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CASE REPORT

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Solitary Fibrous Tumor of the Masticator Space with Unusual Extension: a Case Report

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ABSTRACT

Background: *We present the case of a patient with solitary fibrous tumor of the masticator space with unusual extension.*

Case presentation: *A 43-year-old woman presented with a painless mass with intraoral extension on the right cheek. The B-scan sonograph and magnetic resonance imaging revealed the extension of the tumor. The biopsy performed under local anesthesia raised the suspicion of a solitary fibrous tumor. Tumor excision included a preoperative tumor embolization. The surgical removal of the tumor included a partial parotidectomy on the right side, insertion of masseteric and temporalis muscle, resection of the middle part of the zygomatic bone and stabilization of the bone with a plate, mobilization of the tumor from the maxillary sinus and the pterygopalatine fossa through an endoscopic approach and an approach via partial resection of the anterior wall of the maxillary sinus after identifying and sparing the infraorbital nerve. The histological findings confirmed the diagnosis of solitary fibrous tumor. The patient's treatment completed with radiation therapy, and 2.5 years later, there was recurrence in the right temporal area.*

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Conclusions: *To our knowledge, this is the second reported case of solitary fibrous tumor arising in the masticator space and the only case with extension intraorally and in the paranasal sinuses. Tumor embolization and complete surgical excision are the most frequently recommended treatments.*

Keywords: solitary fibrous tumor, masticator space, mesenchymal tumor.

INTRODUCTION

Solitary fibrous tumor (SFT) is a rare mesenchymal tissue neoplasm that occurs at all anatomic sites, commonly at the pleura. It is estimated that less than 2% of all soft tissue masses are solitary fibrous tumors (SFTs). The extra thoracic detection is rare and the incidence in the head and neck region is exceedingly rare (1-16).

The first detailed histological description of SFTs was reported by Wagner in 1870 (17). Later in 1931, Klemperer and Rabin described SFTs in pleural tumors (18). Although various opinions have been expressed about SFTs and hemangiopericytomas, both tumors are considered separate entities, according to 2002 WHO classification (19).

It is estimated that 25% of extra pleural SFT and 6-18% of all SFTs arise from the head and neck region (20-22). Within this anatomic region, it usually offends the orbit and the oral cavity. Less frequently, it affects the nasal cavity and paranasal sinuses, the salivary gland and larynx (23). Nevertheless, only few cases have been reported to SFT departing from the buccal mucosa (24-27).

These tumors grow slowly, show minimal malignant variants, while metastases appear in tumors outside the head and neck area (28, 29).

The STAT6 nuclear protein and the NAB2–STAT6 fusion gene appear to play an important role in immunohistochemistry and molecular diagnosis. The 2020 WHO classification recommended risk models to determine the prognosis of SFT more clearly (6, 14, 30-32).

We report the case of a solidary fibrous tumor of the buccal space which presents an unusual extension to adjacent tissues. ❑

CASE PRESENTATION

In July 202, a 43-year-old woman presented with inability to open the mouth and abnormal right lateral jaw dislocation for about two months. As the patient had difficulty in eating properly, she previously referred to a local dental infirmary in the Greek island where she lives. Following the dentist's medical instructions, she received muscle relaxant therapy without permanent effect.

During clinical examination, a well-circumscribed, smooth and painless tumor mass was palpated on the right cheek. The mass had intraoral extension (Figure 1). The patient also presented trismus and lateral right displacement of the lower jaw. On B-scan sonography we detected a well-circumscribed inhomogeneous tumor with high peripheral vascularity and a maximum diameter of 3 cm. Magnetic resonance imaging (MRI) revealed a solid mass into the right masticator space, showing mild high intense signal on T2 sequences and intermediate intense signal on T1 sequences. The mass with a maximum diameter of 4.1 cm was coming out of the right temporalis muscle, causing erosion of the posterior wall of the right maxillary sinus. Under local anesthesia, four pieces of tissue were obtained from the lesion and adjacent structures. The histological analysis raised the suspicion of a cell neoplasm compatible with SFT.

