

CASE REPORT: PRIMARY INTRAOSSEOUS NEUROFIBROMA OF MAXILLA

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ABSTRACT

The present article is a case report of solitary neurofibroma of the oral cavity in a 50-year old male patient. Patient reported with a chief complaint of swelling in relation to upper front teeth region since two months. Based on clinical, radiographical and histopathological findings the diagnosis of neurofibroma was made. Neurofibroma involving oral cavity as the primary lesion is very rare in occurrence. It mainly involves tongue, palate and buccal mucosa. Surgical excision is performed in the present case.

KEYWORDS: *Solitary Neurofibroma, Intraosseous Neurofibroma, Soft Tissue Tumor of Palate*

INTRODUCTION

Neurofibroma is a genetic disorder with autosomal dominant inheritance, rarely occurring as a solitary lesion in the oral cavity. The etiopathogenesis of this rare variant remains unknown. The present case report adds to the literature and the findings of which help in understanding the lesion.

Neurofibroma is a benign tumor arising from the peripheral nerve sheath. The symptoms of which can range from localized swelling to multiple cutaneous swellings affecting skin. Majority of neurofibromas occur as an autosomal dominant type of inheritance with strong genetic predisposition. This article presents a case report of a solitary intraoral neurofibroma with no known family history and absence of other cutaneous manifestations.

Case Report

A 50-year old male patient reported to the department of oral medicine and radiology, G. Pulla Reddy Dental College, with a chief complaint of pain and swelling in relation to upper front teeth region since two months. Extraorally mild diffuse ovoid swelling was present at the upper lip region with obliteration of normal contour of the philtrum. No history of trauma was present. Lymph nodes were not palpable. On intraoral examination, a diffuse swelling was present in relation to 11 and 21 in the midline. The surface of the lesion was smooth, normal in color with no signs of inflammation and ulceration. The lesion extended from labial mucosa to palatal surface with swelling occupying palatal rugae region, with obliteration of labial vestibule and frenum at the involved site. The swelling measured 3 × 2 cm in size labially. On

palpation, the swelling was sessile, firm in consistency and was fixed to underlying structures. Hard tissue examination revealed grade 1 mobility in relation to 12, 11, 21 and 22 and teeth involved were vital. A patient was known diabetic and is under medication. No relevant medical and dental history was reported. No other systemic manifestations were evident.

OPG revealed radiolucency in relation to 11, 12, 21 and 22 with irregular borders. Resorption of roots of involved teeth is evident. Teeth with respect to 11 and 21 showed distal migration. CT was performed for the present case which revealed a well-defined round to oval expansile lytic lesion (measuring 34×34×30 mm) with partially calcified/corticated peripheral wall seen in the alveolar process of the anterior maxilla and anterior hard palate in the midline causing anterior contour bulge. Based on clinical and radiographic features a provisional diagnosis of leiomyoma, schwannoma and hemangioma were considered.

Fine needle aspiration revealed negative aspiration. An incisional biopsy was done in the present case. The specimen was whitish grey in color with irregular borders and firm in consistency. The tissue was processed and H&E stained. Microscopic examination of the H&E stained tissue under 10x revealed fibrocellular connective tissue with prominent nerve bundles. The nerve fibers were with wavy nuclei and connective tissue is densely arranged collagen fibers with inflammatory cell infiltrate predominantly lymphocytes. Immunohistochemistry findings were positive for S-100 protein and CD34.

Complete excision was performed in the present case. Excision was carried out under general anesthesia. A labial flap was elevated and a vestibular incision was given as shown in the figure. Vertical and horizontal bony cuts were given to mobilize the tumor segment. The complete premaxillary segment was removed from canine to canine. After excision cauterization was performed to achieve hemostasis. Nasolabial flap was sutured to palatal and labial mucosa. The intraoral soft tissue defect was reconstructed with nasolabial flap. Postoperative ryles tube was advised for two weeks. After one month we planned for obturator.

