

Up-to Date Review And Case Report

Oral nodular fasciitis – A case report with a diagnostic schema

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Abstract – Introduction: The spectrum of myofibroblastic lesions of the oral cavity ranges from reactive to benign to malignant lesions with overlapping histopathologic and immunohistologic characteristics posing a diagnostic dilemma. **Observation:** A 30-year-old male presented with a spontaneous swelling over the right lower buccal gingiva giving a clinical suspicion of a benign mesenchymal tumor. The lesion presented with a varied biphasic microscopic appearance that posed as a challenge for diagnosis. **Commentaries:** The incisional biopsy of the lesion showed a highly collagenous stroma with spindle-shaped cells, while the excision biopsy revealed myxoid and hyalinized stroma. A panel of markers comprising of SMA (smooth muscle actin), CD-34, β -Catenin, and Alcian blue stain was employed to arrive at a diagnosis. **Conclusion:** Most myofibroblastic lesions present with diverse histological appearance which warrants a thorough assessment of the cellular and stromal components for an accurate diagnosis.

Introduction

Nodular fasciitis (NF) is a benign reactive soft tissue lesion that was first described by Konwaler in 1955 as a subcutaneous pseudosarcomatous fibromatosis [1]. NF most commonly present in the extremities, trunk, head and neck region. The incidence of NF in the oral cavity is very rare and it occurs mostly in the buccal mucosa, alveolar mucosa, tongue and upper lip [2–4]. Nodular fasciitis occurs equally among both males and females in the fourth and fifth decades of life. It presents as a rapidly growing mass that can grow at a rate of approximately 4 cms in one month. It is firm and fixed on palpation and the surface of the growth may be ulcerated [3,5].

The pathogenesis of NF is unknown, although trauma followed by inflammation is implicated as a causative factor [6]. Although NF is a benign reactive lesion, due to its rapid growth, high cellularity and high mitotic rate it is often misdiagnosed as a sarcoma [7]. NF has to be differentiated from schwannoma, myofibroma, solitary fibrous tumor, fibrosarcoma, leiomyosarcoma, neurofibroma and malignant fibrous histiocytoma [4,6,8]. The positivity of tumor cells for

smooth muscle actin (α -SMA), muscle specific actin (HHF-35) and vimentin has established the myofibroblastic origin of NF [9].

Here, we present a case of NF that occurred on the alveolar mucosa, a rare site for the lesion. The lesion presented with a biphasic histological pattern which posed as a challenge for its diagnosis. The histochemical and immunohistochemical analysis that was carried out to arrive at the diagnosis is described.

Observation

A 30-year-old male patient visited the dental clinic with a complaint of a growth in the right lower teeth region since 3 months, which gradually increased in size. There was no history of pain, pus discharge or mobility of teeth. The medical history was otherwise not remarkable.

On clinical examination, the lesion presented as a lobulated growth measuring approximately 3 × 3 cm in the buccal vestibule in relation to the right mandibular second premolar, first molar and second molar (Fig. 1). The lesion was pink in color, with well-defined margins, was firm in consistency and mildly tender on palpation. There was no evidence of induration or discharge from the lesion. Based on the clinical

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Fig. 1. A 3 × 3 cm nodular growth on the right mandibular alveolus.

