Case Report

Solitary intraosseous neurofibroma of the mandible: Report of an extremely rare histopathologic feature

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ABSTRACT

Neurofibroma (NF) is a benign tumor derived from the peripheral nerve sheath. Neurofibromas may present either as solitary lesions or as part of the generalized syndrome of neurofibromatosis or von Recklinghausen's disease of the skin. The intraosseous variant of NF is very rare. We report a case of a 32-year-old female who was diagnosed with a solitary intraosseous neurofibroma of the mandible. The present case is rare with respect to its unique histopathologic feature.

KEY WORDS: Benign tumor, intraosseous, mandible, nerve sheath, solitary neurofibroma

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INTRODUCTION

Neurofibroma is a benign, well-circumscribed soft tissue tumor of the peripheral nerve sheath phenotype with mixed cellular components which includes Schwann cells, perineural hybrid cells, and intraneural fibroblasts.[1-4] Neurofibroma occurs as a single tumor or associated with neurofibromatosis type 1 (NF1), an autosomal dominant genetically inherited disease.[1-6] A solitary neurofibroma is a single lesion that occurs in a patient who does not have neurofibromatosis.[7] Neurofibromas are mostly observed on the skin and very few cases of neurofibromas have been reported in the oral cavity.[1,2,4] The tongue, buccal mucosa, lip, palate, gingival, and major salivary glands are the most common intraoral sites whereas intraosseous or central neurofibromas of the mandible are very rare.[1,2,4,5] In 1954, Bruce gave the first description of solitary of the oral cavity. Since then, less than 50 cases have been documented in the literature.[8]

We report a case of an intraosseous neurofibroma of the mandible in a 32-year-old female patient who showed a unique histological feature of the tumor.

CASE REPORT

A 32-year-old female patient visited the department of oral medicine of Semnan dental school with a chief complaint of swelling in the lower left side of the face. The patient gave a history of a slow-growing swelling for 6 months with intermittent dull aching pain for the past 3 months. Intraorally there was diffuse swelling in the lingual cortex [Figure 1]. The patient had no medical history and systemic diseases.

On panoramic radiography and CT scan image, there was a well-defined unilocular radiolucency in the left side of the mandible measuring about 3 × 1.5 × 2.4 cm. Lingual cortical expansion and thinning of buccal and lingual cortexes were seen. There was no evidence of root resorption. The loss of lamina dura and PDL widening of the second molar was obvious. Cortical borders of the mandibular canal were lost [Figure 2].

Incisional biopsy was done and the histopathological examination revealed a benign proliferation of spindle-shaped cells with wavy nuclei and densely packed collagen bundles within the fibromatous stroma. The diagnosis of neural tumors was given which was followed by excisional biopsy. A similar histopathological feature was seen in excisional biopsy tissue [Figure 3]. Accordingly, the diagnosis of neurofibroma was confirmed.

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Considering the location of the tumor and radiographical and histopathological features intraosseous neurofibroma was diagnosed.

DISCUSSION

Neurofibroma is a benign soft tissue tumor that originates from the nerve sheath cells.[1] NF is presented as a solitary tumor or as multiple lesions associated with neurofibromatosis type 1 (NF1).[2] In the oral cavity, neurofibroma occurs on the tongue, lip, palate, gingiva, salivary glands, and the jawbones.[2-4]

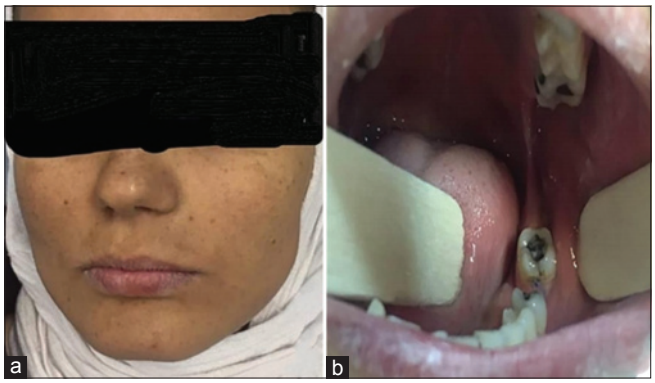

Figure 1: (a) Clinical feature showed swelling in the lower left side of the face (b) Intraoral feature showed diffuse swelling in the lingual cortex

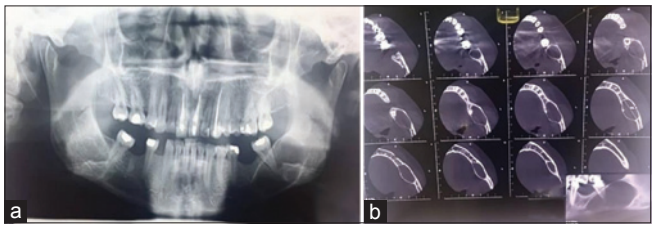

Figure 2: (a) Panoramic radiography showed well-defined unilocular radiolucency in the left side of the mandible (b) CT images

