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CASE REPORT

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Myofibroma of the upper lip free edge mimicking a hyperkeratotic plaque

Abstract:

Myofibroma is a benign, non-encapsulated neoplasm that predominantly affects the skin and subcutaneous tissue of the head and neck. It has been documented in various regions of the oral mucosa and jawbones. However, cases involving the free edge of the lips have not been previously reported. We present a case of myofibroma manifesting as a yellowish-white plaque on the free edge of the upper lip, initially misdiagnosed as a hyperkeratotic plaque. Consequently, myofibromas may also present as flat lesions, rather than as swellings or tumors, potentially mimicking an epithelial disorder. The lesion was completely excised, and after a four-month follow-up, no recurrence was observed.

Keywords: Lip neoplasms; Myofibroma; Oral pathology; Benign tumor.

INTRODUCTION

Myofibroma (MF) is a benign neoplasm of mesenchymal origin composed of myofibroblasts, currently reclassified as a pericytic tumor by the WHO[1,2]. Morphologically, it presents as interlacing bundles of spindle-shaped cells with elongated, oval to wavy nuclei that have blunt ends, consistently associated with blood vessels. The cytoplasm is eosinophilic, indicating differentiation toward smooth muscle and fibroblasts. Functionally, these cells exhibit characteristics of both fibroblasts and smooth muscle cells[3]. Immunohistochemically, tumor cells are positive for smooth muscle actin and vimentin but negative for S-100 protein, desmin, H-caldesmon, and Ki67[4,5].

Statement of Clinical Significance
The clinical significance of this case lies in the atypical presentation of an upper lip myofibroma, which manifested as a hyperkeratotic plaque, potentially leading to a misdiagnosis of a potentially malignant lesion. This report emphasizes the importance of differential diagnosis in oral lesions with uncommon clinical features. The correct diagnosis, based on histopathological and immunohistochemical examination, enabled appropriate treatment and complete resolution without the need for additional invasive procedures.

Since myofibroblasts produce collagen and possess contractile capacity, they are considered to play a crucial role in wound healing[6,7]. The causes of myofibroblastic proliferation are unknown[8]. Myofibroma (MF) can present in three forms: solitary, multicentric, with or without visceral involvement. The tumor most commonly occurs in the skin and subcutaneous tissue, particularly in the head and neck, followed by the upper limbs and trunk[2,9,10].

In the oral cavity, cases have been reported in the tongue, palate, gums, retromolar area, floor of the mouth, buccal mucosa, alveolar ridge, and intraosseously in the maxilla and mandible, often being mistaken for odontogenic cysts and tumors[7,11-24]. They have been described as painless nodules, ulcerated and non-ulcerated exophytic masses, pyogenic granulomas, and irritative fibromas. Their size can range from one to several centimeters. Occasionally, they have been confused with malignant neoplasms[24-26]. In most cases, the diagnosis is made after a benign lesion is sent for histological examination[20,27,28].

We report a case of myofibroma in a 36-year-old female that presented as a yellowish-white plaque, rather

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than the common papule or tumor usually associated with this neoplasm. Furthermore, the lesion was located on the free edge of the upper lip, an extremely rare site for this tumor. Clinically, it was initially misdiagnosed as a hyperkeratotic plaque.

CASE REPORT

A 36-year-old female was referred to the Oral Medicine and Oral Pathology Unit of the Faculty of Stomatology at the Universidad Peruana Cayetano Heredia with a 4-month history of a lesion on the free edge of the upper lip. She reported a painless yellowish-white plaque, round-shaped with irregular borders and a rough texture, approximately 1 cm in diameter (Figure 1).

Her medical history was unremarkable, and she denied smoking or alcohol consumption.

Clinically, the differential diagnosis included a hyperkeratotic plaque, a benign tumor of vascular components, and a reactive connective tissue papule. Since all these proposed diagnoses referred to benign lesions, the lesion was excised completely and sent for histological examination.