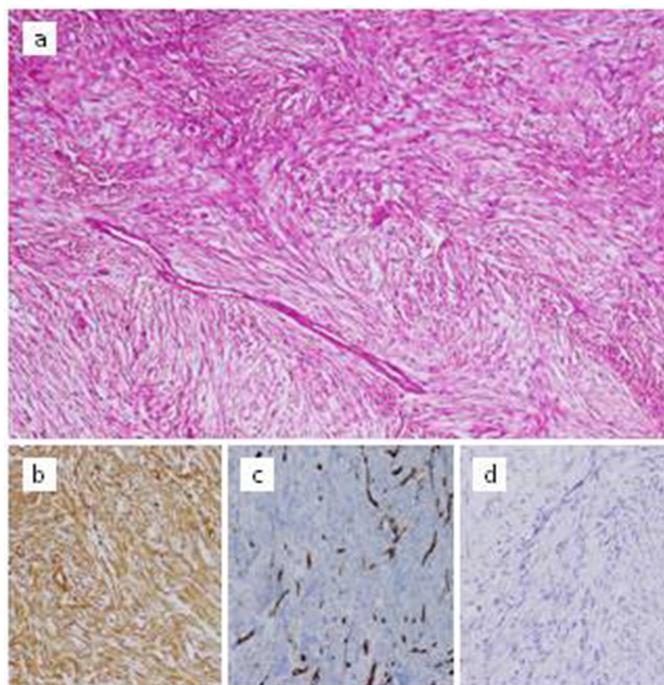


Fig. 2. Photomicrograph showing a) A collagenous stroma comprising of numerous proliferating spindle cells arranged in a biphasic pattern (H&E 4X), (b) Spindle cells showing cytoplasmic expression of smooth muscle actin (IHC-10X), (c) CD 34 expression present in endothelial cells but absent in spindle cells (IHC-10X), (d) Spindle-shaped cells lacking the expression of β -Catenin (IHC-10X).

presentation, a provisional diagnosis of benign mesenchymal tumor such as fibroma, irritational fibroma and peripheral giant cell granuloma was proffered. An incisional biopsy of the soft tissue mass was obtained for histopathological examination.

The hematoxylin and eosin stained sections revealed a highly cellular and collagenous stroma comprising spindle-shaped cells arranged in a storiform pattern (Fig. 2a). The spindle-shaped cells were elongated with an oval nuclei and eosinophilic cytoplasm. The connective tissue stroma comprised of plump fibroblasts, fibrocytes, variable number of blood vessels, and areas of hyalinization. Chronic inflammatory cells were seen in the perivascularly.

On Immunohistochemical analysis, the cytoplasm of the spindle cells expressed SMA (clone IA4, Dako) (Fig. 2b) but did not express CD-34 (clone QBE 10, Dako) (Fig. 2c) or β -Catenin (EP 35, Pathnsitu) (Fig. 2d). The findings were in favor of solitary myofibroma. The entire lesion was subsequently excised.

The hematoxylin and eosin stained tissue section of the operative piece showed immature-appearing myofibroblasts admixed with myxomatous and hyalinized areas. The cells had an oval pale staining nuclei with prominent nucleoli (Figs. 3a and 3b). The intervening matrix was rich in mucopolysacchrides which stained readily with Alcian blue (Fig. 3c). Based on the histochemical and immunohistochemical profile of the lesional

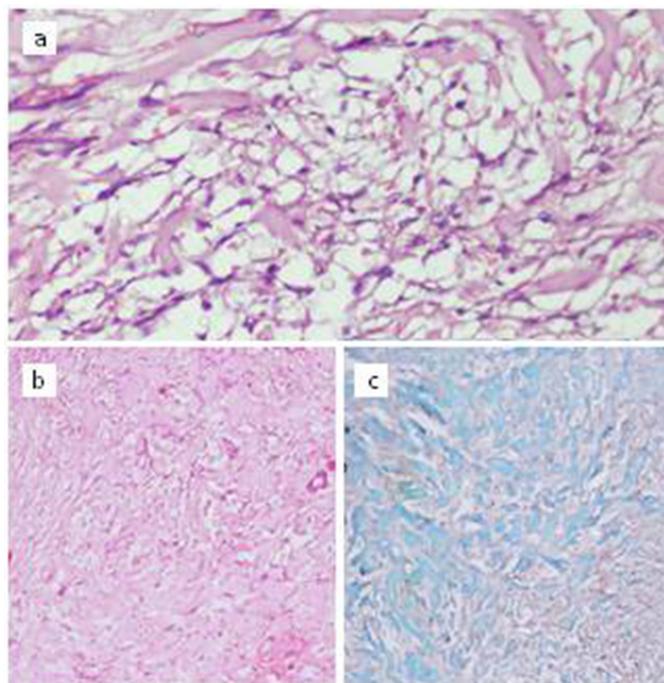


Fig. 3. Photomicrograph showing a) Fascicles of proliferating spindle cells in a hyalinized stroma with extravasated RBCs and cleft-like spaces (H&E-20X), (b) Hyalinized stroma (H&E 10X) and (c) stroma rich in mucopolysaccharide (Alcian blue stain - 10X).

