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Solitary Fibrous Tumor of the Lower Lip; A Rare Case Report

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A B S T R A C T

Solitary Fibrous Tumor (SFT) is a rare benign neoplasm primarily associated with the pleura. However, it has also been identified in diverse locations, including the skin, deep soft tissues, genitourinary system, gastrointestinal tract, and the head and neck region, including the oral cavity. There is no gender predilection for SFT of the oral cavity and it can occur at any age. We herein present a case of SFT on the lower lip of a 19-year-old male patient and describe our diagnostic approach. The histopathological findings showed typical features of SFT with immunoexpression of CD34, CD99, BCL2, and STAT6, along with low expression of Ki67 in 10% of the tumor cells.

Keywords: Solitary Fibrous Tumors; Lip; Pathology; Neoplasms

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INTRODUCTION

The Solitary Fibrous Tumor (SFT) is a rare benign spindle cell neoplasm that was first described in 1937 by Klemperer and Coleman [1]. Initially, it was described as a pleural neoplasm derived from mesothelial or submesothelial fibroblast [2]. After the World Health Organization (WHO) revised the classification of pleural tumors in 1999, SFT was eventually classified as a distinct entity and was removed from the mesothelioma subgroups. In 2002, it was determined that most neoplasms previously diagnosed as hemangiopericytoma (HPC) could be reclassified as other soft tissue tumors, such as solitary fibrous tumors (SFT), a distinction that had been under scrutiny since the 1990s [3]. SFT has also been found to occur in other anatomic locations, including the skin, deep soft tissue, genitourinary system, gastrointestinal tract, and head and neck region, including the oral cavity (4].

SFTs of the oral cavity occur as slow-growing masses, mostly involving the buccal mucosa, lip, tongue, and alveolar mucosa [5]. It is often an incidental finding without symptoms, but sometimes they are symptomatic due to pressure on adjacent structures . Consequently, it cannot be distinguished from other reactive and neoplastic lesions that mostly affect these areas, and a complete histological and immunohistochemical

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examination, mainly with STAT6, CD34, CD99, and BCL2, may be required to identify the final diagnosis [4,6].

SFTs exhibit a broad histological spectrum, often presenting with alternating hypercellular and hypocellular regions, accompanied by varying degrees of myxoid background changes. Their diverse histological architecture poses a diagnostic challenge, as they are composed of spindle to oval-shaped nuclei with minimal cytoplasm, interspersed with irregularly arranged collagen fibers in a patternless distribution. While some areas display a high tumor cell density, others are hypocellular, characterized by an increased proportion of stromal collagen [7]. In this report, we present a rare case of SFT occurring on the lower lip of an adult male patient.

CASE PRESENTATION

A 19-year-old man was referred to our clinic for an excisional biopsy and a pathological examination. His chief complaint was pain and swelling of the lower lip but was otherwise healthy with no systematic disease or history of oral cavity surgery or trauma. However, he reported Coronavirus disease two weeks before he was diagnosed with the lesion. Clinically, the lesion appeared as a solitary ulcerative exophytic mass on the inner right side of the lower lip, measuring 2×2cm. Differential diagnosis included reactive lesions and mucocele. It was surgically excised under local anesthesia, revealing a pink-gray mass with a firm consistency, measuring 2×2×1.5cm [Figure 1].

Histopathological examination revealed a well-circumscribed, ulcerated neoplastic lesion composed of haphazardly arranged cells with round to oval nuclei, indistinct cytoplasmic borders, and an abundance of blood vessels. The tumor cells exhibited either a spindle shape arranged in short fascicles, or a disorganized pattern (Figure 2, A and B). Immunohistochemically, the tumor cells showed membranous positivity for CD34 (QBEnd/10, dilution:1:50-1:100), cytoplasmic positivity for CD99 (HO36-1.1 , dilution1:25-1:50), and focal positivity for BCL2 (rMM-B1, dilution 1:50-1:100) and STAT6 (EP325 ,dilution1:50 -1:100) as shown in Figure 2 C-H. Ki67 (rMM-K1 ,dilution 1:100-1:200) staining was weak and observed in 10% of tumor cells [Figure 2, I]. All antibodies were obtained from Long-island Antibody Diagnosis Company, Shanghai, China. The tumor was negative for desmin (Figure 2, J) and CD68 (Figure 2, K). Based on the combined histopathological and immunohistochemical findings, the lesion was diagnosed as a “solitary fibrous tumor”.

