

Solitary fibrous tumos of the hard palate: a rare entity in oral Cavity

Tumor fibroso solitário do palato duro: um achado raro na cavidade bucal

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ABSTRACT

A case of solitary fibrous tumor is reported. Solitary fibrous tumor is a rare neoplasia characterized by the proliferation of fusiform cells of mesenchymal origin accounting for at least 2% of all soft tissue tumors. In this present case, the initial diagnosis was salivary gland tumor because of the location in the hard palate. Histologically, the tumor was composed by conjunctive tissue with proliferation of oval and fusiform cells. The immunohistochemical analysis was positive for CD34 and CD99. The lesion was treated by surgical resection. The clinical, imaging, histological and immunohistochemical data are discussed in this study.

KEYWORDS

Solitary fibrous tumor; Mouth; Palate, hard; Diagnosis, differential.

RESUMO

Um caso de tumor fibroso solitário em cavidade oral é reportado. O tumor fibroso solitário é uma neoplasia rara caracterizada por uma proliferação de células fusiformes de origem mesenquimatosa e é responsável por menos de 2% de todos os tumores de tecidos moles. No presente caso, o diagnóstico clínico inicial foi direcionado para um tumor de glândula salivar em decorrência da localização em palato duro. Histologicamente, o tumor era composto por tecido conjuntivo com proliferação de células fusiformes e ovaladas. Imunoistoquímica da lesão foi positiva para CD34 e CD99. A lesão foi tratada por ressecção cirúrgica. Os dados clínicos, imaginológicos, histológicos e imunoistoquímica são discutidos neste estudo.

PALAVRAS-CHAVE

Tumor fibroso solitário; Cavidade oral; Palato duro; Diagnóstico diferencial.

BRIEF LITERATURE REVIEW

The Solitary Fibrous Tumor (SFT) was initially described as a pleural lesion in 1931, but overtime, it was recognized by affecting other organs at many anatomical sites of the human body. Over 50% of these tumors occur inside the chest cavity; however, the extra-thoracic tumors have been reported in many sites, including the liver, adrenal gland, skin, head, and neck [1].

The SFT terminology is confusing, and normally based on the histological characteristics, location, and histogenesis of the lesion. SFT of head and neck area is rare and the tumor behavior is not fully understood [1,2].

Clinically, intraoral SFT appears as an asymptomatic lesion of slow growth, a well-circumscribed submucous mass normally involving the oral mucosa which makes difficult to distinguish SFT from other soft-tissue tumors, such as synovial sarcoma, benign

fibrous histiocytoma, dermatofibrosarcoma protuberans, neurofibroma, schwannoma, fibroids, and myelofibrosis [3]. SFT affects mainly female adults[1-6].

This study aimed to report a rare case of SFT in a 35-year-old woman, emphasizing the resources of imaging, clinical-pathologic, and immunohistochemical evaluations for the diagnosis of these tumors.

CASE REPORT

A female patient aged 35 years was referred to the Service of Oral Diagnosis of the District Hospital of Araguaína(Tocantins, Brazil), with main complaint of volume increasing in “the roof of the mouth”, without pain symptomatology and with a slow growth. Intraoral clinical examination revealed a volume increase on right hard palate of 5cm, with bleeding by touching and tooth mobility (Figure1).

On the panoramic radiographic, a diffuse radiolucent image close to the roots of the right first and second molars was identified(Figure2A) mimicking a possible



Figure 1 - Clinical appearance of the lesion. Large mass of soft tissue on right hard palate.

periapical lesion. Aiming to evaluate the spatial locations and the possible effects on the corresponding bone cortical, computed tomography examination was executed. CT axial images demonstrated a well-defined tumor mass isodense to muscle on the right hard palate (Figure 2B). The examination performed after the use of intravenous contrast evidenced a solid nodular image, with intravenous contrast enhancement, of lobulated outline, and partially-defined limits, located at the topography of the mucous-pharynx space, on the right hard palate.

