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Annals of Surgical Case Reports & Images

Volume 1 | Case Report

Oral solitary fibrous tumor in the retromolar region; rare localization with molecular and clinical aspects

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Abstract

Solitary Fibrous Tumor (SFT) is a rare type of soft tissue tumor that typically exhibits benign clinical characteristics. SFT of the oral cavity is a rare condition. The differential diagnosis can be challenging due to the histomorphologic diversity and clinical similarity to other mesenchymal tumors.

This article examines a case of SFT found in the retromolar region of the mandible. It discusses the clinical presentation and diagnosis, as well as the histological and immunohistochemical characteristics of the tumor.

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Received: May 30, 2024

Accepted: Jun 21, 2024

Published Online: Jun 28, 2024

Journal: Annals of Surgical Case Reports & Images

Online edition: https://annscri.org

Cite this article: Secen IA, Yakuboglu SK, Toraman M, Gultekin SE. Oral solitary fibrous tumor in the retromolar region; rare localization with molecular and clinical aspects. Ann Surg Case Rep Images. 2024; 1(4): 1035.

Introduction

Solitary Fibrous Tumor (SFT) is an uncommon benign neoplasm that arises from mesenchymal tissue. Initially documented by Klemperer and Rabin in 1931 [1], it was believed affect only pleura and peritoneum, it is now recognized in various locations, including the oral cavity [2]. Oral SFT can develop in individuals of all ages and typically affects female adults [2]. Based on the most recent categorization by the World Health Organization (WHO) for head and neck tumors, SFT is categorized as a borderline/low-grade mesenchymal tumor [3]. From a clinical standpoint, it is not possible to distinguish it from other reactive and neoplastic lesions of the oral cavity. As it is referred "the many face tumor" in some papers [4], it shows very similar histological features with other soft tissue tumors. Thus, the diagnosis should rely on clinical, histomorphological, Immunohistochemical (IHC), and molecular findings. The objective of this research is to report an uncommon occurrence of a SFT in the oral cavity and to highlight the significance of IHC and molecular analysis in differential diagnosis.

Case report

A 63-year-old female patient presented to the Oral and Maxillofacial Radiology Department needing dentures. The patient had a medical background of systemic hypertension and diabetes mellitus and adhered to her medication schedule consistently. The intraoral examination revealed a pedunculated, vascularized, painless nodular exophytic mass with a rubbery-firm texture. The mass measured 1.4 cm in its largest dimension and was located in the retromolar region of the mandible (Figure 1). The patient stated that she had no awareness of the lesion and had not experienced any previous traumas. The extraoral examination yielded normal results. The preliminary diagnosis was a traumatic fibroma. The lesion was excised under local anesthesia, fixed in 10% buffered formalin, and sent to the Department of Oral Pathology.

Grossly, the specimen was 1.4 x 0.9 x 0.7 cm in size with an encapsulated, whitish, firm-cut surface. Histopathological examination revealed the proliferation of spindle cells with thin

fibrous encapsulation. The tumor cells had fusiform nuclei with a pale cytoplasm and no atypia. There was no evidence of mitosis or necrosis. The spindle-shaped tumor cells in a storiform pattern and thin-walled, branching-staghorn pattern vascular structures were observed in the collagenous stroma (Figures 2A,2B). To distinguish from soft tissue sarcomas, an IHC panel and FISH were conducted. The tumor exhibited positive immunoreactivity for CD34 and STAT6, while negative for S-100 and beta-catenin. ki67 proliferation index was less than 1%. Furthermore, using chromosome 18q11.2 as a translocation partner, sections from paraffin blocks were examined for chromosomal translocations by FISH with dual-color, break-apart probes. SYT-translocation was yielded negative. Based on these results, the final diagnosis was oral SFT. There was no recurrence detected three months after removal (Figure 3).

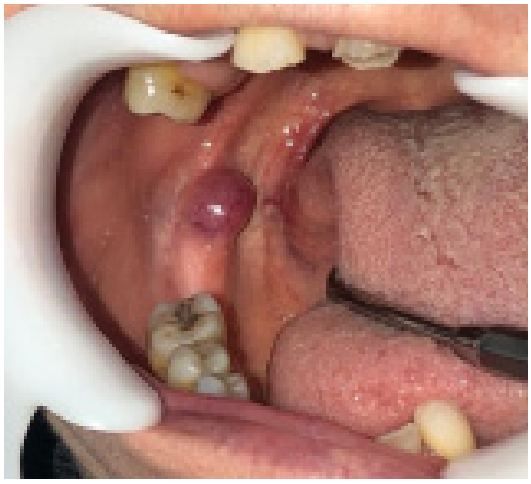

Figure 1: Clinical view of the lesion.

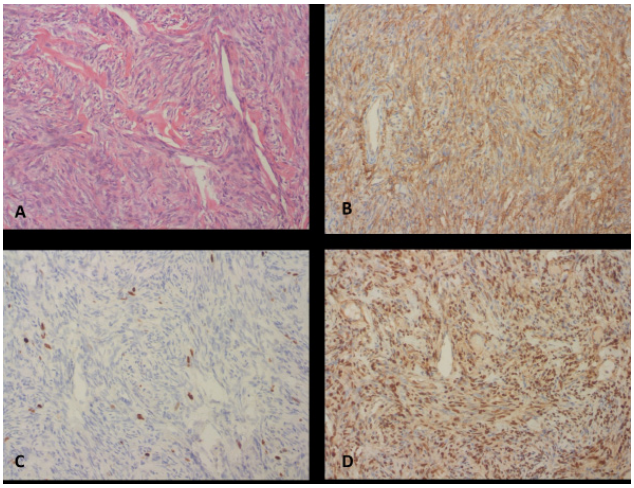

Figure 2: Histopathological view of the case. **(A)**: Hematoxylin & eosin (x200 magnification). **(B)**: CD34 (x200 magnification, DAB). **(C)**: ki67 (x200 magnification, DAB). **(D)**: STAT6 (x200 magnification, DAB).