In February 2021, the patient decided to be treated for tumor exclusion. The MRI, which was performed for the evaluation of tumor resectabi-

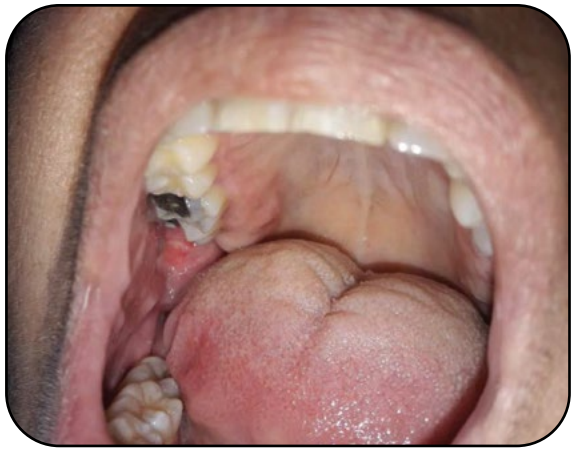

FIGURE 1. Intraoral extension of the tumor

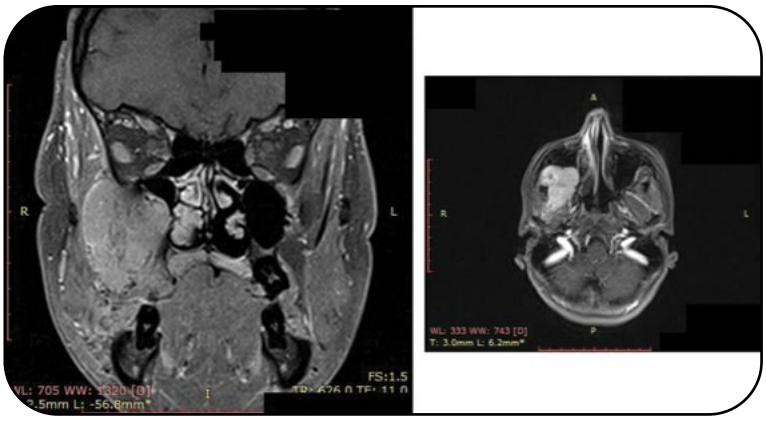

FIGURE 2. Magnetic resonance performed for the evaluation of the resectability of the tumor, showing a vascular mass with maximal diameter 5.7 cm that pushes the posterior wall of the right maxillary sinus and extents to the orbital floor

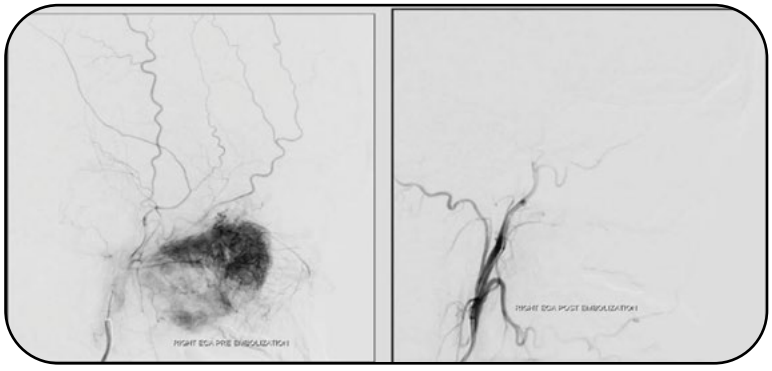

FIGURE 3. Right external carotid artery angiography, pre-embolization and post embolization

lity, revealed a vascular mass with a maximum diameter of 5.7 cm that pushed the posterior wall of the right maxillary sinus and extended to the orbital floor (Figure 2).

Because both imaging tests and histological analysis revealed a vascular tumor, we decided to proceed to a preoperative tumor embolization. Under general anesthesia, the right femoral artery was punctured, a 6 F sheath was placed and a 6 F guide catheter was advanced into the right external carotid artery. Through this, selective catheterization of branches and the internal maxillary artery (transverse facial, inferior alveolar, buccal, sphenopalatine, descending palatine) was performed. They were embolized using 300-900 μm microparticles (Embosphere, Merit Medical). Immediate angiographic control showed the almost complete elimination of the blood supply to the tumor (Figure 3).

