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Case Report

Solitary Fibrous Tumor: A Case with a Review

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Abstract:

Introduction: solitary fibrous tumor (SFT) is a rare intraoral occurring tumor. Buccal mucosa is among the most commonly affected sites. SFT lesions are linked to a *NAB2-STAT6* fusion gene. The lesion exhibits well circumscribed feature in both radiographical and histopathological examinations. The lesion shows STAT 6, CD34, bcl-2 and CD99 positive expression. Unfortunately STAT6 and CD 34 expression are lost in highly malignant lesions, making the diagnosis challenging.

Case report: a 43 –year- old female presented with a painful lesion in the left buccal mucosa. Upon surgical excision and histopathological examination the lesion revealed a bland spindle cell lesion with nuclear STAT6 and beta catenin cytoplasmic reactivity. After excision the case was followed up for 6 months and showed no recurrence.

Discussion: the presented case showed a typical presentation the same as the literature reported cases. However, the associated pain was coming in contrast to the most published cases. Although being usually negative in SFT, beta catenin showed cytoplasmic reactivity in our presented case.

Conclusions: SFT is usually not associated with high recurrence rate, but the hypercellularity, pleomorphism and necrosis should be taken into consideration as they may be associated with high recurrence and metastatic potentials.

Keywords: Solitary Fibrous Tumor (SFT), Hemangiopericytoma, Spindle cell lesion.

Review of literature:

Solitary fibrous tumor (SFT) is a rare tumor of mesenchymal origin. It is proposed to have a fibroblastic origin. The lesion was firstly described as a pleural lesion in 1931. Extra-pleural locations were documented in the visceral organs, being the most common sites followed by extremities, abdominal cavity and retroperitoneal sites. Head and neck lesions are rare accounting for about 10- 15% of published cases. In head and neck region orbits are the most common sites, however sinonasal lesions are also a frequent site of incidence. Oral cavity and salivary glands are also considerable sites of occurrence **[1]**.

In 2017, the lesion was classified in the fourth edition of WHO classification of head and neck tumors as a sinonasal lesion. This categorization was changed in the latest head and neck tumors classification made by WHO, as the lesion is now introduced in the soft tissue tumors**.** SFT is specifically defined by a *NAB2-STAT6* fusion gene that is responsible for its pathogenesis and almost detected in all documented lesions. This fused gene could be detected by in situ hybridization to facilitate the diagnosis of such lesions **[2].**

Clinically, SFT is mostly presented as a deep seated lesion and rarely as a superficial lobulated mass. SFT mass grows in a painless and slowly growing fashion, however nasal obstruction, epistaxis, sinonasal discharge, pain, headache, visual changes and proptosis are site specific findings. Males are affected with SFT more than females with wide age range of occurrence. Radiographically, CT and MRI imaging demonstrate SFT as a well circumscribed lesion with non-infiltrative pattern. Although being rare, dedifferentiated or poorly differentiated lesions show slightly infiltrative borders **[3].**

Histopathological examination of SFT usually demonstrates a non-encapsulated but well circumscribed spindle cell lesion with little areas of slightly ovoid cells. The lesion exhibits patternless architecture with alternative hypo and hypercellular areas. The cells are bland and are arranged in sheets around largely dilated blood vessels in the form of stag horn pattern or hemangiopercytomatous pattern. Keloid type collagen deposition is focally detected. Hypercellularity accompanied by pleomorphism, necrosis, increased mitosis and infiltrative borders are indications of aggressive lesions. Lipomatous subtype shows mature lipocytes and giant cell rich one shows tumor giant cells, but both are rare in head and neck region.

