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CASE REPORT

Solitary fibrous tumor of the upper lip: an uncommon case presentation

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Abstract:

Solitary Fibrous Tumor (SFT) is a rare mesenchymal neoplasm, account approximately 3% of mesenchymal tumors in the oral cavity. We present the case of a 65-year-old female patient with a painless swelling in the left upper lip. Clinical examination revealed a firm, smooth-surfaced submucosal lesion without ulceration, non-tender on palpation, and exhibiting a slightly yellowish hue. Computed tomography demonstrated a well-defined, isodense mass measuring 25 mm in diameter, with no evidence of bone invasion. The lesion was surgically excised, and histopathological analysis confirmed the diagnosis of SFT, supported by positive immunohistochemical staining for CD34 and STAT6. The tumor was classified as low risk for metastasis, and the patient demonstrated favorable postoperative recovery. The patient remains under follow-up with the medical team and has been recurrence-free for 07 months. This case underscores the rarity of oral cavity SFT and emphasizes the importance of comprehensive differential diagnosis to guide effective management and reduce recurrence risk.

Keywords: Solitary Fibrous Tumor; Immunohistochemistry; Diagnosis; Lip.

INTRODUCTION

SFTs were first described by Klemperer and Rabin in 1931. Initially, they were considered mesenchymal neoplasms primarily confined to the pleura and lungs[1]. However, it is now well established that SFTs can occur in various parts of the body[2,3]. A multi-institutional clinicopathological study revealed that SFTs exhibit no gender predilection, affecting individuals with a mean age of 52 years, irrespective of sex. The most commonly affected anatomical sites include the orbit and the sinonasal tract, followed by the oral cavity and salivary glands[4].

In the oral cavity, SFTs account for approximately 3% of all mesenchymal tumors, representing a rare entity with significant diagnostic challenges[5]. These neoplasms may present as solitary, well-defined submucosal masses or slow-growing nodules[6,7]. While radiographic features of SFTs are infrequently discussed in the literature, computed tomography (CT) scans are commonly used for evaluation, typically revealing isodense, circumscribed lesions with well-defined margins[7,8].

Statement of Clinical Significance
This case highlights the importance of differential diagnosis in the context of rare oral tumors, such as Solitary Fibrous Tumor, emphasizing the need for a comprehensive and precise diagnostic approach to ensure appropriate clinical management and avoid diagnostic errors.

Benign oral SFTs predominantly occur in the buccal mucosa, whereas malignant variants are more frequently localized to the tongue[5]. Macroscopically, these lesions appear as firm masses, circumscribed and unencapsulated if benign. Malignant SFTs often demonstrate infiltrative growth patterns and may exhibit necrotic areas[9]. Due to these overlapping features with other reactive and neoplastic oral lesions, clinical evaluation alone is insufficient for definitive diagnosis, necessitating histopathological and immunohistochemical confirmation.

Therefore, for an accurate diagnosis, clinical features must be correlated with immunohistochemical analysis techniques and microscopic evaluation. Markers

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such as CD34, BCL-2, and CD99 demonstrate high sensitivity. CD34 and the STAT6 transcription factor are essential for confirming the diagnosis of SFT, although in certain cases, their expression may be lost in malignant variants[5,9,10].

This manuscript details the diagnostic process of a clinical case involving an SFT located in the upper lip region, highlighting the critical role of histopathological methods and immunomarkers in achieving precise tumor characterization.

CASE REPORT

A 65-year-old female patient present to the Oral and Maxillofacial Surgery and Traumatology Department at João de Barros Barretos University Hospital (HUJBB) with a chief complaint of a progressive left upper lip swelling developing over approximately two months.

During history-taking, the patient reported chronic alcohol consumption spanning over 30 years and a 20-year smoking history (cessation occurred two decades prior). No history of trauma or prior lesions in the affected region was noted. The patient denied allergies or significant comorbidities.

The patient's medical history indicates that she is currently undergoing diagnostic evaluation for Alzheimer's disease and is being treated with Donepezil Hydrochloride (10 mg) on a regular basis. The family history did not reveal any relevant information or conditions that could contribute to the present diagnosis.

