Non-Syndromic Solitary Neurofibroma in floor of the mouth: A case report

Balan N, Sudhaa Mani M, Yasmeen Ahamed S, Sivaraman G.S, Ezhilarasi V

From Vivekanandha Dental College for Women, Elayampalayam., Tamil Nadu, India

Correspondence to: Dr. Ezhilarasi V, Dental College for Women, Elayampalayam., Tamil Nadu, India. E-mail: appaarasi@gmail.com

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ABSTRACT

Neurofibromas are rare in the head and neck region, but most frequent tumor of neural origin. Oral hard and soft tissue is affected by the tumor. In this paper, we describe an unusual case of non-syndromic solitary neurofibroma of the floor of the mouth in a 70-year-female patient with a chief complaint of growth in the floor of the mouth for the past 3 months. An occlusal, intraoral periapical radiograph and CT imaging were done. After confirming the diagnosis, the lesion was excised under local anesthesia and the specimen was submitted for histopathological examination. On subsequent follow-up, the patient was asymptomatic. Intraoral neurofibroma although uncommon, deserve special attention because of their similarity with other inflammatory neoplastic condition, and their tendency to undergo malignant transformation.

Keywords: *Mouth, Neurofibroma, Oral cavity, Syndromic.*

Neurofibroma is a benign neurogenic tumor, which can be either single or multiple [1,2]. A solitary neurofibroma (SNF) is a single lesion that occurs in an individual who does not have hereditary neurofibromatosis [3]. It is a benign, unencapsulated, well-circumscribed lesion containing Schwann cells, endoneurial fibroblasts, and peripheral cells. The presence of neurofibroma is rare in the oral cavity and is even rarer in the floor of the mouth [4]. Neurofibrosarcoma is a major complication which has a poor prognosis [5]. Here, we report the case of a non-syndromic solitary neurofibroma arising from the floor of the mouth in a 70-year-old female patient.

CASE REPORT

A 70-year-old female patient visited the dental hospital with the chief complaint of growth in the floor of the mouth for the past 3 months. The patient stated that the growth had started insidiously 3 months back, which was initially small and had grown gradually to reach the present size. There was no history of an increase in the size of swelling during meal time, with no associated pain, paresthesia, or any discharge. There was no difficulty in swallowing or similar swellings or growths elsewhere in the body. Her medical, surgical, and dental histories were noncontributory.

On general examination, the patient was conscious, cooperative with moderate build, normal gait and posture. There were no positive signs of pallor, icterus, cyanosis or clubbing. On extraoral examination, vital signs include blood pressure 140/90 mmHg, pulse rate71 beats /minute and temperature 37.2^0c. No gross facial asymmetry detected. A single right submandibular lymph node was palpable, which measured approximately about 1x1.5cm, roughly oval in shape, non-tender, firm in consistency and mobile. On intraoral examination, a dome-shaped, sessile swelling measuring approximately 4x3cm, red in color with dilated blood vessels was present on the floor of the mouth (Fig. 1). It extends anteroposteriorly from mandibular anterior teeth to lingual frenum, and mediolaterally extends in relation to 31 to 32 regions, it crosses the midline. The borders were well-defined; surface appears smooth and no other secondary changes were seen. On palpation, all inspection findings were confirmed with respect to the site, size, shape and extension as it was, non-tender, firm to hard in consistency. Bimanual palpation of the submandibular salivary gland was done which does not reveal any positive findings. The salivary flow rate was normal and flowing from the Wharton's duct.

Based on the history and clinical features, the lesion was provisionally diagnosed as a benign minor salivary gland tumor

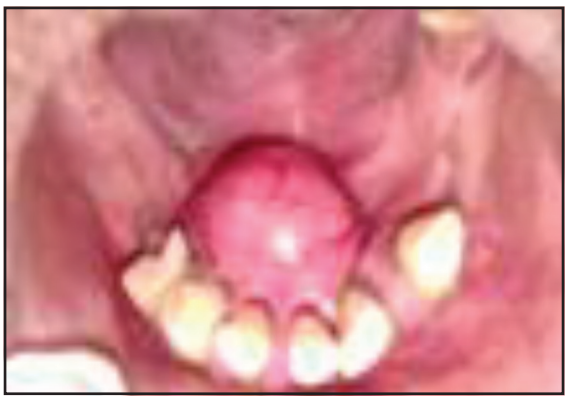

Figure 1: Clinical photograph showing dome-shaped swelling in the floor of the mouth