Immunohistochemical staining shows highly specific STAT6 nuclear staining and occasional positivity of CD34, bcl-2 and CD99. Malignantly transformed lesions may show loss of both nuclear STAT6 and CD34 staining. S100, SOX10, desmin, actins and nuclear beta catenin are all negative in immunohistochemical examination. This pattern of immunohistochemical staining is highly valuable in the differential diagnosis of SFT from other spindle cell lesions **[4].**

The majority of SFT lesions are benign, but lesions featuring infiltrative borders, pleomorphism and hyper cellularity are linked to an aggressive behavior. SFT shows low recurrence rates, however recurrent lesions show a more destructive growth pattern. Recently, dedifferentiated SFT lesions are described as lesions that exhibit a high grade tumor component in combination with a conventional tumor component. The lesions are diagnosed as dedifferentiated lesions when abrupt tarnation from low to high grade pattern is detected. The diagnosis of dedifferentiated lesions is crucial as they are exhibiting a highly destructive behavior, worse prognosis and require more aggressive treatment approach **[5].**

Case presentation:

The presented case is for a 43 -year- old female who attended to The Oral and Maxillofacial Surgery Department, Cairo University with a cheek soft tissue mass. The patient had a painful cheek mass measuring about 2x2 cm **(fig.1).** The lesion appeared one year before the presentation and was covered by normally colored mucosa. The patient had no contributory dental or medical history. The submandibular lymph nodes were examined, and they were palpable and not fixed. An excisional biopsy was performed and the gross examination of the lesion revealed, an irregular firm mass with solid and greyish white cut surface. The lesion was measuring about 3x 3.5 cm on macroscopic examination **(sup.1)**

Histopathological examination of the lesion revealed a well circumscribed connective tissue lesion. The lesion was exhibiting evident hypercellularity **(fig.2)**. Areas of loose arranged hypocellular pattern were observed **(fig. 4).** The lesion was formed of bland spindle cells dispersed along thin parallel collagen bands. Numerous endothelial lined stag horn shaped blood vessels were detected. Inflammatory cell infiltrate was focally observed **(fig.2- 3).** Areas of cells with slightly ovoid outline and clear cell change were evidently detected focally **(sup.2-3).** The areas of clear cell changes were accompanied by keloid type collagen deposition **(sup.3)**. Immunohistochemical staining was performed and the lesion showed diffuse nuclear STAT6 **(fig. 4 and sup. 6-7)** and cytoplasmic beta catenin positivity **(sup. 4)**. SMA and ALK immunostaining were carried out and SMA showed scattered positivity **(sup. 5)** however, ALK showed negative reaction. The patient was followed up for 6 months and showed no evidence of recurrence.

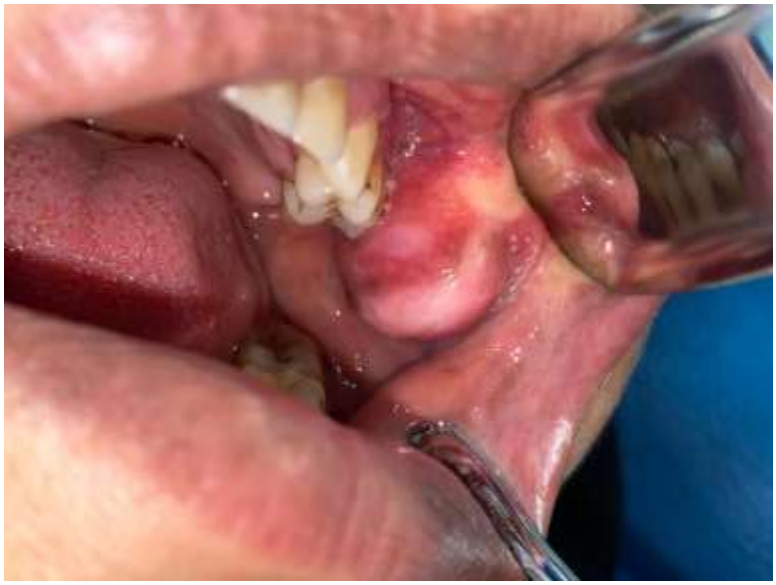

Fig. 1 Clinical presentation of the lesion showing a well circumscribed lesion originating from the buccal mucosa.