Physical examination showed the patient to be in good general condition, with preserved consciousness, oriented in time and space, breathing normally, communicative, ambulatory, and afebrile. Extraoral inspection revealed edema in the nasogenian region extending into the submalar area, accompanied by mild flattening of the nasolabial fold (Figure 1A). Intraoral examination showed normochromic mucosa with a painless, slightly yellowish mass located in the left maxillary vestibular region (Figure 1B). The lesion was firm upon palpation, with no signs of fluctuation or instability.

Imaging investigations using conventional computed tomography revealed an isodense image in the mid-lateral region of the left maxilla. Coronal sections showed a well-defined lesion measuring approximately 25 mm, with no signs of involvement of the adjacent maxillary sinus (Figure 2C). Axial sections displayed a round lesion with regular contours in the submalar region (Figure 2A). The 3D reconstruction and sagittal sections further confirmed a well-defined lesion with clear margins and no evidence of involvement of surrounding structures (Figures 2B–D). Complementary laboratory tests indicated no clinically significant abnormalities.

Based on the clinical and radiographic characteristics, the main differential diagnoses, listed in decreasing order of probability, were canalicular adenoma, lipoma, polymorphous adenocarcinoma, schwannoma, and mucoepidermoid carcinoma. The proposed treatment involved total surgical excision of the lesion under local anesthesia, which was performed without

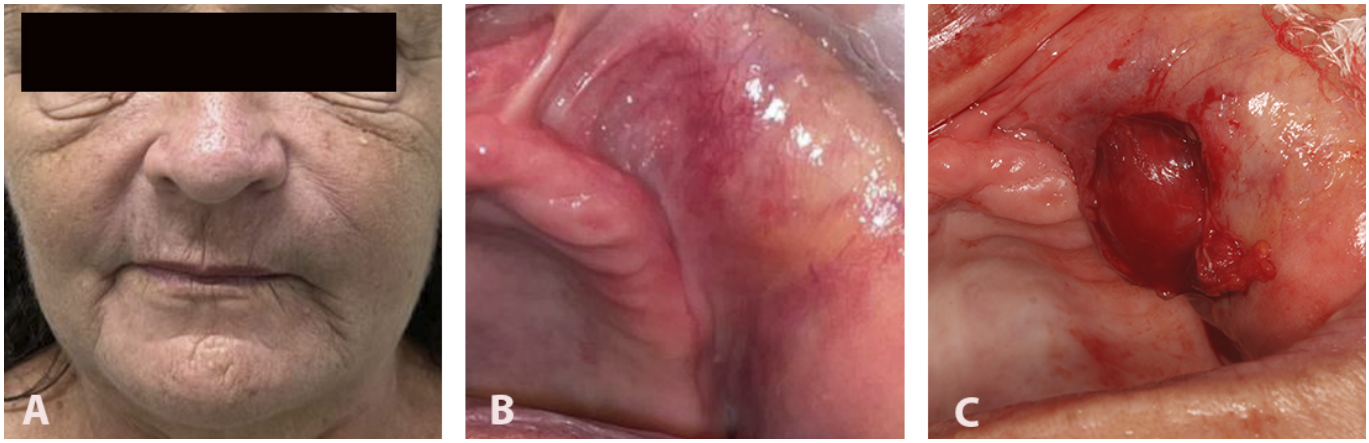

Figure 1. Clinical presentation. (A) Extraoral clinical examination demonstrating edema in the nasogenian region, extending to the submalar region, with a slight obliteration of the nasolabial fold on the left side. (B) Intraoral clinical examination demonstrates a slightly yellowish swelling in the vestibular fold region and a surrounding edematous area. (C) Immediate postoperative period following excisional biopsy in the maxillary vestibular fold region.

complications during or after the procedure (Figure 1C). The patient had a satisfactory postoperative recovery with no complications

Microscopic examination of the excised lesion revealed a mesenchymal neoplasm characterized by spindle cells arranged randomly within a stroma exhibiting a

