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# **Case Report**

# Malignant solitary fibrous tumor of the floor of the mouth in a 71-year-old male: A case report $\stackrel{\star}{\sim}$

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#### ABSTRACT

Solitary fibrous tumors are rare mesenchymal tumors originally described in the pleura that infrequently metastasize. We present a 71-year-old male complaining of hemoptysis and a mass with the characteristic appearance of a hemangioma in the floor of the mouth. The mass had nonspecific imaging features on CT and MRI. After unsuccessful fine needle aspiration, surgical excision and biopsy with histological analysis revealed a solitary fibrous tumor, high risk variant. CT Imaging and lymph node biopsy showed gross total resection and no metastatic adenopathy. Given the high risk for malignancy, the patient received adjuvant radiation without subsequent clinical or imaging signs of recurrence. This case report demonstrates the presentation of this rare entity that can often be confused with other tumors in this region, given its nonspecific clinical and imaging findings.

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## Introduction

Solitary fibrous tumors (SFT) are rare mesenchymal tumors, of which only 10% occur in the oral cavity [1]. The differential diagnosis of SFT is broad, and its MRI findings are variable and nonspecific. Tissue diagnosis is required, with immunohistochemical stains commonly used to aid in diagnosis. Surgical resection is the most common method of treatment, with adjuvant therapy reserved for unresectable, recurrent, or malignant variants [2].

## **Case report**

A 71-year-old male with past medical history of hypertension, hyperlipidemia, diabetes mellitus, coronary artery disease with coronary artery bypass graft 5 years ago, atrial fibrillation treated with rivaroxaban (Xarelto), intermittent complete heart block, prostate cancer (radiation therapy 14 years prior), and gout presented for intermittent hemoptysis over the preceding years. His vital signs and labs were largely normal, including a hematocrit of 41% (normal: 41-50) and MCV of

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Fig. 1 – A blue-purple soft tissue mass along the right side of the floor of the mouth (black arrow).

80 fl (normal: 80-100), but his bicarbonate was 19 mEq/L (normal: 23-30). On exam, he was found to have a large, erythematous, soft, nontender floor of the mouth lesion on the right with intact overlying mucosa. On clinical inspection, the lesion was bulging with a blue-purple discoloration, typical of a hemangioma (Fig. 1). Oozing was noted on minor manipulation of the lesion.

CT of the neck with intravenous contrast showed a large, heterogeneously enhancing soft tissue mass in the right sublingual space extending posteriorly and inferiorly to the submandibular space. The lesion displaced the genioglossus muscle to the left (Fig. 2). MRI of the neck demonstrated the lesion to be isointense to muscle on T1-weighted images, hyperintense on T2-weighted images, and heterogeneous enhancement on postcontrast images (Fig. 3). On contrast-enhanced MRA using time-resolved imaging, the lesion demonstrated heterogenous late arterial enhancement with neovascularity and without early dilated draining veins (Fig. 4).

Prior to surgical excision of the mass, embolization of his right facial artery was performed. Subsequently, the mass was accessed by way of the platysma superficially, the right sternocleidomastoid laterally and the right anterior belly of the digastric medially. The right submandibular gland was resected and sent for histologic analysis, and the facial vein and facial artery were ligated. The hypoglossal and lingual nerves were preserved. Grossly involved mucosa were circumscribed around the mass and sent, along with the mass, for biopsy. The mucosal defect was closed primarily.

Postoperative CT demonstrated total gross resection (Fig. 5).

The excisional biopsy showed a mass that measured  $10.1 \times 5.0 \times 3.9$  cm. On immunohistochemical analysis, the mass was positive for CD34 (diffuse), STAT6 (patchy) (Fig. 6), and SMA (focally), but negative for AE1:AE3, desmin, CD31, and EMA. The mass showed up to 7 mitoses per 10 high powered fields, increased cellularity, focal cellular pleomorphism, stroma hyalinization, and focal necrosis; however, no vascular invasion was identified. Based on these findings, the mass was diagnosed as an SFT. The tumor was also described as high risk on the bases of age greater than 55, size larger than 5 cm, and a mitotic index of more than 4 per 10 high powered fields. No metastatic adenopathy was identified.