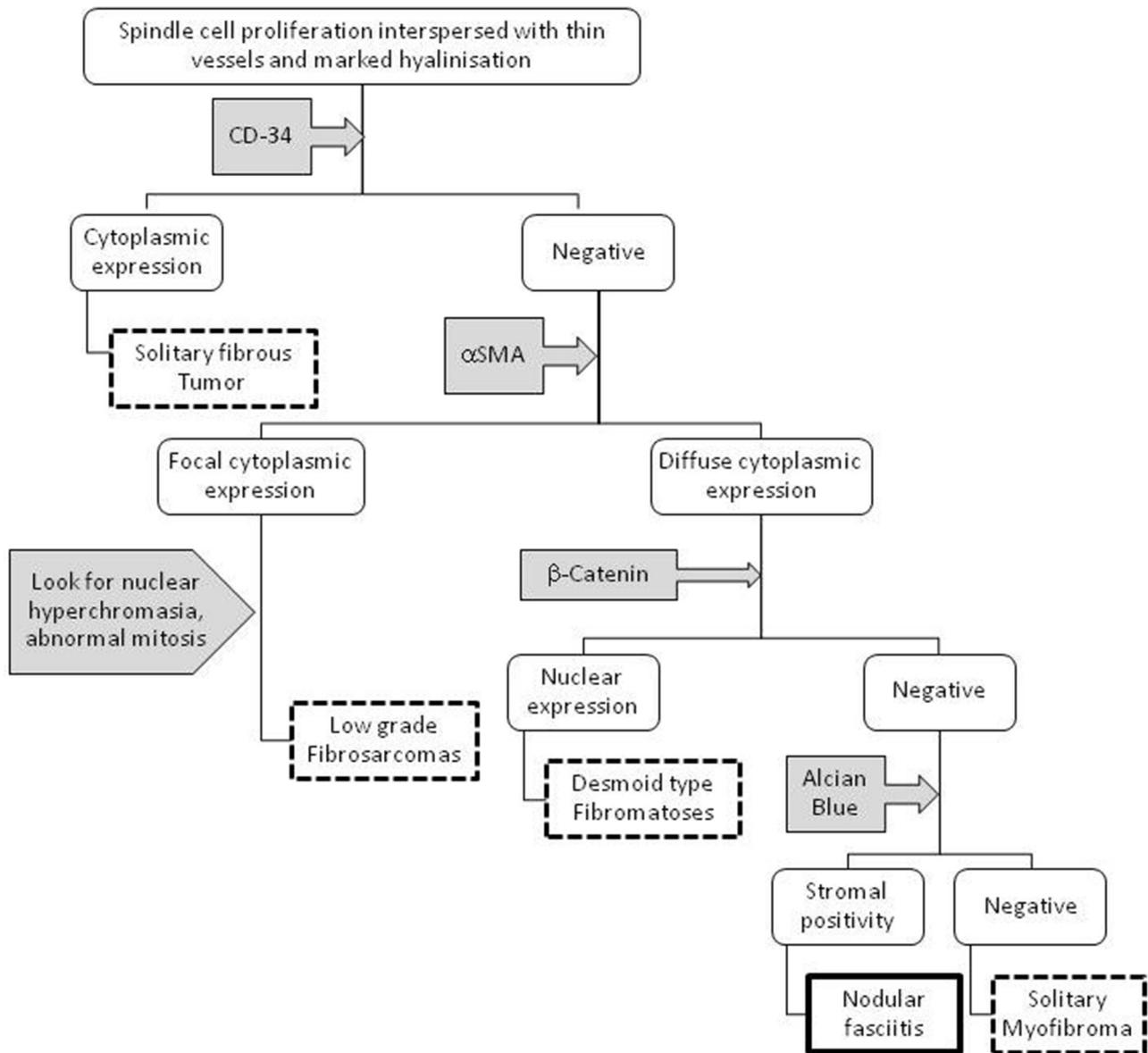


Fig. 4. A decision tree depicting the diagnostic approach.

tissue, a final diagnosis of nodular fasciitis was given (Fig. 4). The patient is on regular follow-up and there is no sign of recurrence since the last 8 months.

Commentary

Nodular fasciitis is a benign reactive lesion of the fibrous connective tissue with proliferation of fibroblasts and myofibroblasts arranged in short fascicles [5]. This benign reactive lesion is said to arise from the fascia and lesion extends into the septae of the subcutaneous fat [10,11].

Allen *et al.* had described four typical histological features of NF, numerous spindle-shaped fibroblasts arranged in whorled fascicles, small clefts separating the fibroblasts, scattered extravasated red blood cells and a mucoid interstitial ground

substance [12]. The spindle cells exhibiting mitotic activity, areas of myxoid degeneration, fibrosis and hyalinization, and the presence of giant cells are additional features of NF [13,14]. The myxoid stroma gives rise to a “feathery pattern” in early lesions and with time the lesion gets progressively hyalinised [15]. The histological features that were seen in the present case also included spindle-shaped cells, areas of hyalinization, areas of myxoid degeneration and a mucoid stroma.