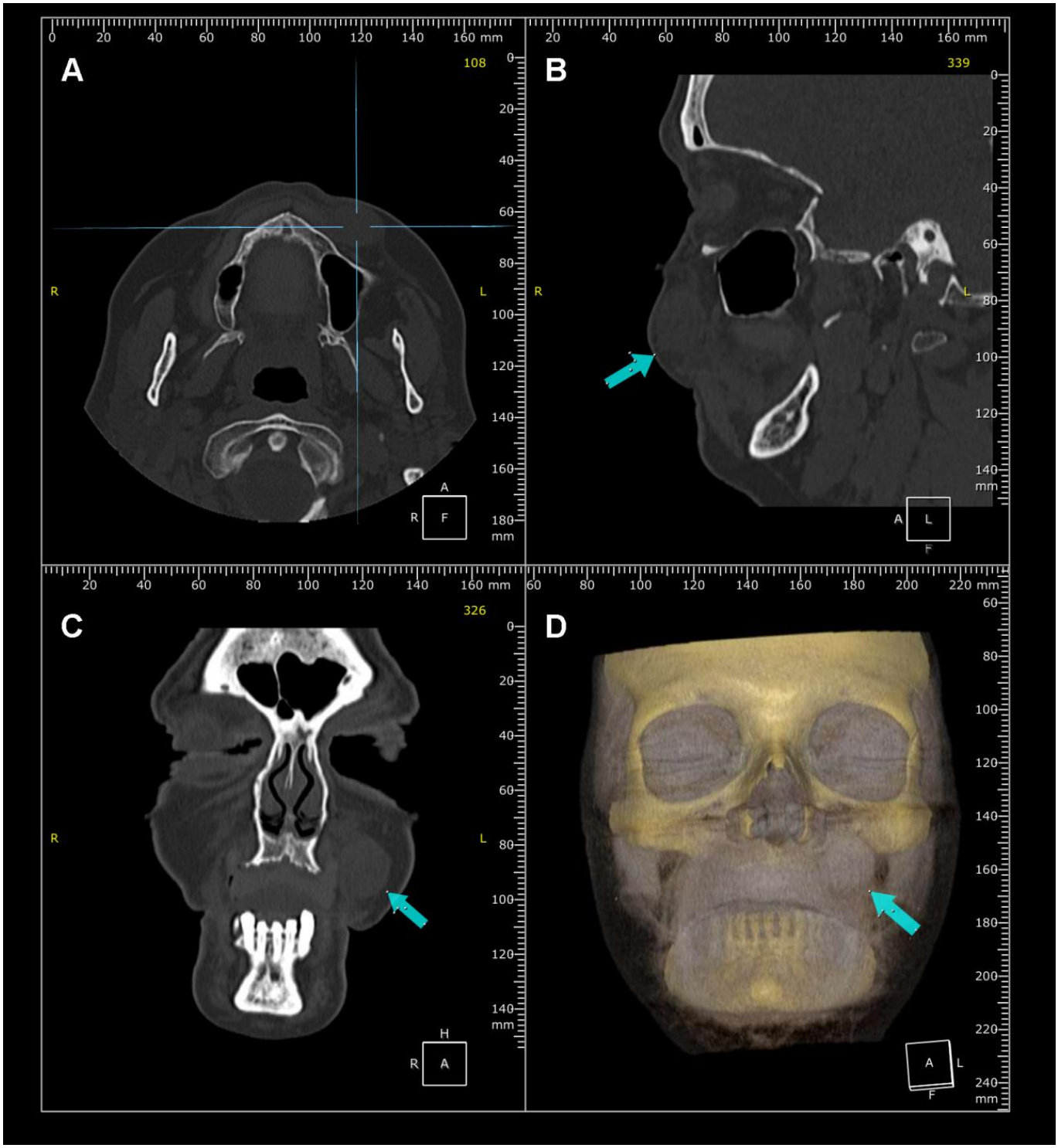

Figure 2. Imaging exams. (A) Conventional Computed Tomography demonstrating an isodense lesion relative to soft tissues, circumscribed, well-marginated, and with no evidence of adjacent structural involvement. Axial View. (B) Sagittal View. (C) Coronal View and (D) 3D reconstruction.

significant degree of collagenization. Numerous tortuous blood vessels displaying a "hemangiopericytoma-like" architectural pattern were also observed. While no marked cytological atypia was noted, areas of localized hypercellularity were evident (Figure 3A). The mitotic index was quantified at two mitoses per 10 high-power fields (HPF), corresponding to an approximate area of 1.96 mm^2.

