Pedunculated Desmoplastic Fibroblastoma (Collagenous Fibroma) of the Oral Cavity: A Previously Unreported Clinical Presentation

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Abstract

Desmoplastic fibroblastoma is a rare, benign, soft tissue tumour. To our knowledge, six cases of Desmoplastic fibroblastoma in the oral cavity have been described so far, all with a sessile clinical configuration. Here, we describe the seventh case of oral Desmoplastic fibroblastoma, the first one presenting as a pedunculated nodule. A 56-year-old female sought care for evaluation of a nodule on the palate. No radiographic changes were noted. Excision of the lesion was performed and microscopic examination revealed a fibroblastic proliferation in the lamina propria. Immunohistochemistry showed positivity for vimentin and some positive cells for α-smooth muscle actin and factor XIIIa, confirming the diagnosis of desmoplastic fibroblastoma. This paper expands the spectrum of clinical manifestations of oral Desmoplastic fibroblastoma and demonstrates that this lesion should be included in the differential diagnosis of pedunculated intra-oral lesions.

Keywords: Case Report; Collagenous Fibroma; Desmoplastic Fibroblastoma; Oral Cavity; Vimentin; Pedunculated.

Introduction

Desmoplastic fibroblastoma (DF), also known as collagenous fibroma, is a rare, benign, soft tissue tumour. Since its characterization by Evans, fewer than 100 cases have been reported in the English literature. DF normally occurs in the subcutaneous tissue or skeletal muscle of adults. Patients in their fifth or sixth decades of life are affected more commonly. The disease is three to four times more common in males than in females. The upper (shoulder, upper arm and forearm) and lower extremities are the most common sites. Rarely, lesions may arise in the head and neck. DF is rare in the oral cavity. To our knowledge, only six cases described so far. Clinically, all previously reported cases presented as a sessile, painless, solitary, slow growing and well-circumscribed mass.

The aim of the current case is to describe the seventh case of oral DF. This case is particularly novel because it is the first report of a DF with a pedunculated clinical configuration.

Case report

A 56-year-old Caucasian female sought care at our Oral Medicine Clinic with chief complaint of an asymptomatic growth in the palate, with five years of evolution. The patient reported the lesion was initially small, but gradually increased in size over time. She disclosed a 30-year smoking history. Medical, familial and socioeconomic histories were non-contributory. Intra-oral examination demonstrated a 35mm, non-ulcerated, smooth-surfaced, normal coloured, well-defined, nodule on the palate (Fig 1A). The lesion was fibrous and painless on palpation; at this stage, it was also possible to observe that the lesion was pedunculated and that its insertion base was located between teeth number #13 and #12 (Fig 1B). The patient wore a removable partial denture, which was, however, unrelated to the lesion. In fact, as the lesion grew, it actually adapted to the denture (Fig 1A).

Panoramic and occlusal radiographs revealed no osseous abnormalities. Based on a clinical hypothesis of a benign mesenchymal lesion, an excisional biopsy was performed (Fig 1C). Upon removal of the lesion, it was possible to more clearly observe the lesion’s insertion region (Fig 1C and 1D). The specimen was fixed with 10% formaldehyde and submitted for histopathological analysis. Microscopic
examination revealed a fragment of oral mucosa with a non-encapsulated, fibroblastic proliferation in the underlying lamina propria, characterized by paucicellularity within a highly collagenous matrix (Fig 2A and 2B). At high power magnification, fibroblasts presented spindle to stellate shapes, with large and rounded nuclei exhibiting thin chromatin and small nucleoli (Figure 2C). Binucleated (Fig 2D) or multinucleated (Fig 2E) fibroblasts were also observed. Blood vessels were few and small, and neither mitosis nor necrosis was present. In the deep region of the specimen, lesional tissue trapped adipocytes (Fig 2F). Immunohistochemical evaluation revealed intense positivity for vimentin (Fig 3A) and some positive cells for α-smooth muscle actin (Fig 3B) or factor XIIIa. The cells were immunonegative for desmin, HHF-35, S100, CD34, CD68 and AE1/AE3. The final diagnosis was desmoplastic fibroblastoma. The patient is currently being followed-up and no recurrence has been observed after 16 months.

Figure 1: Clinical and gross aspects. (A) Intra-oral examination revealed a 35-mm, non-ulcerated, smooth-surfaced, normal coloured, well-defined nodule on the palate; (B) Upon elevation of the lesion with a tongue blade, its insertion base (arrow) was located between teeth 13 and 12; (C) Immediate post-operatory view, showing the lesion’s insertion base (arrow); and (D) Gross aspect of specimen, again indicating site of insertion.

Discussion
The current case presents clinical, histopathological and immunohistochemical features of DF, a rare benign soft tissue tumour. DF has been reported in several locations, such as the arm, posterior portion of the neck, and upper back; however, the lesion is rare in the oral cavity, with the current case being, to our knowledge, the seventh report in the literature. Oral DF is more common in females and the mean age of patients is 54 years. The palate is the most frequently affected site. Our patient was a 56-year-old female, with a palatal lesion, in agreement with the literature. Clinically, oral DF appears as a painless, solitary, slow growing, well-circumscribed mass. Typically, no precipitating trauma is present. Regarding colour, DF may be yellowish, reddish, or similar to the normal adjacent mucosa. The lesion may grow to over 1cm and cause difficulty in speaking, eating, and closing of the mouth. In the current case, the lesion was adapted to the patient’s removable partial denture, but unrelated to it. The consistency is described as firm to elastic. The current case is in accordance with these clinical aspects, except that all cases of DF reported to date have been described as sessile masses, whereas the current case presented as a pedunculated mass. This unique clinical presentation makes our case the first of its kind.