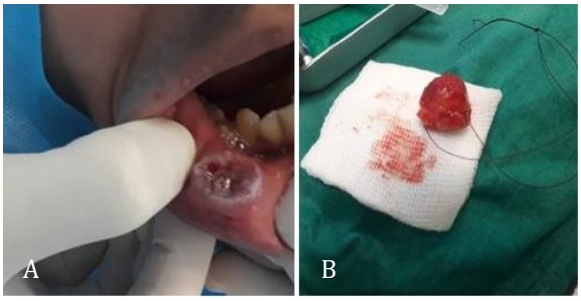

Fig 1. A) Exophytic ulcerative soft tissue lesion on the lower lip; B) Macroscopic findings showed a pink-gray nodular mass

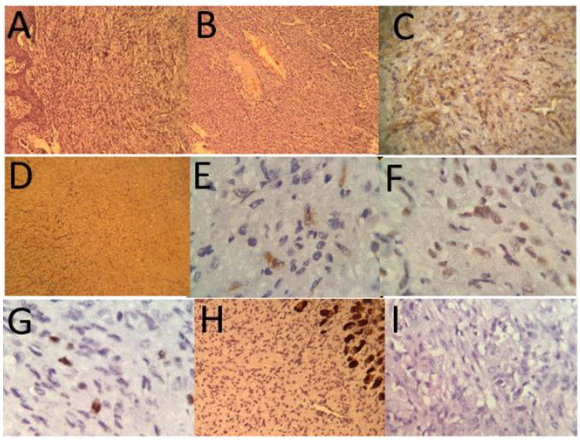

Fig 2. Histopathological and immunohistochemical findings; A and B) Proliferation of spindle cells in a vascular stroma containing dilated vessels and mononuclear inflammatory cells. The squamous lining is seen on the left (×40, H&E); C) The spindle cells show diffuse positivity for CD34. An internal control of vascular epithelial cells is evident (×100); D) Tumor cells show positive staining for CD99 (×40); E) Strong diffuse immunostaining for BCL2 (×400); F) diffuse nuclear expression of STAT6(X400); G) Ki67 proliferative index is less than 10% (×400); H) The tumor cells are negative for desmin. Muscle around the tumor serves as positive internal control (upper right) (×40); I) The tumor cells are negative for CD68 (×100).

The patient underwent complete surgical excision and was followed for 12 months, during which no evidence of recurrence was observed [Figure 3].

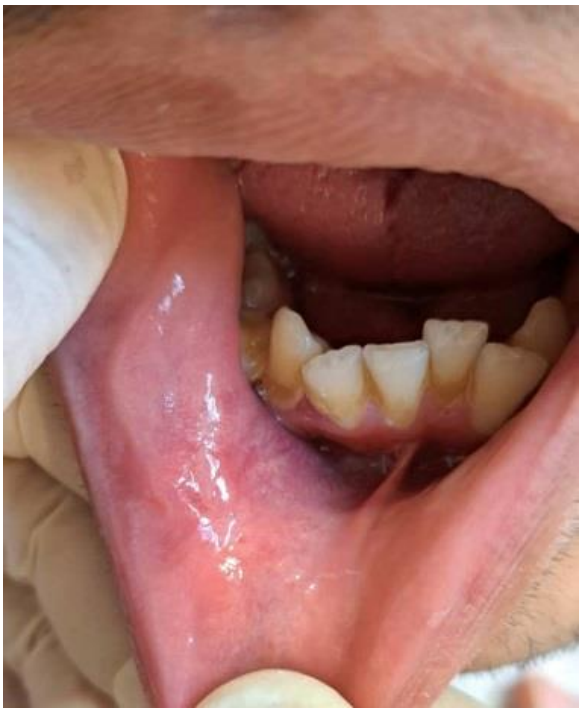

Fig 3. 12-month follow-up showed good healing and no evidence of recurrence

DISCUSSION

SFT was initially mischaracterized as originating from mesothelial tissue and has been referred to by various names over time such as localized fibrous tumor, localized mesothelioma, benign mesothelioma, solitary fibrous mesothelioma, pleural fibroma, submesothelial fibroma, and subserosal fibroma. Nevertheless; recent studies have indicated that SFT is a mesenchymal neoplasm of fibroblastic origin [7,8] and therefore, the lesion can be located in all mesenchymal tissues of the body [9]. The head and neck, including the oral cavity, are among the areas susceptible to this lesion, and are affected in only 3% of cases [5,10]. To date, only two cases (1.3%) have been reported in the Middle East.