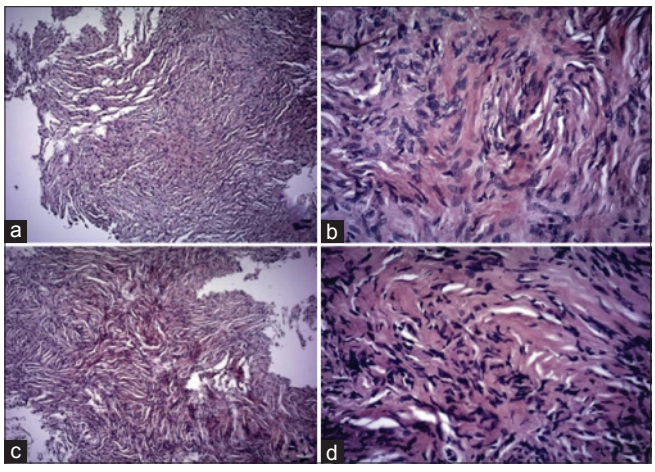

Figure 3: (a) Microscopic feature of the incisional biopsy showed benign proliferation of spindle-shaped cells with wavy nuclei and densely packed collagen bundles within the fibromatous stroma. (H and E × 100) (b) Microscopic feature of incisional biopsy (H and E × 400) (c) Microscopic feature of the excisional biopsy showed benign proliferation of spindle-shaped cells with wavy nuclei and densely packed collagen bundles within the fibromatous stroma (H and E × 100) (d) Microscopic feature of excisional biopsy (H and E × 400)

A literature search of intraosseous NFs of the jaws showed few reported cases. Che *et al.* noticed the number of NFs occurring in the posterior part of the mandible, which could be the reason for the passing of bundles of inferior alveolar nerve in the mandibular canal.[2,9] Neurofibroma occurs at various ages between 14 and 45 years old. A male: female ratio of 1:2 has been observed [Table 1].

Most of the intraosseous NFs are asymptomatic in the initial stages. As the tumor increases in its size, it starts compressing on the adjacent vital structures and begins to destroy the bone.[10] Later on, pain and numbness of the affected side of the lip occur. So far, few cases of symptomatic intraosseous neurofibroma have been reported.[9] Similar to the symptoms reported in the literature search, the present case also depicted the symptoms of swelling and bone loss.[2,4,5]

Radiologically, the tumor appears as a nonspecific, unilocular or multilocular, poorly defined or well-demarcated, radiolucency.[6] Alatli *et al.* reported an intraosseous neurofibroma with no abnormality in radiographic features.[11]

Histopathologically, NFs exhibit an irregular pattern with interlacing bundles of spindle-shaped cells with round or fusiform nuclei and eosinophilic cytoplasm within a loose matrix of delicate fibrillary collagen and a variable amount of myxoid matrix.[1,2,12] Ide *et al.*[13] also recognized that neurofibroma is composed of a complex proliferation of Schwann cells, perineural cell, endoneurial fibroblasts, and intermediate cells. The researcher's distinguished three types of neurofibromas (type I, II, and III) based on their reactivity to different markers and ultrastructural features. This subdivision is useful and represents the variable possibility for different markers. This case should be considered as an intraosseous neurofibroma of controversial diagnosis because it showed no histological features typical of NF. The histopathology feature of this case is remarkable because it showed a fibro collagenous stroma rather

Table 1: ???

than myxoid matrix, and it was similar to some cases which had been reported as central NF.[12,14-21]

It is important to differentiate NFs from other spindle cell tumors such as Schwannoma. Although both have a neural origin, there are slight anatomical considerations associated with them. Neurofibroma attempt to encase the nerve fiber while Schwannomas will typically displace the root of nerve they are associated with.[7]

Neurofibromas are immunoreactive for S100 protein, indicating its neural origin.[1,2] Mast cells are numerous and can be a helpful diagnostic feature.[1] Despite the above features and considering the histopathological architecture of the lesion, the anatomic area (periphery of the inferior alveolar nerve), and the biological behavior, we confirm the diagnosis of neurofibroma.

Complete surgical excision is often possible for solitary intraosseous neurofibroma. On the other hand, complete surgical removal should not be attempted if the neurofibromas do not cause obvious impairment of function and if the patient's condition would not be improved by surgery. Complete excision can be performed if the tumor is small and deteriorating or if the lesion is growing and if the patient has an acceptable hearing on the other side.[22-24]

CONCLUSION

Neurofibromatosis is a relatively common disorder but intraosseous neurofibroma is a rarity. As a solitary lesion, it might present diagnostic difficulties. Hence, it is essential for oral diagnosticians to be aware of even the rare presentations of relatively common disorders which might also be the first indicator of the disease process.

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Conflicts of interest
There are no conflicts of interest.

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