Due to having the high-risk variant of SFT it was recommended that the patient undergo adjuvant radiation therapy with a total dose of 70 Gy in 35 fractions. Follow up 18fludeoxyglucose positron emission tomography/CT showed

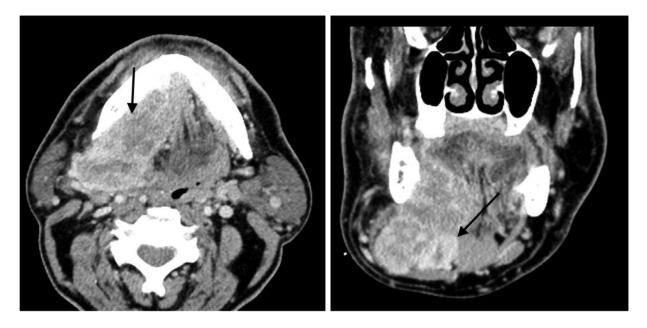


Fig. 2 – Axial and coronal postcontrast CT neck soft tissue. There is a heterogeneously enhancing soft tissue mass along the floor of the mouth on the right (black arrows).

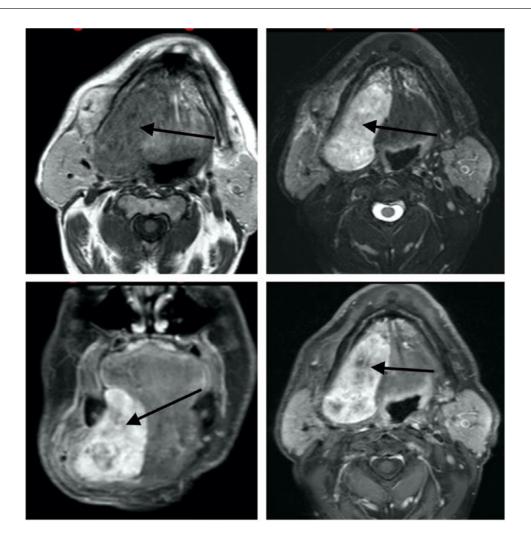


Fig. 3 – MRI neck soft tissue series, clockwise from top left: axial precontrast T1 without fat saturation, axial T2 with fat saturation, axial postcontrast T1 with fat saturation, coronal postcontrast T1 with fat saturation. The soft tissue mass is well-defined, isointense to muscle on the T1-weighted image, hyperintense on the T2-weighted image, and demonstrates heterogeneous enhancement on postcontrast images (black arrows).

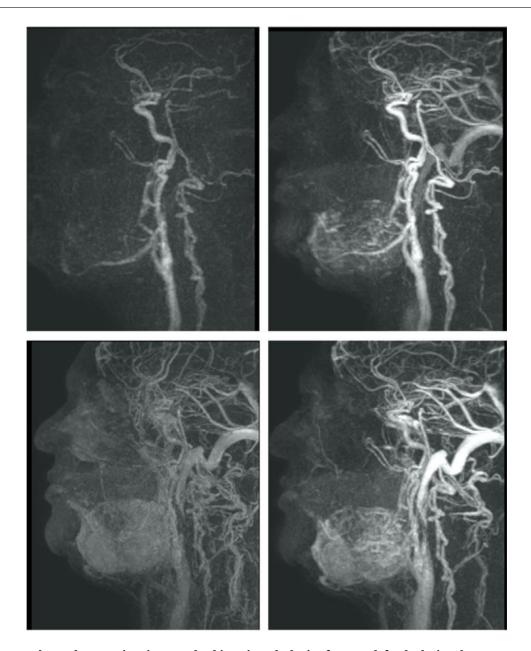
no focally intense activity to suggest residual disease. The patient is currently being monitored clinically with regular follow-up. At his last visit 14 months postoperatively, he denied any new complaints.

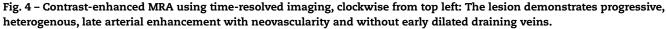
## Discussion

SFT are mesenchymal tumors first described in the pleura and lungs in 1931 [3]. Extrathoracic cases of SFT were not identified until 60 years later [4] and have since been found in the mediastinum, peritoneum, spinal cord, soft tissue, and head and neck [1]. They are classified as fibroblastic neoplasms with intermediate (rarely metastasizing) behavior by the World Health Organization (WHO) [5]. They are further risk stratified into high-risk, intermediate-risk, or low-risk for metastasis, on the basis of patient age, tumor size, mitoses, and necrosis criteria [6]. Only 10% of all SFT are found in the head and neck [1]. One study of 150 cases of SFT of the oral cavity by Nunes et al. found that they are slightly more common in females (56%), predominantly benign (90.1%), usually under 2.6 cm (63%), and that the mean age is 49.4. The most common sites were the buccal mucosa (46%), the tongue (15%), and the palate (7%). However, the tongue was found to be the most common site of malignant SFT of the oral cavity [2].