Recent studies have identified two fusion genes, NAB2-STAT6 and NFIX-STAT6, with several variants that are relatively effective in differentiating SFT from histologically similar tumors [11,12]. STAT6 plays a critical role as a transcriptional activator, mediating tyrosine phosphorylation in response to IL-4 or IL-13. The NAB2-STAT6 fusion facilitates STAT6 target gene activation through the nuclear translocation of the STAT6-NAB2 transcript, while NFIX, similar to NAB2, supports the nuclear retention of STAT6 [13]. Emerging evidence highlights the oncogenic potential of STAT6 in SFT development, independent of the tumorigenic roles of NAB2 and NFIX [14]. A recent study reported diffuse and robust nuclear expression of STAT6 in all 52 SFT cases examined immunohistochemically [15]. Similarly, in a review by Doyle et al., STAT6 nuclear staining was observed in 59 of 60 (98%) SFT cases, while all other tumor types tested negative [12].

Although SFT of the oral cavity has no sex predilection and can affect any age, a review of 21 oral cases, reported a male to female ratio of 12/9 and a median age of 51 years [3]. Another study mentioned an equal distribution between both sexes, and a higher prevalence in the 5th and 6th decades of life [7]. A systematic review in 2019, showed more oral lesions in females (55.9%) with an average age of 49.4 years [5]. Although most studies report that this lesion can develop across all age groups, it is more commonly observed in patients aged 50 to 60 years. This makes the current case, involving a 19-year-old patient, an uncommon presentation.

Grossly, SFTs of the oral cavity are usually reported to be firm, submucosal, well-circumscribed, and measuring 0.1 to 9.3cm in size. The most common areas are described as buccal mucosa (45.6%), tongue (15.4%), and palate (6.6%). Patients generally have no pain; however, in some individuals, as in the present case, painful lesions have been reported [5].

Diagnosing SFT can be challenging due to its varied histological features across cases. Rodríguez-Gil described several diagnostic criteria, including circumscription, alternating hypercellular and hypocellular sclerotic foci, spindle-shaped or ovoid cells with scant, poorly-defined cytoplasm, interwoven collagen fibrils, and CD34 positivity [8]. Historically, the diagnosis of HPC was questioned due to its morphological overlap with SFT. Currently,

tumors previously classified as HPC are recognized as SFTs, with the term HPC reserved for sinonasal lesions with myxoid differentiation, attributed to pericytes [3,16].

Microscopically, SFTs exhibit diverse patterns, ranging from round-to-spindle cell arrangements to alternating cellular and hypocellular regions separated by collagen fibers [8]. In our case, the tumor displayed a patternless architecture, with spindle cells organized in short fascicles or a disorganized arrangement. Additional features observed in some cases, such as giant cells, mast cells, lymphocytes, and myxoid changes, were present in our case, along with mononuclear inflammatory cells.

Immunohistochemically, SFTs are usually positive for CD34, CD99, BCl-2, and vimentin, but cytokeratins, desmin, CD68, and S-100 protein are typically negative in these lesions [16,18]. CD34 is one of the most important diagnostic markers. One study demonstrated that 97.2% of benign and malignant tumors showed positivity for this marker, while a small subset (2.8%) were negative [5]. Our case was positive for CD34 and Ki67, and negative for desmin and CD68. Ki-67 expression has shown correlation with poor prognosis in different malignancies like breast and prostate cancer. A MIB-1 proliferation index (Ki-67) of ≥10% has been linked to adverse outcomes in SFT [19], suggesting that malignancy is unlikely in our case.

SFTs are typically managed through complete surgical excision with wide resection margins, as achieving clear margins significantly lowers the risk of recurrence [20]. In some studies, radiotherapy and chemotherapy have been proposed as adjunctive treatments following primary surgery [7]. There were 150 cases reviewed in 2019, of which 14 were malignant, and among those, there was one incident of metastasis and one recurrence. Two Middle Eastern cases showed no malignancy, similar to our case [5]. No additional treatment beyond the usual excision is required for malignant SFTs, However, long-term follow-up is required due to the risk of recurrence and metastasis [21]. Our patient was followed and after 12 months, there was no evidence of recurrence [Figure 3]. In summary, we presented a benign oral cavity SFT with rare features, including occurrence in a young age and an atypical site. The lesion demonstrated a favorable outcome following complete surgical excision.

CONCLUSION

This report documents a rare case of SFT in the Lower lip as a rare oral cavity neoplasm. Clinical evaluation combined with immunohistochemical and molecular analysis is vital for accurate diagnosis. An effective treatment for SFT in the oral cavity is surgical excision, with prolonged follow-up being essential to evaluate the risk of recurrence.

CONFLICT OF INTEREST STATEMENT

None declared.

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