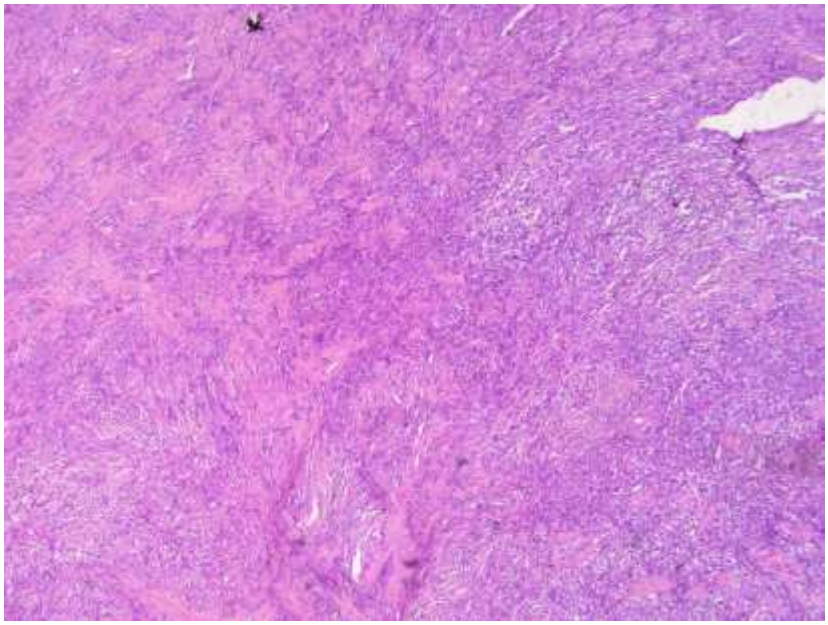

Fig. 2 Histopathological examination of H and E stained sections showed hypercellular areas of bland spindle shaped cells.

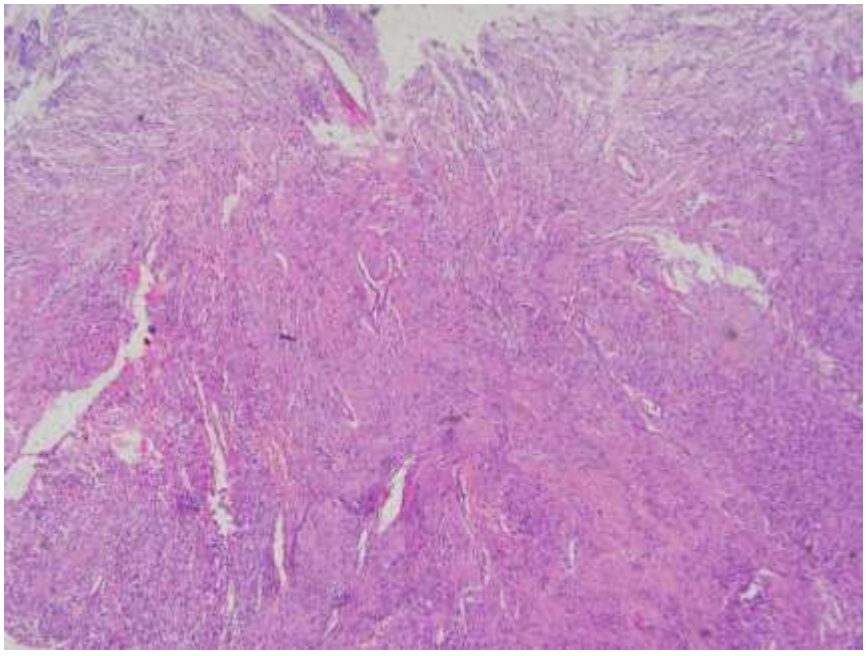

Fig. 3 Hypocellular areas were evident and accompanied by multiple stag- horn vessels.