Immunohistochemical analysis revealed positivity for the markers CD34, CD99, and STAT6, supporting the histopathological diagnosis of SFT based on both morphological and immunophenotypic findings (Figures 3B–D). Additionally, the tumor was negative for the endothelial marker ERG, which helps exclude vascular neoplasms, further reinforcing the diagnosis.

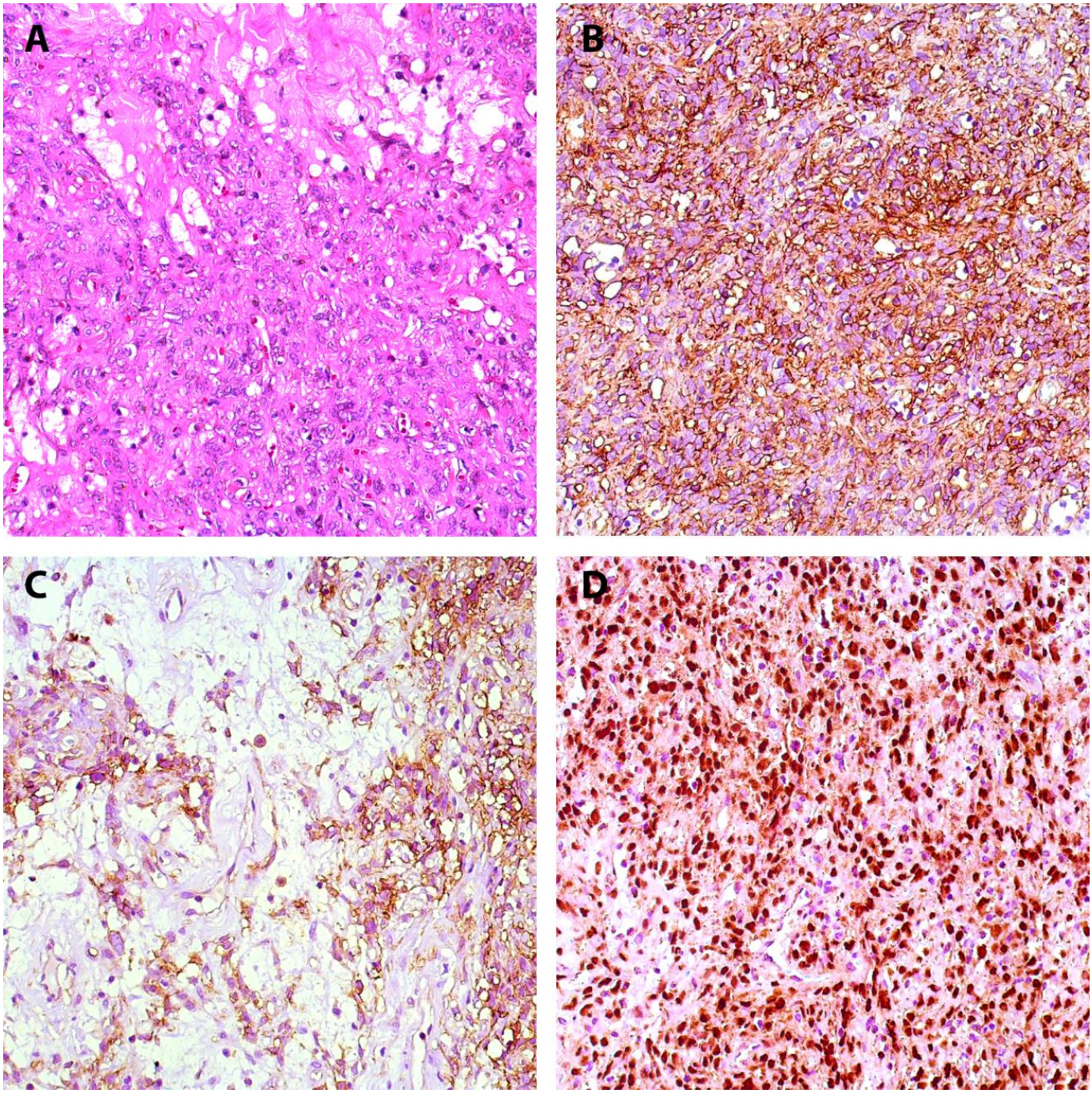

Figure 3. Microscopic and immunohistochemical features. (A) Spindle cells randomly arranged in a collagenized stroma containing numerous tortuous blood vessels with a "hemangiopericytoma-like" pattern (H&E; 100X). (B) Expression of CD34 protein (DAB; 100X); (C) Expression of CD99 protein (DAB; 100X). (D) Expression of STAT6 protein (DAB; 100X).

