A case of oral dermatofibrosarcoma protuberans relapsing in the skin

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Summary
Dermatofibrosarcoma protuberans comprises roughly 0.1% of all malignant tumors and approximately 1 to 2 percent of all soft tissue sarcomas. Slowly growing and locally aggressive, dermatofibrosarcoma protuberans can reach an enormous size, underlying in fascia and musculoskeletal structures including bone and also becoming superficial. It is characterized by high rates of local recurrence and low risk of metastasis. Dermatofibrosarcoma protuberans may involve the head and neck area in 10-15% of cases, but it is rarely described in the oral mucosa. Here, we describe a case of oral dermatofibrosarcoma protuberans relapsing in the skin after 18 years. The lineage of the cell type comprising dermatofibrosarcoma protuberans remains a matter of controversy, but this case is consistent with the opinion that the progenitor cell is not only present in the skin but also in the submucosal tissue of the oral cavity.

KEY WORDS: dermatofibrosarcoma protuberans; skin cancer.

Introduction
Dermatofibrosarcoma protuberans (DFSP) is an uncommon infiltrative dermal spindle cell tumor, which represents a cutaneous-origin low-grade sarcoma (1). This soft tissue neoplasm, in many cases, affects young people or adults with a previous long preoperative history due to the slowly and indolently growing (over years) of the lesion (2). DFSP generally develops as a multifocal reddish-blue plaque on the trunk. Occurrence at sites of previous trauma has been reported (3), whereas mucosal involvement is an exceptional circumstance. DFSP is characterized by high rates of local recurrence, for its extensive subclinical growth, a low risk of metastasis and a favorable prognosis (4).

We report an unusual advanced case of DFSP in an elderly patient, localized in the oral cavity, arising almost 18 years before our final diagnosis.

Case report
An 80-year-old male attended our dermatology outpatient clinic with rapidly growing, nodules on the nasolabial fold of his left cheek appeared about 1 year before. Physical examination revealed two nodules, a larger lesion 2 cm in diameter and a smaller one 0.5 cm in diameter (Figure 1 A, B). Under these two nodules an area of induration was present which extended into the lower internal part of the cheek. The biopsy carried out on both the skin and the mucosa, showed a dense proliferation of spindle cells, arranged in a storiform pattern, strongly positive for CD34 and negative for S100, CD68, CD31, CK AE1/AE3 (Figure 2 A-D) and a diagnosis of DFSP was made. The patient reported that 10 years before an excisional biopsy had been performed for a long standing (8 years) nodular lesion of the oral cavity. The histological features were consistent with the diagnosis of lipoma. Two years later an ultrasound scan of the cheek revealed a diffuse ill-defined subcutaneous thickening. No other investigations were performed before the development of the two nodular lesions. We decided to review the original slides and a diagnosis of DFSP was made. The patient reported that 10 years before an excisional biopsy had been performed for a long standing (8 years) nodular lesion of the oral cavity. The histological features were consistent with the diagnosis of lipoma. Two years later an ultrasound scan of the cheek revealed a diffuse ill-defined subcutaneous thickening. No other investigations were performed before the development of the two nodular lesions. We decided to review the original slides and a diagnosis of DFSP was made. The patient was referred for reconstructive surgical procedures; a wide surgical excision was performed, with a full-thickness resection of the buccal mucosa and closure of the buccinators by upper hinge. At present the patient is in good health and no recurrences have been observed.

Discussion
Differential diagnosis of DFSP includes several conditions, notably dermatofibroma, dermatofibrosarcoma, fibrosarcoma, angiosarcoma, invasive spindle cell squamous cell carcinoma, spindle cell melanoma, malignant
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Figure 1, A, B - A 2 cm diameter, well-defined, smooth exophytic nodule and a smaller one on the nasolabial fold (A). A plaque of increased consistency can be observed under the nodules (B).

Figure 2, A-D - Dense proliferation of packed monomorphus spindle cells arranged in a storiform pattern in the derma and hypoderma of the cheek (A) and of the oral mucosa (B) (hematoxylin-eosin. Original magnification x 20). (C) Spindle cells encircle old individual adipocytes ("string of pearls pattern") and the entire fat lobules ("honeycomb pattern") (hematoxylin-eosin. Original magnification x 40). (D) Spindle cells with strong, diffuse CD34 immunoreactivity (Original magnification x 20).

fibrous histiocytoma (also called pleomorphic sarcoma), smooth muscle tumor and leiomyosarcoma (2,5,6). In our case the initial diagnosis was lipoma of the oral cavity. The subsequent revision of the histological slides was consistent with the diagnosis of oral DFSP. The histological pattern of the original lesion simulated a spindle cell lipoma, which is histologically and immunohistochemically similar to DFSP. Spindle cell lipoma is com-
posed of mature fat and bland spindle cells, strongly positive for CD34 and negative for desmin, which typically occurs on neck and head in predominantly elderly males. To the best of our knowledge only two cases of oral DF-SP have been described in literature (7, 8). Our case is the first demonstration of an oral DFSP relapsing in the skin. It is unlikely that the patient presented two independent DFSP. The area involved was the same and considering the slow growth of DFSP and the amount of time between the oral localization and the skin relapse, the clinical history is consistent with this hypothesis.

Even though 10 to 15% of DFSP involve the head and neck, this neoplasm is exceptional in the oral cavity. Moreover, the location on the head and neck is associated with a greater risk of morbidity and local recurrence, but the course of mucosal DFSP seems less aggressive. The lineage of the cell type comprising DFSP remains a matter of controversy, but this case is consistent with the view of Meehan et al., that the progenitor cell is not only present in the skin but also in the submucosal tissue of the oral cavity (7).

References