Tumor embolization was followed by its surgical removal. Surgery started with a partial parotidectomy on the right, with a more cranially positioned incision, in order to gain better access to the zygomatic, orbital and frontal branches of the facial nerve. Using intraoperative facial nerve monitoring, the above branches were identified and protected. Afterwards, the superficial portion of the parotid gland was resected (Figure 4). The tumor was extending posteriorly up to the right temporalis muscle and the right pterygoid muscle and anteriorly/inferiorly to the alveolar process of the maxilla. The cranial insertion of both the superficial and the deep portion of the masseteric muscle and the inferior insertion of the temporalis muscle have been detached from

FIGURE 4. Partial parotidectomy on the right using intraoperative facial nerve monitoring

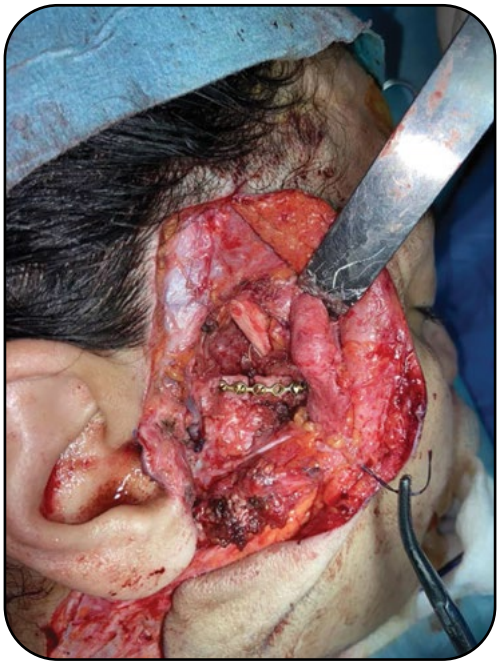

FIGURE 5. Re-stabilization of the middle part of the zygomatic bone with a plate

FIGURE 6. External approach via partial resection of the anterior wall of the maxillary sinus after identifying and sparing the infraorbital nerve

the zygomatic bone as the middle part of the zygomatic bone has been resected. So, the tumor was mobilized from the above region. The middle part of the zygomatic bone was re-stabilized afterwards with a plate (Figure 5). Through an endonasal endoscopic approach and 45-degree-endoscopic visualization, the medial and posterior wall of the maxillary sinus have been resected and the tumor was mobilized from the maxillary sinus and the pterygopalatine fossa, respectively. Due to extensive tumor attachments towards the oral cavity in the area of the right third molar posteriorly and inferiorly, another approach via partial resection of the anterior wall of the maxillary sinus – after identifying and sparing the infraorbital nerve – was needed to fully mobilize and resect the tumor (Figure 6). Complete excision of the tumor and satisfactory cosmetic results were achieved. The specimen was sent for histological analysis (Figure 7). The findings confirmed the diagnosis of SFT. The patient's treatment completed with radiation therapy, with good effect on locoregional recurrence, at two-years follow up. However, 2.5 years later, there was recurrence in the right temporal area, that extended intracranially and pressed the dura mater. The recurrence also extended up to