According to the aforementioned characteristics, the diagnosis hypothesis was of Pleomorphic adenoma and Mucoepidermoid carcinoma. The incisional biopsy was carried out (Figure3) and the material sent to the anatomic-pathologic examination. For this purpose, the specimen was fixed in 10% formalin, and paraffin sections were prepared for light microscopy. The initial sections were

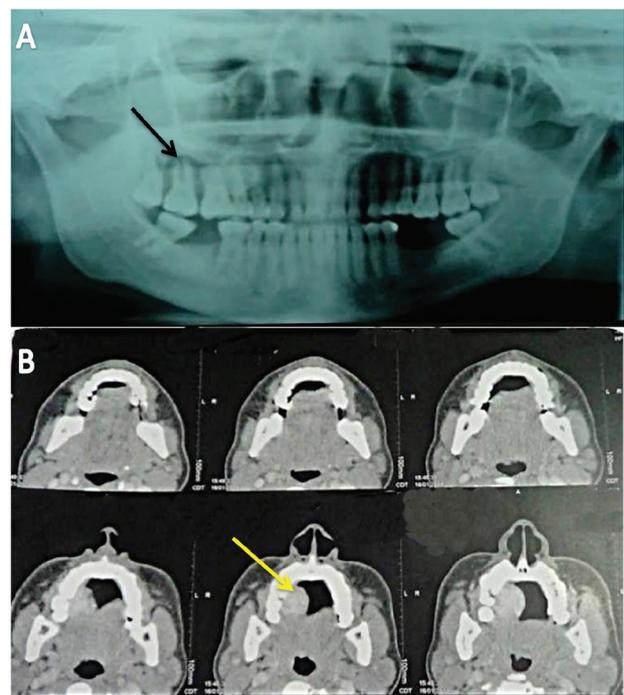


Figure 2 - Panoramic radiograph (A) and Computed tomography of the head: Solid expansive lesion with intravenous contrast enhancement, located at the topography of the mucous-pharynx space on the right hard palate(B).

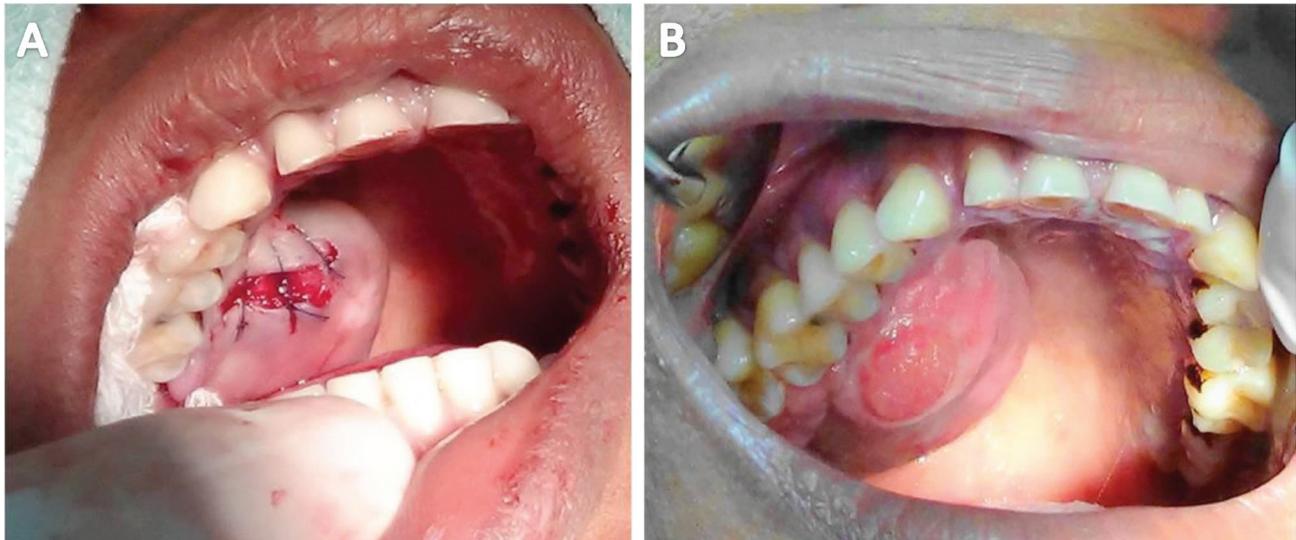


Figure 3 - Incisional biopsy (A) and clinical view after surgery (B).

stained in Hematoxylin and eosin (H & E). The material sent for histopathological examination was composed of three fragments of soft tissue measuring together 15mm x 10mm x 03mm, of irregular shape, smooth surface, brown color, and fibrous consistency.