The combination of CD34, CD99, and STAT6 positivity, along with ERG negativity, aligns with the characteristic immunoprofile of SFT, aiding in its differentiation from other mesenchymal neoplasms.

The metastatic risk classification was performed using the Fifth World Health Organization's (WHO) classification criteria published in April 2020, which incorporate key prognostic factors such as patient age, tumor size, and mitotic count[9,10]. Among the available models, the Demicco model is the most widely applied in clinical practice. It exists in both three- and four-variable versions, with the latter chosen for this case. This model assesses metastatic risk based on age, mitotic activity per mm^2, tumor size, and presence of necrosis, assigning a risk score (low: 0–3 points, intermediate: 4–5 points, high: 6–7 points). For this patient, aged 65, the classification resulted in a total of 3 points — 1 for age, 2 for mitotic count (2 mitoses per mm^2), and 0 for both tumor size (25 mm) and absence of necrosis — categorizing the tumor as low-risk for metastasis[11,12].

After receiving the diagnostic report, the patient provided detailed postoperative care instructions, including general monitoring measures and preventive strategies to prevent complications. She continues to be monitored by the Oral Surgery and Pathology team and is currently 7 months free of signs of lesion recurrence.

DISCUSSION

SFTs are rare mesenchymal neoplasms consisting of spindle cells with an uncertain pathogenesis. These tumors are pluripotent and can differentiate into various cell types, such as mesothelial or fibroblastic cells[5-7]. Often mistaken for other soft tissue tumors, their behavior is notoriously unpredictable, with 10 to 15% of cases showing malignant potential[12,13].

Approximately 5% of SFT cases occur in the head and neck region, with maxillomandibular involvement being particularly rare[13]. Intraoral SFTs are equally distributed between sexes and predominantly affect individuals in their sixties, with an estimated incidence of 1 new case per million people annually. They can arise in various intraoral locations, most commonly affecting the buccal mucosa, followed by the lips and tongue[11,14].

Clinically, SFT present as solitary, well-circumscribed, slow-growing submucosal mass or nodule. They may be symptomatic or painless, with durations varying from 2 months to 5 years. Tumor size range from 5 cm to 8 cm. Oral SFTs resemble a variety of submucosal neoplasms, including schwannoma, myofibroma, and salivary gland neoplasms, making clinical identification particularly challenging[2,5-7,11,13].

Differential diagnosis of SFT in the upper lip can be challenging due to its rarity and the overlap of clinical and epidemiological features with other neoplasms. Various benign and malignant tumors should be considered in this context. For example, Canalicular Adenoma, a benign salivary gland neoplasm, typically presents as an asymptomatic nodular enlargement, often affecting the upper lip, particularly in elderly women[15]. Lipoma, a common mesenchymal tumor, usually appears as a circumscribed, firm or soft mass with a slightly yellowish or pinkish coloration and can occur in various regions, including the upper lip[16]. Polymorphous Adenocarcinoma, a rare malignant neoplasm, commonly affects women around the age of 50 and presents as a slow-growing, painless submucosal nodule without distinctive features, primarily affecting the palate but also potentially occurring in the upper lip[17]. Neurilemoma, or Schwannoma, is a benign tumor of Schwann cell origin, typically presenting as a solitary, encapsulated, slow-growing mass with a smooth surface and a well-defined appearance on imaging studies[18]. Lastly, Mucoepidermoid Carcinoma, the most common malignant tumor of the salivary glands, primarily affects women in their fifth decade of life and manifests as a floating, painless submucosal mass with color variation from red to blue, which may lead to confusion with benign lesions[19].

These hypotheses were initially considered due to the location, clinical history, and pathological characteristics of the lesion. Therefore, a comprehensive histopathological analysis combined with immunohistochemistry is essential for an accurate SFT diagnosis.