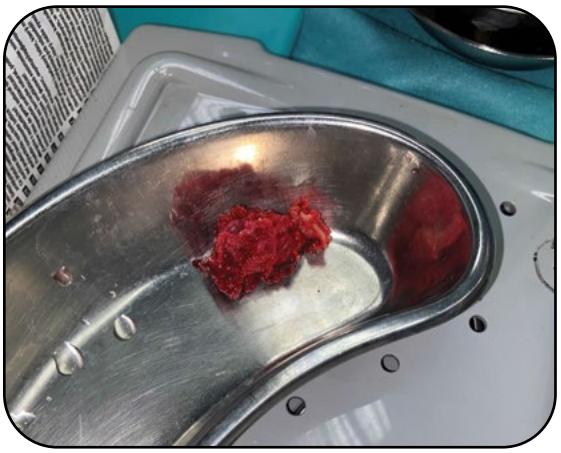

FIGURE 7. The specimen after complete excision of the tumor

the orbital floor, pressing the external rectus muscle. ❑

DISCUSSION

Solitary fibrous tumor is a type of mesenchymal neoplasm that previously termed and diagnosed as hemangiopericytoma, localized mesothelioma, orbital fibrous histiocytoma, giant cell angiofibroma.

Based on the 2002 World Health Organization (WHO) classification system, these tumors have intermediate biological potential (rarely metastasizing). The risk for metastasis was up to 35-45% in series with a longer follow-up, as recommended in the 2020 WHO classification (1, 6, 11, 33).

Luke Stanisce et al, who conducted the most robust and recent review of the literature about SFT of the head and neck, noticed that over 587 of SFTs were found in this area since 1991. Soft tissue tumors appear to have no predilection for gender and mainly occur in fifth and sixth decade of life (34-36).

Soft tissue tumors of the head and neck usually present as painless masses that grow up slowly, often with local compressive symptoms (33). In the head and neck, the most commonly affected areas include the oral cavity, followed by the orbit, nose and paranasal sinuses (37-41). The nasopharynx, larynx, parapharyngeal space, salivary glands and thyroid are less commonly affected areas (42-46). Patil *et al* described the first case of SFT of the masticator space extending to the infratemporal-temporal fossa and parapharyngeal spaces (46). In our case, SFT was arising in the right masticatory coming out the right tem-

poralis muscle. The mass had intraoral extension, eroded the posterior wall of the right maxillary sinus and extended up to the infratemporal fossa.

Regarding differential diagnosis *via* imaging, SFT should be concerned in cases of hypervascular tumors (33). According to Demicco's model age at presentation, tumor size and mitotic index are the strongest predictors of time to metastasis and death (1).

Concerning the genetic signatures that are implicated in the onset of STF, the discovery of fusion genes between NAB2 and STAT6 in 2013 was determinant to expand knowledge on the molecular subtypes involved in SFT (6). More specifically, the fusion gene variants, NAB2 exon 4-STAT6 exon 2/3 and NAB2 exon 5/6/7-STAT6 exon 16/17/18 are identified in meningeal STFs, but all of them could play an important role for future therapies independently of the histological origin of STFs (47). Interestingly, STFs that are derived from the salivary glands and cases that demonstrate ectopic salivary glands are very rare in the intracranial environment. Molecular studies based on sophisticated and novel techniques including next-generation sequencing (NGS)-based assay with targeted RNA fusion sequencing. In one of them, the study group reported a variety of NAB2-STAT6 fusions such as NAB2ex4-STAT6ex2, NAB2ex2-STAT6ex5, NAB2ex6-STAT6ex16, NAB2ex6-STAT6ex17, NAB2ex3-STAT6ex18 and NAB2intron6-STAT6Ex17 (48). ❑

CONCLUSION

Solitary fibrous tumors of the head and neck are extremely rare. To our knowledge, this is the second reported case of SFT arising in the masticator space and the only case with extension intraorally and in the paranasal sinuses. Differential diagnosis is difficult and impossible without immunohistochemical evaluation. Tumor embolization and complete surgical excision are the most frequently recommended treatments. Besides therapeutic approaches, genetic analysis for detecting the specific NAB2/STAT6 fusion subtype should be made for completing the whole diagnostic procedure. ❑

Conflicts of interest: none declared.

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Data availability: Authors have used third party data (from another individual) and therefore do not own the data.

Ethics approval and consent to participate: The present report has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

Authors' contributions: EG, AZ, VR, GP and ET analyzed and interpreted the patient data. VR, SD, EG and GP performed the surgical treatment. ET performed the histological examination. AZ, EG, ET and GP contributed in writing the manuscript. All authors read and approved the final manuscript.

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