The microscopic and histologic examinations revealed fragments of mucosa covered by parakeratinized, stratified epithelium. The lamina

propriawas composed of conjunctive tissue with proliferation of fusiform and oval cells randomly arranged, with hypocellular foci, prominent blood vessels with irregular ramifications (Figure 4A, B and C). In the immunohistochemistry analysis, the neoplastic cells showed strong and diffuse positivity to CD34 and CD99. These cells showed negativity to actin of smooth muscle, S-100, and AE1/AE3 (Figure 4D).

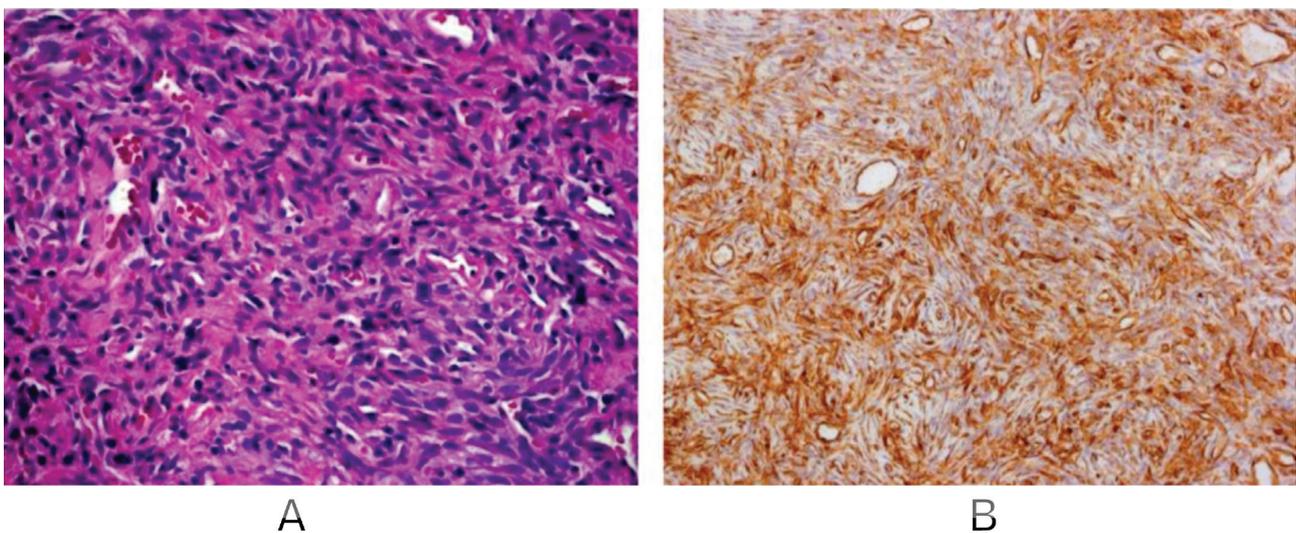


Figure 4 - Histopathological and immunohistochemical examination of the lesion. A: Microscopy showing the conjunctive tissue with fusiform and oval cells (HE staining; x20 magnification). B: Immunohistochemical staining demonstrating strong positivity to CD34 (HE staining; x20 magnification).

Based on these characteristics, the final diagnosis was of solitary fibrous tumor and the treatment of choice was surgical excision with long-term following-up. The tumor was completely removed (Figure 5). The surgical margins were free, and local signs of relapse were not observed, at 5-month post-surgical following-up appointment.

DISCUSSION

In 1942 and 1949, Stout and Murray defined the hemangiopericytoma as a neoplasia of prominent ramified vessels surrounded by pericyclic tumor cells and included it in the studies on real tumors as myofibroma/myopericytoma and conventional hemangiopericytoma, which later was accepted

as cellular variant of SFT [2,3]. Originally described by Klemperer and Rabin as a “primary pleural neoplasia” on their first reports, these lesions were considered as SFT of mesothelial origin. This point of view was gradually changed by observing that SFT might affect many other body sites [7,8].

Currently, SFT has been described in almost all human body organs: head and neck, including the nose and paranasal sinuses; nasopharynx; greater salivary glands; larynx; thyroid; skin; oral cavity; parapharyngeal space; and orbit. According to the literature, the oral cavity is the most common site of occurrence in head and neck, specifically on the oral mucosa, as seen in this case report, followed by the tongue and lower lip [9].