Microscopic examination revealed a non-encapsulated tumor within the papillary and reticular layers of the lamina propria. It was composed of spindle-shaped and rounded cells, organized in fascicles around blood vessels with prominent endothelial cells. The cytoplasm of the cells was eosinophilic, and they exhibited blunt-ended nuclei. In some areas, the cells showed spiral-shaped proliferation. The surface epithelium appeared slightly thickened due to hyperorthokeratosis. No inflammatory reaction was observed. Surrounding the tumor cells was a collagen matrix, as demonstrated by Masson's trichrome stain (Figure 2).

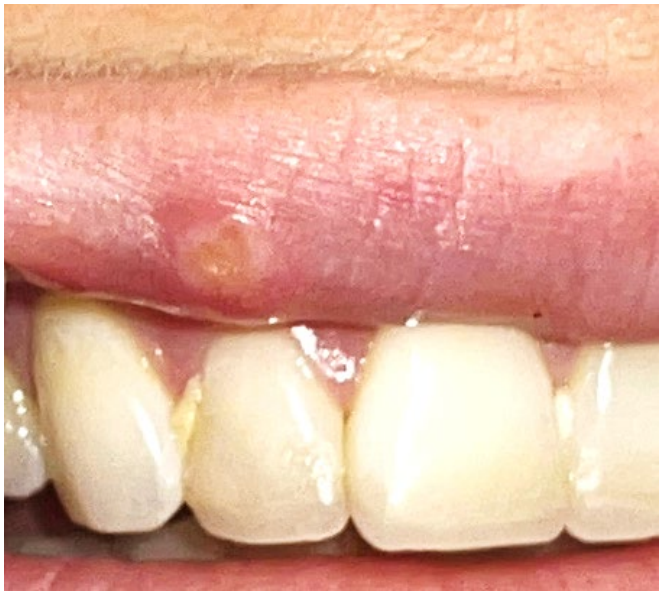

Figure 1. A round-shaped, yellowish-white plaque located on the free edge of the upper lip.

Given the shape of the tumor cells, it was necessary to rule out a peripheral nerve or muscle origin. Therefore, immunohistochemical staining with α-SMA, S-100 were performed. Additionally, Ki67 staining was requested to assess the degree of cell proliferation. Immunohistochemical staining was strongly positive for α-SMA (Figure 3), while S-100 and Ki67 were negative. Based on these histological findings, a diagnosis of benign myofibroma was made.

The treatment consisted of the complete removal of the lesion. At the four-month follow-up, no recurrence was observed.

DISCUSSION

Myofibromas are benign tumors composed of myofibroblasts. According to the WHO, they are perivascular neoplasms originating from pericytes. They are considered rare in the oral cavity and can resemble a variety of lesions, such as irritational fibromas, fibrous hyperplasias, or pyogenic granulomas[2,7,24–27]. The most common location in the mouth is the gingiva, followed by the tongue and buccal mucosa[4,11,12]. The lip is a less common location, with fewer than 30 cases currently reported in the literature[5,7,16–23], only two of which mention the upper lip[7,19]. This case is likely the first reported instance of involvement on the free edge of the upper lip.

Clinically, they appear as asymptomatic papules or masses, sometimes with superficial ulceration, ranging in size from one to two centimeters. Larger cases have been reported in the jaws, which can radiographically be confused with odontogenic cysts and tumors[4,9,11,13–15,29]. Epithelial plaque-like lesions have not been reported on the upper lip.

The clinical appearance and location of the reported case were quite peculiar. It was situated on the free edge of the upper lip and was clinically mistaken for a hyperkeratotic plaque. However, hyperkeratotic lesions typically occur on the free edge of the lower lip, primarily in male patients with a history of smoking or those exposed to sunlight[30,31]. Therefore, this case does not fit that diagnosis.