Clinical differential diagnoses of oral DF, when in its sessile form, include schwannoma, neurofibroma, myofibroma, and lipoma. More remote possibilities include benign fibrous histiocytoma, granular cell tumour and leiomyoma. Given the
tendency to affect the palate, salivary gland tumours, such as pleomorphic adenoma, may be considered in the differential diagnosis of sessile DF.\textsuperscript{9} Considering the pedunculated clinical presentation, clinical differential diagnoses are more restricted; remote possibilities include giant cell fibroma, leaf-like denture fibroma and pyogenic granuloma. Absence of precipitating trauma is an important factor to differentiate DF from the above-mentioned lesions.

Figure 2: Microscopic features. (A) Oral mucosa fragment with a non-encapsulated, fibroblastic proliferation in the underlying lamina propria (Haematoxylin-eosin, x50 original magnification); (B) The lesion was characterized by paucicellularity within a highly collagenous matrix (Haematoxylin-eosin, x200 original magnification); (C) Fibroblasts presented spindle to stellate shapes, with large and round nuclei exhibiting thin chromatin and small nucleoli (Haematoxylin-eosin, x400 original magnification); (D) Binucleated; (E) Multinucleated fibroblasts were also observed (Haematoxylin-eosin, x400 original magnification); and (F) Lesional tissue trapped adipocytes in the deep area of the specimen (Haematoxylin-eosin, x200 original magnification).

Histopathological examination is required to achieve the diagnosis of DF. Microscopically, the tumour is composed of stellate or spindle shaped cells, with a round or elongated nucleus, delicate chromatin and small nucleoli.\textsuperscript{4,10} Binucleated cells and
few multinucleated cells are also occasionally observed. The cells are embedded in a prominent collagenous background with inconspicuous blood vessels. DF may infiltrate both adipose tissue and skeletal muscle and entrap nerves. In our case, skeletal muscle involvement was not observed. This was expected, since the lesion was located in the hard palate, an area in which muscle fibres are absent. Although DF is a relatively paucicellular tumour, some increase in cellularity is frequently observed at the periphery of the lesion. However, mitotic figures are rare or absent, and necrosis is not observed. Focal calcifications are also uncommon findings.

Figure 3: Immunohistochemistry of desmoplastic fibroblastoma (collagenous fibroma) demonstrated intense positivity for vimentin (A) and some cells positive for α-smooth muscle actin (B) (Streptavidin-biotin, x400 original magnification).

The microscopic differential diagnosis includes mainly inflammatory fibrous hyperplasia and giant cell fibroma. Less common conditions to be considered include solitary fibrous tumour, juvenile hyaline fibromatosis, calcifying fibrous pseudotumour, low grade fibromyxoid sarcoma, elastofibroma and long standing nodular fasciitis. Although not mandatory, immunohistochemistry can aid in the exclusion of other diagnoses. DF is strongly and diffusely positive to vimentin and negative to S-100, CD34, desmin, cytokeratins, epithelial membrane antigen, and CD68. The majority of studies, as well as the current case, show focal positivity for α-smooth muscle actin and factor XIIIa. In contrast, the giant cell fibroma is negative for α-smooth muscle actin and rarely positive for factor XIIIa. Solitary fibrous tumours are positive for CD34 and negative for α-smooth muscle actin. Juvenile hyaline fibromatosis are negative for actins and show CD68 positive macrophages between the spindle cells. Calcifying fibrous pseudotumor are diffusely positive for factor XIIIa and negative for smooth muscle actin, muscle-specific actin, and CD34. Low grade fibromyxoid sarcoma shows positive staining with vimentin, but no immunoreactivity for keratin, desmin, muscle specific actin, S100 protein, CD34 or CD31. Elastofibroma is positive for vimentin, elastic stains and CD34 in spindle cells, but negative for S100, desmin, smooth muscle actin, p53. Nodular fasciitis typically stains for smooth muscle actin and muscle-specific actin, whereas cytokeratin and S-100 are consistently negative.
The immunohistochemical profiling of DF supports a myofibroblastic origin. Indeed, Nielsen et al. (1996)\textsuperscript{13} demonstrated that the cells have ultrastructural features characteristic of fibroblasts and myofibroblasts. Nonetheless, the pathogenesis of DF is still unclear and there is debate whether it represents a peculiar reactive process or a neoplasm.\textsuperscript{10,12} Evans (1995)\textsuperscript{5} and Nielsen et al. (1996)\textsuperscript{21} stated that DF represents a neoplasm due to absence of a clinical inciting event or specific microscopic cause. Cytogenetic studies suggested a translocation t(2;11)(q31;q12) to constitute a recurrent and non-random event in DF, providing further evidence for a neoplastic disorder.\textsuperscript{11} The current treatment of choice for DF is total surgical excision, and the prognosis is favourable. No patient to date has experienced tumour recurrence or metastasis in several follow-up studies.\textsuperscript{10,21,25} Likewise, our patient was treated by complete surgical excision, with no recurrence or metastasis during a 14-month follow-up period.

**Conclusion**

In conclusion, we described the seventh case of oral DF in the literature, the first to present clinically as a pedunculated protruding nodule with a stalk. This paper expands the spectrum of clinical manifestations of oral DF and demonstrates that this lesion should be included in the differential diagnosis of pedunculated intraoral lesions.

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Pedunculated Desmoplastic Fibroblastoma (Collagenous Fibroma)


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