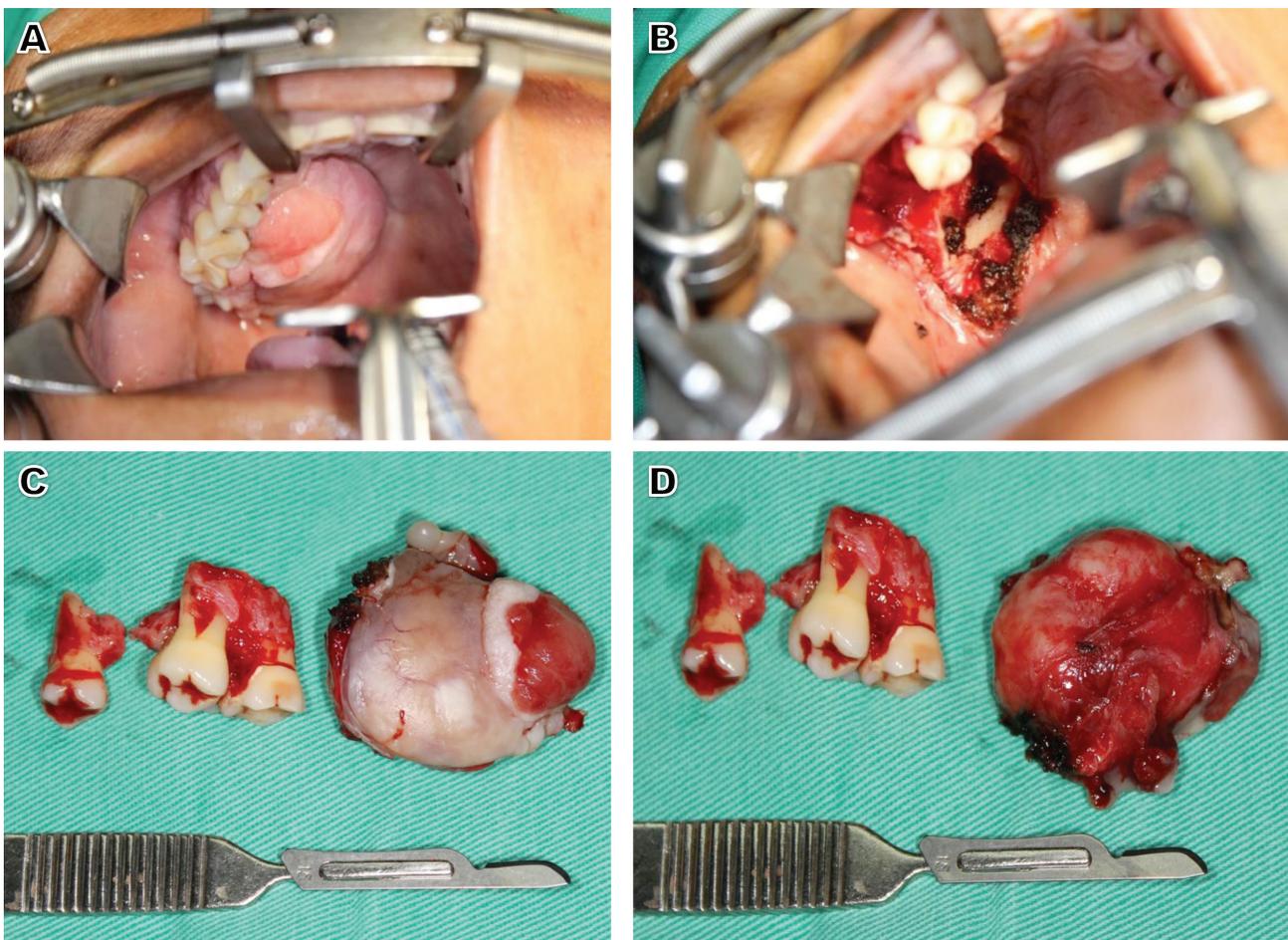


Figure 5 - Local surgical excision.

SFT association with oral cavity, especially oral mucosa, lead to the suggestion that they might be associated with trauma[10], because oral mucosa is frequently susceptible to many traumas from external or internal origin, caused by many physical, chemical, and biological agents. In this present case report, both the patient and the clinical examination did not reveal during anamnesis any information that would support such hypothesis.

Clinically, oral SFT is a well-circumscribed mass on submucosa of normal color, asymptomatic and of slow growth [2,10], which corresponded to the clinical characteristics seen in this present clinical case, except for the presence of bleeding and tooth mobility, which were atypical characteristics reported in other studies. SFT can be mistaken to be other lesions as mucocele, salivary gland tumors, lipoma, vascular malformations, leiomyoma, among others [10]. In this present clinical case, SFT had large dimensions and was located on the hard palate reflecting on the alveolar bone, and presented a fast growth after the incisional biopsy[11].