The clinical differential diagnosis included a hyperkeratotic plaque, a benign vascular tumor, and a

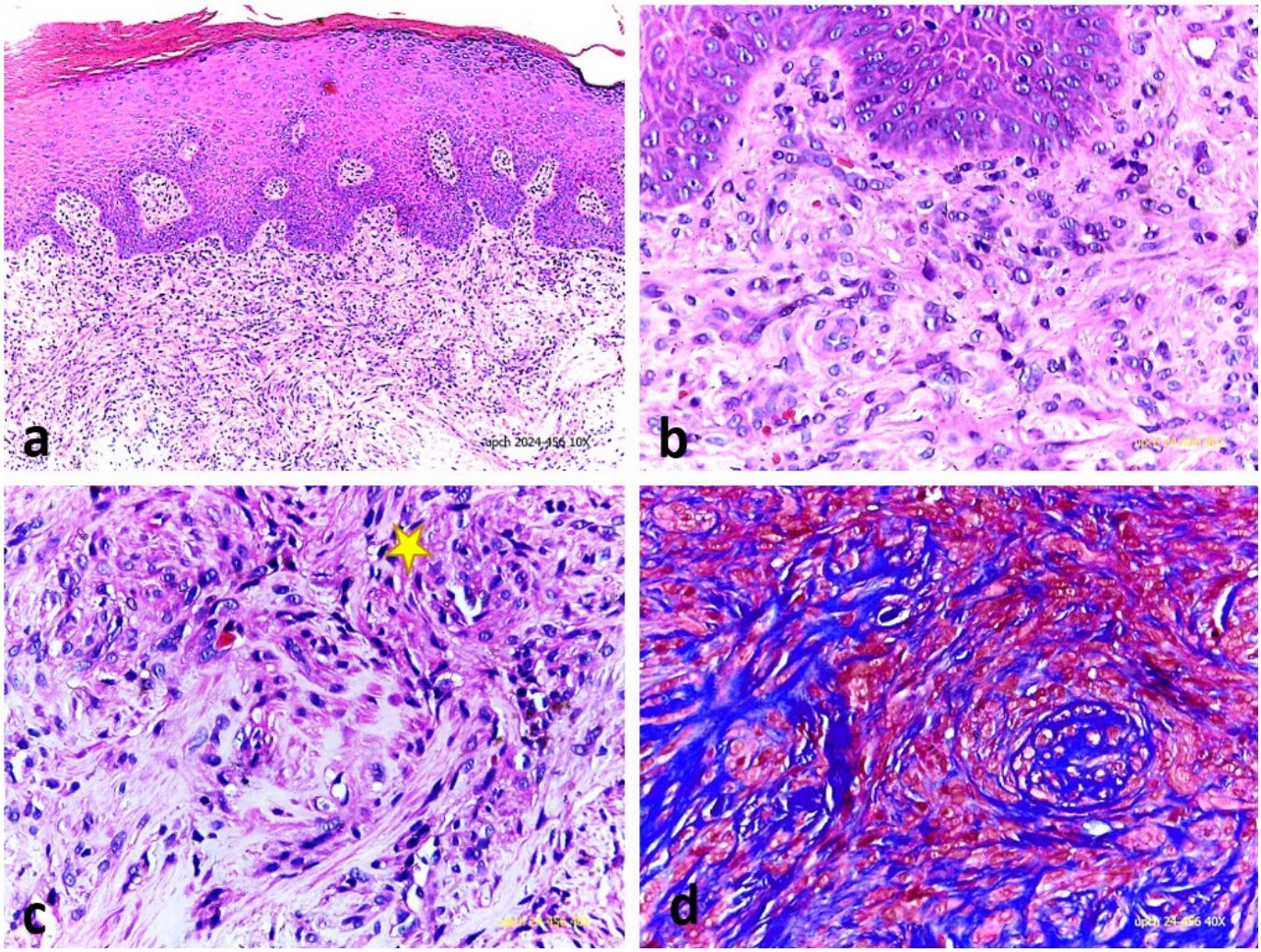

Figure 2. a) Non-encapsulated tumor composed of spindle-shaped and round cells arranged in fascicles within the reticular and papillary layers of the lamina propria. The epithelium showed hyperorthokeratosis. b) Tumor cells at high magnification. Endothelial cells of blood vessels were prominent. c) Spindle-shaped and round cells exhibited eosinophilic cytoplasm with blunt-ended nuclei. In addition to the fascicular arrangement, some areas showed myofibroblasts demonstrating a whorled pattern. A collagenous matrix surrounded the cells (★). d) Trichrome stain depicted collagen in blue, while nuclei and myofibers appeared red.