SFT, displays marked histological diversity, characterized by spindle or oval cells arranged in haphazard or storiform patterns within a variably collagenous stroma. A hallmark feature is the presence of "staghorn" vascular patterns (resembling hemangiopericytomas/HPC), with perivascular fibrosis and focal myxoid changes. However, staghorn vasculature is not pathognomonic and can occur in other mesenchymal tumors, including soft tissue sarcomas. Intratumoral heterogeneity often includes alternating hypocellular zones and hypercellular regions, contributing to diagnostic challenges[11,13,14]. Tumors historically classified as hemangiopericytomas are now recognized as SFTs due to distinct molecular and immunohistochemical profiles[11].

The 2020 WHO Classification redefines SFT based on the molecular hallmark NAB2-STAT6 gene fusion, detectable via nuclear STAT6 immunohistochemistry

with 98% sensitivity and specificity[10]. STAT6 immunohistochemistry has superseded older markers like CD34 (positive in 75–95% of cases but less specific and often lost in malignant variants) and BCL-2/CD99[11,13]. Additional markers such as cytokeratin, desmin, EMA, α-SMA, and S-100 are consistently negative, aiding in differential diagnosis[5].

The fifth WHO classification published in April 2020, introduced new risk stratification criteria for SFT, categorizing them as low, intermediate, or high metastatic risk based on clinical variables (e.g., age) and pathological features (e.g., tumor size, mitotic index, necrosis)[9,10]. The widely adopted Demicco model evaluates age (<55 vs. >55 years), tumor size (<5 cm, 5–10 cm, 10–15 cm, ≥ 15 cm), mitotic count (0, 0.5–1.5, ≥2 mitoses/mm^2), and necrosis presence (<10% vs. ≥ 10%) to predict metastasis risk and mortality[10,11,12]. These models guide clinical management, with surgical excision achieving negative margins remain the standard treatment for localized SFTs[7,11,13]. Recurrence can occur in up to 10–40% of localized SFT cases, underscoring the need for rigorous follow-up[11]. Adjuvant radiotherapy is recommended for malignant SFTs >5 cm or with positive surgical margins. While tumors without malignant components generally have favorable post-resection prognoses, continuous monitoring remains critical due to recurrence risks[11,12]. Antiangiogenic therapies are under investigation for advanced cases, targeting angiogenesis, driven by NAB2-STAT6 oncogenic fusion and VEGF overexpression, which promote tumor growth and metastatic. Metastatic disease warrants systemic treatment, though no standardized medical approach exists yet[11].

Diagnosis challenges arise from SFTs' nonspecific clinical and radiographic presentation, which overlap with other oral soft tissue lesions. The absence of distinctive features complicates early identification, necessitating thorough evaluation and definitive histopathological analysis to differentiate SFTs from mimics and ensure appropriate management.

CONCLUSION

This report documents a rare case of SFT in the upper lip, highlighting its exceptional rarity. Given its status as a rare oral cavity, neoplasm thorough clinical evaluation combined with immunohistochemical and molecular analysis is essential for accurate diagnosis. Surgical excision is an effective treatment for SFT in the oral cavity, with prolonged follow-up being vital to assess the risk of recurrence.

AUTHORS' CONTRIBUTIONS

HARP: Conceptualization, Writing – review & editing. CBA: Writing – original draft. ECM: Writing – original draft. IML: Data curation, Writing – original draft. NRG: Data curation, Resources. NCM: Writing – review & editing. RHAS: Investigation. SALCU: Investigation. TFF: Supervision. FSCP: Conceptualization, Supervision.