Although completely distinct images are not available, resources as the radiograph and computed tomography with window for soft tissue is of fundamental importance for the diagnosis of rare tumors because some imaging features such as the isodensity similar to that of the muscle led to the inclusion of SFT in the differential diagnosis[14]. Although not pathognomonic, the most prominent feature of SFT is that this lesion is densely reinforced by CT and MRI with the use of contrast, a sign probably justified by the presence of prominent blood vessels with irregular ramifications, similar to this case report. These features corroborate previous reports in literature. In this case report, SFT was well-defined and isodense on CT [15].

SFT diagnosis in extra pleural sites may be difficult because of its large histological spectrum. Moreover, the SFT diagnosis of a small biopsy specimen is difficult because of

the extreme intratumoral variability and close similarity of isolated parts of the soft-tissue tumors[16]. Classically, the lesions show areas with increasing of cellularity and a proliferation pattern, while others show either spread occasional mitosis or hyalinized fibrous tissue with little fusiform cells. The most common findings include areas of hypercellularity, pattern of vascularization of hemangiopericytoma, and similar corrugated neural appearance[17]. In this present case, hypocellular foci and prominent vascularization with some ramifications were present corroborating the literature.

The SFT tumor cells are immunoreactive to CD34 and variably positive to CD99, which was very evident in the immunohistochemistry performed in this present case. The anti-CD34 antibody, which recognized a glycoprotein from transmembrane cell surface found in myeloid progenitor cells, is the most consistent and reliable immunohistochemical marker for SFT. The CD99 is a membrane glycoprotein used in the diagnosis of Ewing sarcoma and peripheral primitive neuroectodermal tumors, also positive in many SFT cases. The positivity is membrane pattern, varying from area to area and from cell to cell. On the other hand, the neoplastic cells are negative to cytokeratin, S-100 protein, the antigen of epithelial membrane, actin of smooth muscle, and Factor VIII, which were the characteristics that guided the final diagnosis of this present case[1-4]. The immunohistochemical profile of this present case report is in agreement with this information.

Many features support the hypothesis that the tumor mass is characterized by zones of plenty stroma rich in collagen fibers. In 2002, Kumagai and Cols showed through immunohistochemistry, the positivity for collagen IV of SFT of parotid gland. In this case report, the reactivity to collagen II and IV was not tested [18]. Of the 49 SFT cases found in literature, 32 originated from the face or oral mucosa. The considerable healing activity of this area is necessary due to the frequent presence of

micro traumas caused by mastication. But, this intense healing activity might favor the tumor formation[1,2,17,19].

The difficulty of complete excision might be affected by the anatomical complexity of head and neck, as illustrated in some cases in literature[17]. Although radiotherapy may help in preventing relapse, few studies with short follow-up periods used it, suggesting radiotherapy as standard adjuvant therapy[19].

Most commonly, SFT is a benign tumor. Malignancy may occur and is associated with histological features of marked hypercellularity and pleomorphism, inaccurate limits, necrosis, and more than four mitoses per field, at magnification greater than 10-fold, which were not found in this present case [12]. Large sizes are associated with a more aggressive behavior, which may justify the destruction of the alveolar bone, consequently leads to tooth mobility and bleeding. Neoplasias located on the pleura, mediastinum, abdomen, pelvis and retroperitoneum tend to behave more aggressively. Metastases have been described to lungs, bones, and liver, but are very rare. At the palate, three benign cases were reported, with no histologically malignant case associated with any local or distance invasion [13]. The characteristics described in this case report are consistent with those of the previous studies in the literature.

FINAL CONSIDERATIONS

In conclusion, we reported a rare case of SFT involving the hard palate, relatively composed by hypocellular fibrous tissue foci. The CT examination showing characteristics of isodensity of the lesion similar to that of the muscle; the histopathological profile; and the immunohistochemical reaction to CD34 and CD99 were important to establish the final diagnosis. This clinical case suggests that intraoral SFTs at hard palate can be easily mistaken to be other soft-tissue lesions, such as pleomorphic adenoma and mucoepidermoid carcinoma.

CONFLICT OF INTERESTS

The authors declare that there is no conflict of interests regarding the publication of this paper.

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