Due to the high proliferative capacity of the nodular fasciitis, it has to be differentiated from malignant lesions such as myxofibrosarcoma, low-grade fibromyxoid sarcoma, and myofibroblastic sarcoma. The lack of an invasive growth pattern, absence of nuclear hyperchromatism, cellular pleomorphism and atypical mitotic figures and areas of necrosis

Table I. Clinical and pathological characteristics of benign spindle cell tumors.

	Solitary myofibroma	Fibromatosis	Nodular fasciitis	Solitary fibrous tumor
Age Predilection	Up to 4th decade	Children & young Adults	20–40 years	Adults
Intraoral site	Mandible	Paramandibular soft tissue		Buccal mucosa
Capsule	Well-circumscribed	Poorly circumscribed	Well-circumscribed	Well-circumscribed
Cellular component	Plump spindle-shaped cells with eosinophilic cytoplasm	Plump spindle-shaped cells, more cellular at periphery	Plump spindle-shaped cells	Round to spindle
Arrangement	Biphasic	Long Streaming fascicles	Tissue culture/feathery	Short fascicles to patternless
Stromal Characteristics	Hyalinized at periphery	Variable amounts of collagen	Myxoid (early) to hyalinised (long-standing) Mucopolysaccharide rich	Myxoid areas Prominent hyalinisation
Vascularity	HPC-like vascular pattern	Thin walled vessels	Haemorrhagic areas with hemosiderin deposition	Focal HPC-like vascular pattern
Inflammation	Absent	Scattered lymphocytes at periphery	Scattered lymphocytes in centre	Absent
Alcian blue stain	Negative	Negative	Stroma is Positive	Negative
αSMA	+++	+++	+++	–
Desmin	–	–	Rare	+/-
CD-34	–	–	–	+++
β-Catenin	–	+++ (nuclear)	–	+++ (nuclear)

helped in diagnosis of a benign lesion [9]. In addition, the expression of SMA is not uniform and is restricted in these sarcomatous lesions [15].

The histological features that differentiate nodular fasciitis from other benign spindle cell lesions is given in Table I. Cytoplasmic CD-34 expression is consistent with solitary fibrous tumor (SFT), however, in the present case the expression was restricted to the capillaries in between the tumor cells [15]. Moreover, a nuclear expression of β-catenin is characteristic of fibromatosis and SFT which was negative in the present case [16].

In this case, the spindle-shaped myofibroblast-like cells stained positive for smooth muscle actin. This was also observed in the cases that were reported by Lloyd *et al.*, Ikebe *et al.* and Souza *et al.* [4,17,18] however, NF has to be distinguished from other benign tumors of myofibroblastic origin. Inflammatory myofibroblastic tumor (IMT) is generally larger than NF, tends to occur at a younger age, and is composed of longer fascicles of spindle cells in an inflammatory background that is rich in plasma cells. IMT is a tumor that commonly occurs in the lung, however, the presence of this tumor in the mandible has been reported [19].

NF can be distinguished from solitary myofibroma on the basis of its “biphasic zoning” phenomenon. The biphasic zoning phenomenon is seen in solitary myofibroma and refers to the presence of light-staining collagenous, hyalinized areas adjacent to dark-staining hemangiopericytoma-like areas. SMF also lack the mucopolysaccharide rich stroma as demonstrated in the present case thus directing the diagnosis to nodular fasciitis.

Although nodular fasciitis is regarded as a self-limiting lesion, relocation of USP6 gene was detected. This was also recently reported in a case of nodular fasciitis that presented in temporomandibular joint [20].

The clinical course of nodular fasciitis is essentially benign with tendency for spontaneous regression. The oral NF case reported by De Carti *et al.* underwent partial spontaneous regression within a month of follow-up of the patient [5]. However, complete surgical excision is regarded as the treatment of choice. The recurrence rate is 2% with only one case that recurred [15]. Currently, our patient is under regular follow-up for the last 8 months and there have been no further symptoms.

Conclusion

Nodular fasciitis rarely occurs in the oral cavity, and the mandibular alveolus is a less common site. It is important to consider nodular fasciitis in the differential diagnosis of growths that occur on the mandibular alveolus. Because of its rapid growth it can be misdiagnosed and may be over treated. A complete appraisal of both the cellular and stromal components of the lesion will be of great value in the rendering an effective treatment for the management of these lesions.

Conflicts of interest: The author declare that they have no conflicts of interest in relation to this article.

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