CONFLICT OF INTEREST STATEMENT

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REFERENCES

1. Klemperer P, Rabin CB. Primary neoplasms of the pleura. A report of five cases. Am J Ind Med. 1992;22(1):1-31. https://doi.org/10.1002/ajim.4700220103
2. Ganly I, Patel SG, Stambuk HE, Coleman M, Ghossein R, Carlson D, et al. Solitary fibrous tumors of the head and neck a clinicopathologic and radiologic review. Arch Otolaryngol Head Neck Surg. 2006;132(5):517-25. https://doi.org/10.1001/archotol.132.5.517
3. Chung HR, Tam K, Han AY, Obeidin F, Nakasaki M, Chhetri DK, et al. Solitary fibrous tumors of the head and neck: a single-institution study of 52 patients. OTO Open. 2022;6(3):2473974X221098709. https://doi.org/10.1177/2473974X221098709
4. Smith SC, Gooding WE, Elkins M, Patel RM, Harms PW, McDaniel AS, et al. Solitary fibrous tumors of the head and neck: a multi-institutional clinicopathologic study. Am J Surg Pathol. 2017;41(12):1642-56. https://doi.org/10.1097/PAS.0000000000000940
5. Nunes FB, Sant'Ana MSP, Silva AMB, Agostini M, Canedo NHS, Andrade BAB, et al. Solitary fibrous tumour of the oral cavity: an update. J Oral Pathol Med. 2020;49(1):14-20. https://doi.org/10.1111/jop.12953
6. Shmuly T, Ben Zvi Y, Chaushu G, Kaplan I. Oral solitary fibrous tumor: a retrospective clinico-pathological study and long-term follow-up. Medicina (Kaunas). 2021;57(2):152. https://doi.org/10.3390/medicina57020152
7. Stanisce L, Ahmad N, Levin K, Deckard N, Enriquez M, Brody J, et al. Solitary fibrous tumors in the head and neck: comprehensive review and analysis. Head Neck Pathol. 2020;14(2):516-24. https://doi.org/10.1007/s12105-019-01058-6
8. Lu X, Zhang JL, Zhai X, Liu Q, Hang W. A case report on a nasal and oral cavity involving large solitary fibrous tumor and comprehensive review of case literature. Oral Oncol. 2024;150:106715. https://doi.org/10.1016/j.oraloncology.2024.106715
9. World Health Organization. Classification of tumours soft tissue and bone tumours. 5th ed. Lyon: IARC Press; 2020.

10. Anderson WJ, Doyle LA. Updates from the 2020 World Health Organization Classification of Soft Tissue and Bone Tumours. Histopathology. 2021;78(5):644-57. https://doi.org/10.1111/his.14265
11. Bernardi A, Dufresne A, Mishellany F, Blay JY, Ray-Coquard I, Brahmi M. Novel therapeutic options for solitary fibrous tumor: antiangiogenic therapy and beyond. Cancers (Basel). 2022;14(4):1064. https://doi.org/10.3390/cancers14041064
12. Patil V, Vidya K, Paul S, Tirkey A. Solitary fibrous tumor of the masticator space – report of a rare case. Natl J Maxillofac Surg. 2022;13(Suppl 1):S159-61. https://doi.org/10.4103/njms.NJMS_117_20
13. Tariq MU, Din NU, Abdul-Ghafar J, Park YK. The many faces of solitary fibrous tumor; diversity of histological features, differential diagnosis and role of molecular studies and surrogate markers in avoiding misdiagnosis and predicting the behavior. Diag Pathol. 2021;16(1):32. https://doi.org/10.1186/s13000-021-01095-2
14. O'Regan EM, Vanguri V, Allen CM, Eversole LR, Wright JM, Woo SB. Solitary fibrous tumor of the oral cavity: clinicopathologic and immunohistochemical study of 21 cases. Head Neck Pathol. 2009;3(2):106-15. https://doi.org/10.1007/s12105-009-0111-8
15. Nair PK, Varma BR, Veeraraghavan R, Janardhanan M. Canalicular adenoma: palatal presentation of an uncommon lesion. BMJ Case Rep. 2021;14(9):e243319. https://doi.org/10.1136/bcr-2021-243319
16. Dehghani N, Razmara F, Padeganeh T, Mahmoudi X. Oral lipoma: case report and review of literature. Clin Case Rep. 2019;7(4):809-15. https://doi.org/10.1002/ccr3.2099
17. Mauceri R, Coppini M, Alecci G, Cordova A, Florena AM, Magro G, et al. Polymorphous adenocarcinoma: a systematic review of the literature and presentation of two cases in a less-considered anatomical site. Cancers (Basel). 2024;16(1):220. https://doi.org/10.3390/cancers16010220
18. Martins MD, Jesus LA, Fernandes KPS, Bussadori SK, Taghloubi SA, Martins MAT. Intra-oral schwannoma: case report and literature review. Indian J Dent Res. 2009;20(1):121-5. https://doi.org/10.4103/0970-9290.49059
19. Devaraju R, Gantala R, Aitha H, Gotoor SG. Mucoepidermoid carcinoma. BMJ Case Rep. 2014;2014:bcr-2013-202776. https://doi.org/10.1136/bcr-2013-202776