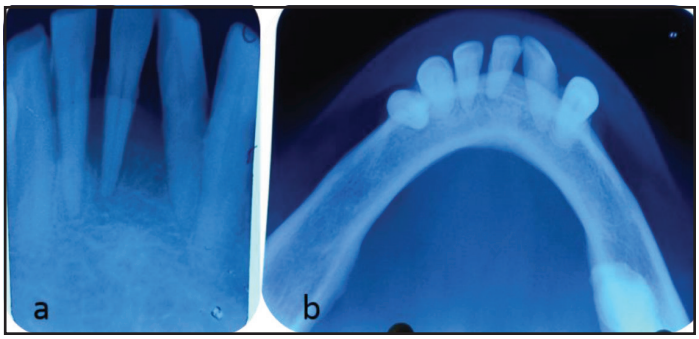

Figure 2: (a) IOPA shows a loss of lamina dura with horizontal bone loss in relation to 31,32,41,42 regions; (b) cross-sectional mandibular occlusal radiograph reveals horizontal bone loss in relation to 31, 32, 33, 41, 42, 43.

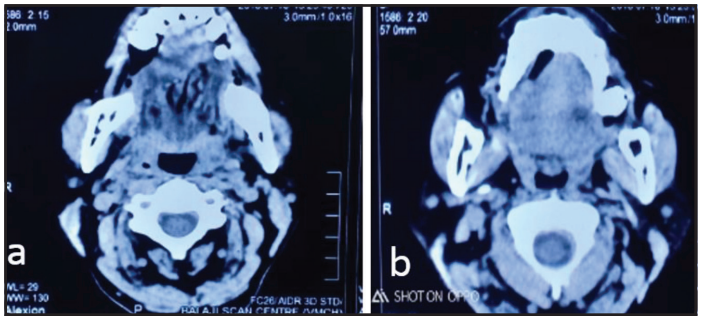

Figure 3: (a and b) Axial view of mandibular CT reveals well-defined hypodense lesion measures 31x 16 mm in the right sublingual space involving the floor of the mouth pushing the anterior genioglossus muscle to the left. No adjacent mandibular erosion was found.

probably pleomorphic adenoma. Differential diagnosis included as fibroma, fibrolipoma, Peripheral giant cell granuloma, schwannoma, (neurilemmoma).

Intraoral periapical radiograph (IOPA) shows loss of lamina dura with horizontal bone loss in relation to 31,32,41,42 regions (Fig. 2a). A cross-sectional mandibular occlusal radiograph shows horizontal bone loss in relation to 31, 32, 33, 41, 42, 43 (Fig. 2b).Computed tomography (CT) mandible reveals a well-defined hypodense lesion measuring 31x16 mm in the right sublingual space involving the floor of the mouth and pushing the anterior genioglossus muscle to the left. No adjacent mandibular erosion was found (Fig. 3a and b)

The lesion was excised under local anesthesia and the specimen was submitted for histopathological examination (Fig. 4a and b).Hematoxylin& Eosin (H&E) stained sections show considerable variation but are generally composed of a proliferation of delicate spindle cells with thin, wavy nuclei intermingled with neurites in an irregular pattern as well as delicate, intertwining connective tissue fibrils. There was no evidence of malignancy in the cells. The histopathologic appearance was consistent with neurofibroma (Fig.5). Immunopositivity for S-100 protein was also seen.

With a histopathologic diagnosis of neurofibroma, the patient was recalled and a thorough general examination was done, to rule out the swelling elsewhere in the body. This excludes neurofibromatosis and the diagnosis of solitary neurofibroma was confirmed. The patient was followed-up after 6 months and there was no recurrent lesion.

DISCUSSION

Neurofibroma is a benign tumor of the peripheral nerve sheath and originates from Schwann cells and perineural fibroblast [6]. According to Salla *et al* in 2009, the prevalence of oral SNF was found to be 3.5%, [7] while Borges *et al* in 2013 reported as 6 % [8]. Hence, the prevalence ranges between 3% and 6%. They occur in a wide age range from 10 months to 70 years, and the mean age is 30 years [9]. Among the gender, a very high female preponderance was seen with the male to female ratio of 1:9. But, Depprich *et al* found an equal gender distribution [10].