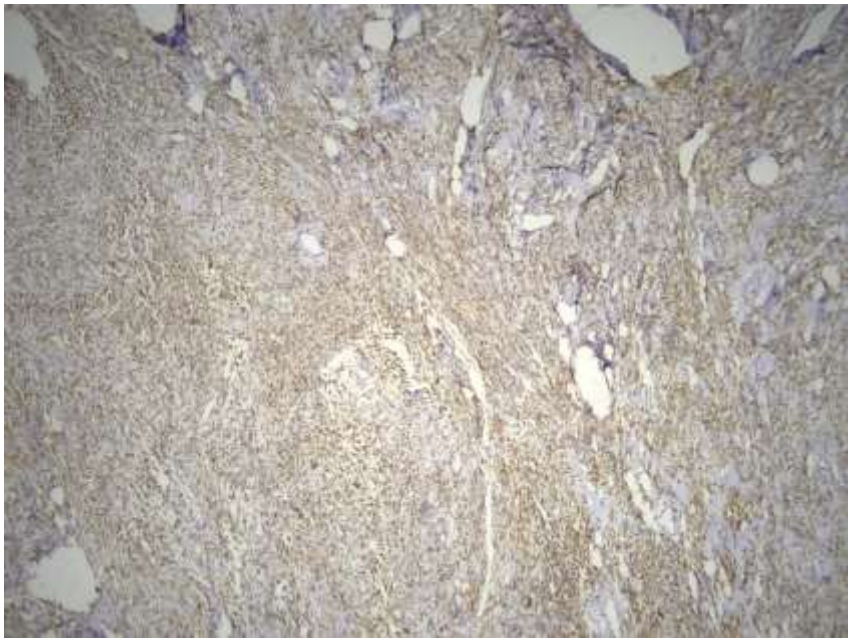

Fig. 4 Immunohistochemical staining was showing a diffuse STAT6 nuclear positive reaction.

Discussion:

SFT is a rare mesenchymal tumor with fibroblastic origin. The oral lesions are much more rare than other sites. The buccal mucosa is the most common site of occurrence intra orally, this finding is consistent with our presented case. Tongue and palatal mucosa are also sites of the lesion's intraoral existence **[6].**

Although affecting males more commonly and has a wide age range, there are few reported cases occurring in females. This came in line with our reported case **[7].** SFT lesions mostly show a slowly growing pattern as we reported in this case **[8]**. Although pain is rarely reported, our patient suffered a painful cheek mass**.**

The typical histopathological findings of SFT are well demonstrated in the presented case as being non encapsulated and well circumscribed spindle cell lesion, besides the existence of multiple stag-horn vessels. The main immunohistochemical staining differentiating SFT from other spindle cells lesions is the nuclear STAT6 positivity, and that was evident in our case. Although being usually negative, beta catenine immunohistochemical staining in the presented case showed cytoplasmic positivity. In the site of presentation of the current lesion, nodular fasciitis is the first lesion in the differential diagnosis list but nodular fasciitis shows positive SMA staining in contrast to the present case **[9].**

Most of SFT lesions are benign with low incidence of recurrence after surgical excision. Recently new insights into the possible risk factors that require close long term follow up are established such as, age of occurrence, tumor size, necrosis and mitotic rate **[10].** In the reported case the lesion showed no features that need a long term follow up for the fear of recurrence or metastasis.

Conclusions:

SFT is rarely presented intraorally, with the buccal mucosa and tongue being the most common intraoral sites. In the oral cavity the lesion should be differentiated from pleomorphic adenoma as both don't possess the same prognosis. Although being not highly recurrent lesions, SFT with hypercellularity, pleomorphism, high mitotic rate and necrosis should take into consideration, as these criteria may increase the rate of recurrence and metastasis.

Conflict of interest:

We have no conflicts of interest.

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Ethical approval:

This work was approved by the Research Ethical Committee at the Faculty of Dentistry, Cairo University, done in compliance with the Helsinki Declaration, and written informed consent was obtained from the patient.

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