Discussion

SFT is a unique tumor originating from fibroblasts, initially found in the pleural cavity, with potential to develop in any anatomical site [1]. Extrapleural cases, affecting 27% of the head and neck region, can be observed in various locations such as the orbit, nasal cavity, paranasal sinuses, thyroid, and salivary glands [5]. SFTs in the oral region are extremely uncommon and often impact the buccal mucosa, tongue and hard palate [6]. SFT rarely occurs in the retromolar region. Recent research estimates a 2.6% incidence of SFT in the retromolar area, with a total of 4 reported cases [7]. To the author's knowledge, this case represents the fifth case of SFT occurring in this area. Lesions are usually characterized by well-defined submucosal growths that might vary in size and duration. The color and texture of the covering mucosa are often uniform [2,6]. Pain in oral SFTs is rarely documented, and ulceration is often caused by local trauma [7]. In this case, there was an elevation in mucosal appearance without local trauma, resembling those described in existing literature.

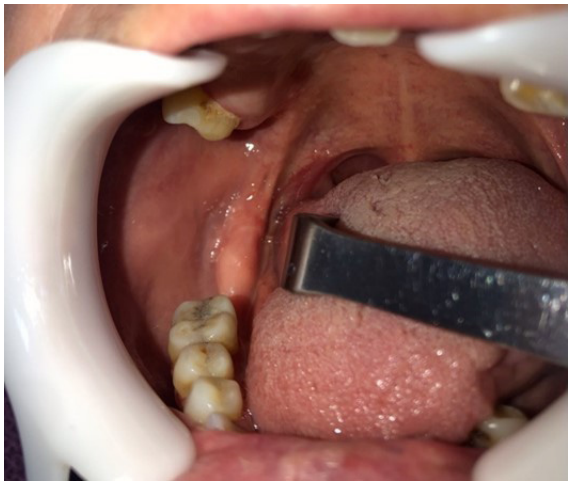

Figure 3: Clinical view after 3 months.

SFTs are histologically unique neoplasms, and the heterogeneity of their microscopic features can be somewhat challenging to diagnose. The diversity of morphological features and the "patternless" growth pattern pose diagnostic challenges for SFT and require differential diagnosis with benign and malign mesenchymal tumors. Oral SFT is histologically characterized by a varying density of spindle tumor cells, extensive collagen deposition, and thin-walled blood vessels resembling hemangiopericytoma. There is no evidence of cytological atypia or necrosis [8]. The histopathological characteristics of our case were comparable to those described in the literature. It is essential to differentiate oral SFTs from other soft tissue lesions, as they can range from a simple traumatic fibroma to a synovial sarcoma [4], due to their non-specific clinical presentation and diverse histologic characteristics [5]. Oral SFTs are often diagnosed using IHC analysis employing CD34, CD99, and Bcl-2 markers [9] or identifying the chromosomal rearrangement NAB2-STAT6 genes with Fluorescent In Situ Hybridization (FISH) [10,11]. SFT is predominantly characterized by a positive expression of CD34, while there are occasional instances when it may exhibit a negative expression. Hence, it is advisable to utilize markers such as CD99 and Bcl-2 in conjunction with CD34. In a recent research [2], it was observed that 72% of oral SFTs had CD99 positivity. However, this particular case was found to be CD99 negative. The S-100 protein tested negative, consistent with the findings reported in the literature [2,8]. By utilizing molecular tools, an intrachromosomal fusion involving NAB2 and STAT6 genes at the 12q.13 locus was identified by DNA sequencing [5,12]. The consistent presence of the NAB2-STAT6 fusion gene in nearly all instances of SFT suggests that this genetic alteration is the main cause of SFT development, regardless of the location and appearance of the tumor [5]. According to the literature, SFT demonstrates STAT6 positive in almost 99% of cases [13,14]. Corroborating these results, this case exhibited strong immunopositivity for STAT6. Given the histological similarities, it was crucial to differentiate from synovial sarcoma, which can

occur in the oral region. FISH is widely regarded as the most dependable technique for detecting synovial sarcoma due to its ability to detect SYT-SSX translocation fusion genes at the molecular level. According to the literature, FISH has been found to yield accurate results in 82% of synovial sarcoma [15]. In our case, the results of beta-catenin immunostaining and the presence of SYT-SSX fusion genes were both negative, leading us to dismiss synovial sarcoma from the list of possible diagnoses.

The literature indicates that excision is sufficient in the case of oral lesions [2,4]. The existence of malignancy in SFTs is typically indicated by the patient's advanced age, a large tumor size, and malignant histological characteristics [4,5]. While malignant oral SFT is rare, it is nonetheless possible. Regarding our case, the tumor size was small, with mitotic activity measuring less than 1%. Despite the low likelihood of malignancy in our case, the patient is being followed-up for 3 months, with no recurrence or evidence of malignant transformation.

Conclusion

To conclude, we reported an uncommon case of SFT in the retromolar region. We discussed the clinical, histopathological, and molecular features. The correct diagnosis is crucial for the appropriate treatment and management of SFTs. Given its rarity in the oral cavity, differential diagnosis with other lesions in the oral mucosa and periodic follow-up are considered necessary.

Declarations

Conflict of interest: None.

Authors contributions: SKY and MT examined the patient and performed treatment and follow-up. SEG and IAS carried out the histological analysis. The laboratory procedures were executed by IAS. First draft of the paper was written by IAS and SKY. SEG and MT reviewed the manuscript's final version.

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