Most frequent location occurs in tongue, although they may occur at any site especially on the palate, cheek mucosa, and floor of the mouth [9]. According to the World Health Organization, neurofibroma is classified into two types as dermal and plexiform types [11]. Clinically, oral neurofibromas usually appear as a pedunculated or sessile nodule, with slow growth. They are usually painless, but pain or paresthesia may occur due to nervous compression [9]. Although neurofibromas

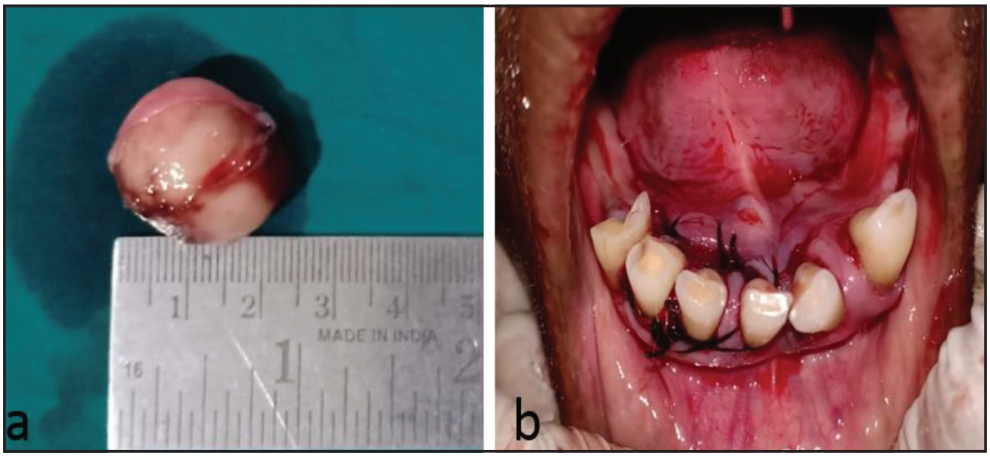

Figure 4: (a) The lesion was excised from the floor of the mouth and (b) post-excisional biopsy done from the floor of the mouth.

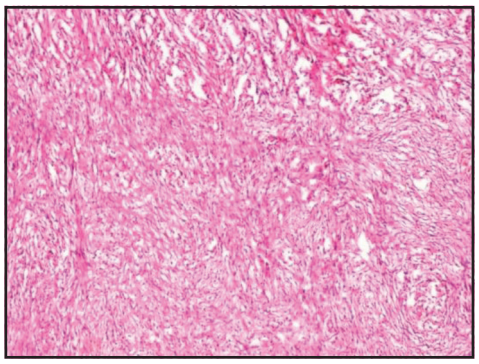

Figure 5: High power view showing Schwann cells with elongated to round nuclei.

are often found in the soft tissue of head and neck region, central variant such as in the jawbones of the mandible or maxilla are rare [10,11].

Under the light microscope, they exhibit numerous spindle-shaped cells with round or fusiform nuclei which are present in interlacing bundles. The cells have eosinophilic cytoplasm, and the cells are distributed in a matrix of fine fibrillar collagen. They have a capsule and have a mix of perineural cells such as Schwann cells and fibroblasts. Immunohistochemically, neurofibromas are S 100 positive [10].

Radiographic findings in oral neurofibroma involving mandibular canal show fusiform enlargement of the canal. Neurofibroma may seldom be primarily intra-osseous, in that case, they usually appear as a unilocular with well-defined radiolucency [12]. Magnetic Resonance Imaging (MRI) reveals a low signal in T1 and a high signal in T2, with a variable highlighting with the contrast. A high peripheral signal with a low central signal in T2 weighted images (bull's eye sign) is a typical sign of neurofibromas. Most neurofibromas show low attenuation in CT scans, although some of them may show soft tissue density. Low-density lesions contain a variable amount of Schwann cells, which are rich in lipids, cystic degeneration, and xanthomatous alterations. High-density areas are thought to represent collagen-rich or cellular areas [13].

Total or partial resection of neurofibromatous lesions is the treatment of choice to solve aesthetic or functional problems; it is advisable to wait for treatment until growth has been completed thus diminishing the risk of recurrence. Total resection with 1 cm margins whenever feasible is the treatment of choice for accessible and small tumors. Usually, radiotherapy or chemotherapy is not recommended for treatment [12].

CONCLUSION

Early diagnosis and treatment in such patients are very important along with regular follow-up during their lifetime to detect recurrences and to rule out the manifestation of vonRecklinghausens disease especially in the central nervous system. So, whenever solitary exophytic growth is encountered in the floor of mouth, oral neurofibroma should be considered in differential diagnosis.

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