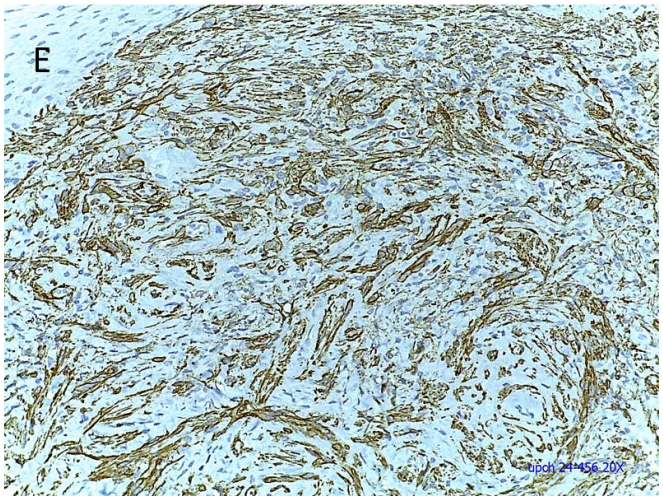

Figure 3. α-SMA is intensely positive in tumor cells (original magnification, 20X). Epithelium (E).

reactive connective tissue papule. As with other cases reported in the literature[5,18,24], the diagnosis of myofibroma was established after histological examination.

In the reported case, the histological study showed a non-encapsulated proliferation of spindle-shaped and rounded cells, distributed around blood vessels and occupying the papillary and reticular layers of the free edge of the upper lip. Essentially, a fusocellular tumor, such as neurofibroma, fibrous solitary tumor, or benign muscle tumor, was considered.

The definitive diagnosis was confirmed through immunohistochemical studies, which demonstrated α-SMA positivity in all tumor cells, while S-100 and Ki67 were negative. These results allowed for the final diagnosis of benign myofibroma, as noted in the literature[2,4,5,11,20,27]. In our opinion, when immunohistochemical

studies are not available, Masson's trichrome stain can be helpful in diagnosing this tumor[1,11,20,28].

When a lesion is composed solely of collagen, no red color will be seen in the fibers with this stain. This indicates that no myofibroblasts are present, and the collagen fibers will appear blue, with the nuclei appearing red[20]. In contrast, the fibers and nuclei of myofibroblasts will appear red (Figure 2d). There are few cases in which molecular studies have been performed, and the results are not conclusive[2,32]. In our case, no molecular studies were conducted.

The epithelium showed hyperorthokeratosis, which could explain the plaque-like appearance of the lesion that led to the incorrect clinical diagnosis of hyperkeratosis (Figure 2). Since recurrent cases have been described after surgical treatment[9,11], the patient has been advised to continue follow-up. Regarding the etiology of myofibromas, it is unknown. In one reported case, mechanical trauma has been suggested[33]. In the present case, no association with any type of mechanical irritation was found.

CONCLUSION

In conclusion, we report a case of myofibroma that developed in a very rare location, with a clinical appearance that was confused with a plaque-like lesion originating in the epithelium of the free edge of the upper lip in a patient without harmful habits. Since the lesion is a non-encapsulated tumor, it needs to be followed up for an extended period. Therefore, the absence of recurrence after a four-month follow-up does not mean that the patient is free from recurrence of the tumor.

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AUTHORS' CONTRIBUTIONS

WADA: investigation, methodology, supervision, validation, visualization, writing – original draft, writing – review & editing. VMAD: conceptualization, validation, visualization. LHMV: data curation, formal analysis. KBTS: conceptualization, funding acquisition, investigation, methodology, project administration, resources, software, supervision, validation, visualization, writing – original draft, writing – review & editing.

CONFLICT OF INTEREST STATEMENT

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Ethics approval: This case report was conducted independently without review by an ethics committee, as it did not involve an intervention or clinical trial. Confidentiality was ensured, and the patient's consent was obtained.

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