Oral cavity SFT are often described as solitary, submucosal, well circumscribed nodules, measuring 0.1 to 9.3 cm, firm to rubbery, and asymptomatic. On gross examination, they are often identified as a whitish nodular mass with a smooth, firm surface [2].

On CT, SFT typically appear as isodense, hypodense, or a combination of both, as compared to adjacent musculature [7]. On MRI, SFT classically display low signal intensity on T1-weighted images and low or mixed-high signal intensity on T2-weighted images. In this case, the SFT exhibited isointense signal intensity on T1 and high signal intensity on T2, not





completely characteristic of SFT. Post contrast T1-weighted images of SFT most commonly demonstrate inhomogeneous enhancement, especially for tumors greater than 10 cm [8], as in our case. On contrast-enhanced MRA using time-resolved imaging, neoplastic lesions classically demonstrate heterogenous late arterial and/or venous enhancement with neovascularity. By contrast, hemangiomas classically demonstrate homogenous early arterial enhancement without neovascularity. Dilated early draining veins are additional findings classically seen with arteriovenous malformation [9].

However, the imaging characteristics of SFT in the floor of the mouth are nonspecific. Therefore, other lesions in this region should be considered; these include squamous cell carcinoma, minor salivary gland tumor (adenoid-cystic carcinoma and mucoepidermoid carcinoma), lymphoma, vascular malformation, myofibroma, neurogenic tumor, sarcoma [1], spindle cell tumor, and liposarcoma [5].

SFT require a tissue diagnosis and often additional immunohistochemical analysis. According to the study by Nunes et al., the histology of SFT of the oral cavity includes spindle (83.1%) and ovoid (27.2%) cells. Additionally, malignant cases demonstrated greater than 4 mitoses per 10 high powered fields 79% of the time and areas of necrosis 21% of the time; perivascular hyalinization was sometimes found as well. Immunohistochemically, among benign cases, 99% were positive for CD34, 93% for BCL2, and 71.4% for CD99. Smooth muscle actin, s100, desmin, cytokeratin are commonly absent [2]. Recently, the NAB2/STAT6 fusion gene was found to be a com-

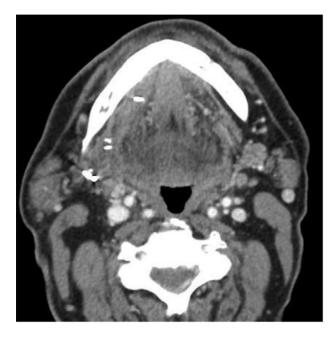


Fig. 5 – Postoperative, postcontrast, axial CT Neck soft tissue. There is total gross resection of the soft tissue mass, without evidence of recurrence.

apy, with pazopanib being the preferred first line agent [11]. According to Nunes et al. [2], 73% were disease free after their last follow up, and the mean time to follow up was 24.7 months.

### Conclusion

We present a 71-year-old male with a malignant SFT located in the floor of the mouth and describe the clinical, imaging, and pathological characteristics of his case. The differential diagnosis of SFT is broad, and its imaging findings are variable and nonspecific. In our case, clinically, the lesion had the characteristic appearance of a hemangioma. This case report demonstrates the presentation of this rare entity that can often be confused with other tumors, given its nonspecific clinical and imaging findings.

#### **Patient consent**

Written informed consent for publication was provided by the discussed patient.

mon finding in almost all SFTs [10], and has since become widely adopted as a reliable diagnostic marker [6].

Surgical resection is the most common method of treatment, and radiation therapy is reserved for malignant [2], unresectable or recurrent cases [1]. Antiangiogenic compounds have been shown to be more effective for SFT than chemother-

### Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2022.09.016.

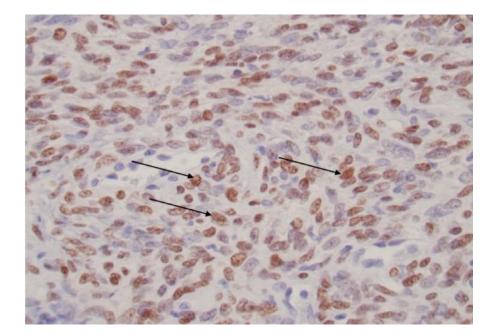


Fig. 6 – STAT6 immunohistochemical stain demonstrating nuclear distribution, a diagnostic feature of Solitary Fibrous Tumor (black arrows).

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