultrasound-guided fine-needle aspiration cytology was suggestive of SFT, positive for signal transducer and activator of transcription 6 (STAT6) and negative for TLE1.

After preoperative embolization of the internal maxillary artery, vascularity of the tumor was significantly reduced. The tumor was excised *in toto* with the posterior segment of the mandible, through a midline lower lip-split approach [Figure 2]. The defect was reconstructed with a titanium reconstruction plate and condylar analog, to prevent postoperative malocclusion and to maintain facial contour [Figure 3]. The patient made an uneventful recovery.

The histopathology was consistent with the diagnosis of SFT, the tumor being STAT6 positive and CD34 negative on immunohistochemistry. In view of increased cellularity, patchy moderate nuclear atypia, larger size of tumor (>5 cm), infiltration of underlying bone, and positive bone resection margin, the multidisciplinary tumor board advised adjuvant radiotherapy.

She was followed up for 1 year after radiotherapy and no recurrence was noted.

DISCUSSION

Klemperer and Rabin first described a SFT as a distinct entity of submesothelial origin arising from the pleura. Although considered to have a mesothelial origin (Stout and Murray),^[2] immunohistochemistry and electron microscopy helped establish SFT's mesenchymal origin, hence widely occurring at extrapleural sites such as the retroperitoneum, proximal extremities, abdominal cavity, and trunk in one quarter of cases. SFTs of the head and neck comprise approximately one-quarter of these extrathoracic SFTs, first reported in 1991.^[2,3]

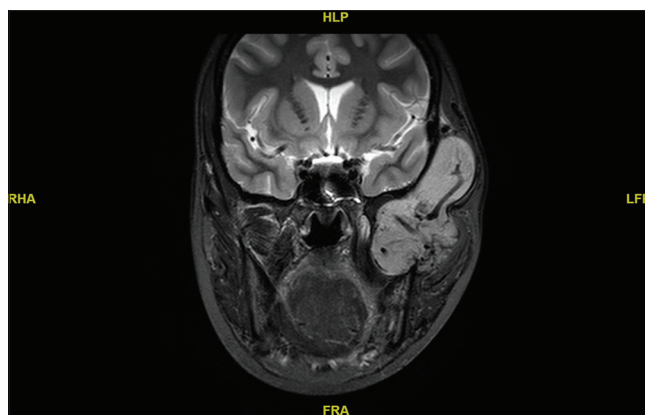


Figure 1: Preoperative magnetic resonance imaging showing the extent of the tumor

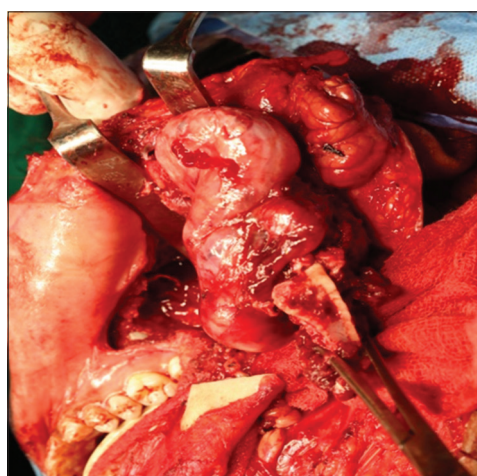


Figure 2: Intraoperative photograph showing the midline lip-split approach and tumor resection *in toto*

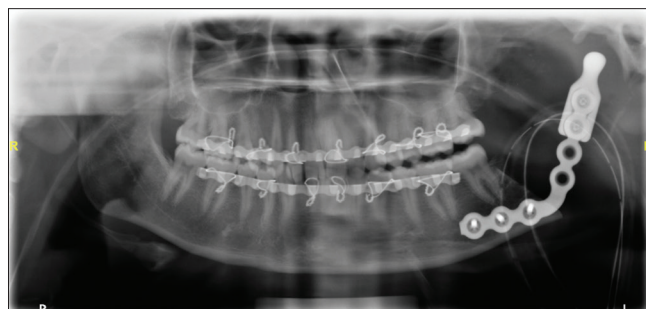


Figure 3: Postoperative orthopantomogram showing the mandibular reconstruction plate and condylar analog

In the head and neck, the most commonly affected site is the oral cavity, most often in the buccal mucosa, followed by tongue and lower lip. Other documented sites of the tumor are the orbit, nose and paranasal sinuses, parapharyngeal space, infratemporal space, larynx, major salivary glands, and thyroid gland.^[4] SFTs in the head and neck present early due to a visible mass or local symptoms, hence more likely detected at a smaller size (<10 cm).^[2] Often confused with other soft tissue tumors,

their behavior is notoriously unpredictable; 10%–15% exhibit malignant potential.^[5]

Definitive diagnosis always requires radiographic and histopathological adjuncts. The presence of regressive remodeling of the adjacent bone due to long-standing pressure of the slow-growing tumor is suggestive of SFT on radiography.^[2] Recent studies have identified inv12 (q13q13) to be a recurrent genetic mutation in SFTs, resulting in NAB2–STAT6 gene fusion, driving STAT6 nuclear expression, a highly sensitive and specific immunohistochemical marker of SFT, among others (positive for CD34, CD99, bcl2, and STAT6 and negative for EMA and S100).^[6]

Demicco *et al.*^[1] proposed a risk stratification model for metastasis from SFT, using age (<55 years vs. >55), tumor size <5, 5 to <10, 10 to <15, or >15 cm), and mitotic figures (0, 1–3, or >4). Total scores are tabulated to determine the risk of aggressive disease. The strongest predictors of time to metastasis and disease-specific death, according to this model, were the age at presentation, tumor size, and mitotic index. Bishop *et al.*^[7] recommended that adjuvant radiation therapy for malignant SFTs of >5 cm and for inadequate surgical margins.

CONCLUSION

SFTs of the head and neck are exceedingly rare and those with aggressive behavior even more so. To our knowledge, this is the only case of SFT arising in the masticator space. Diagnosis is often difficult and not definitive until immunohistochemical evaluation is performed. In most cases, complete surgical excision is the only treatment necessary. Regardless, all patients require close clinical follow-up for several years.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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