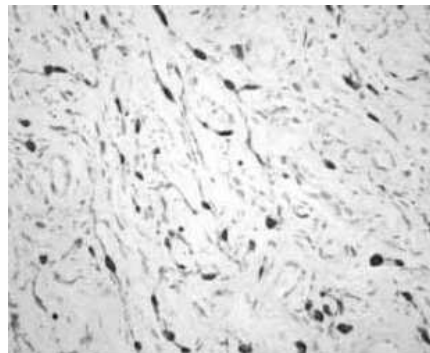


Figure 1

DISCUSSIONS

Neurofibroma is a benign tumor arising from nerve tissue origin, derived from the cells which constitute the nerve sheath. Neurofibroma can arise either as a solitary lesion or as a component of neurofibromatosis type I (NF1, also called as von Recklinghausen's disease of skin).^[1] Neurofibromatosis type I is a neuroectodermal tumors arising within multiple organs and is inherited as an autosomal-dominant inheritance. It is the most common type and accounts for about 90% of the cases, and is characterized by the occurrence of neurofibromas along the peripheral nerves and multiple cafe-au-lait spots.^[2,3] In 1988, National Institute of Health Consensus Development Conference proposed a diagnostic

criterion for neurofibromatosis type 1, if a patient has two or more of the following findings:^[4]

- Six or more cafe au lait macules
- Two or more neurofibromas of any type or one plexiform neurofibroma
- Freckling in the axillary or inguinal regions
- Optic glioma
- Lisch nodule
- Distinctive osseous lesion such as sphenoid dysplasia
- Family history of the first-degree relative with neurofibromatosis.

In the present case, the patient showed a palatal neurofibroma and the other features were not evident. Shklar and Meyer classified neurofibroma as solitary and multiple. Multiple neurofibromas are associated with Neurofibromatosis type 1 whereas solitary neurofibromas are not associated with Neurofibromatosis type 1 and it occurs very rarely in the oral cavity. For solitary neurofibroma, the exact cause is unknown. As many as 50% of cases are reported to be caused due to the spontaneous mutations.^[5] The present case showed a solitary neurofibroma with negative family history.

In the late 19th century, 2 subsets have been identified. One is associated with Neurofibromatosis type-1 (NF1), caused by a mutation of an NF1 gene located on the chromosomal segment on the long arm of chromosome 17 (17q11.2) which encodes a protein known as neurofibromin, which plays a role in neural cell signaling. The other is associated with Neurofibromatosis type-2 (NF2), caused due to the gene mutations of the tumor suppressor genes coding for schwannomin on chromosome 22q12.1.^[6, 7]

World Health Organization (WHO) classified neurofibromas into 2 categories: dermal and plexiform. Dermal neurofibromas are associated with a single peripheral nerve, while plexiform neurofibromas are associated with multiple nerve bundles.^[8]

Solitary neurofibromas appear as soft, painless, circumscribed, slow-growing lesions that vary in size from small nodules to larger masses. The most frequent location for neurofibromas is the skin. Most common intraoral sites are the tongue and buccal mucosa followed by palate, gingiva, alveolar ridge, and vestibule.^[9, 10] The oral lesions appear as discrete, nonulcerated nodules, which tend to be of the same color as the normal mucosa. Due to the diffuse involvement of the tongue, macroglossia is evident. Few cases have been reported with the involvement of submandibular gland, tongue, and lower lip.

In the present case, the intraoral examination revealed a diffuse swelling in relation to 11 and 21 in the midline. The surface of the lesion was smooth, normal in color with no signs of ulceration. The lesion extended from labial mucosa to palatal surface with swelling occupying the palatal rugae region.

In the present case, the histopathology revealed the presence of bundles of nerve fibers with wavy nuclei and densely arranged collagen fibers with inflammatory cell infiltrate predominantly lymphocytes. Immunohistochemistry was done. Immunohistochemical findings were multiple, well-demarcated fascicles of spindle-shaped nerve cells, most of which are positive for S-100 protein. It showed moderate positive immunoreactivity to S-100 protein and CD34. CD34 is a

useful stain for the differential diagnosis, since neurofibromas contain additional cellular components (endoneurial fibroblasts, perineurial-like cells etc.) that are responsible for this CD34 reactivity.

Solitary oral neurofibromas are treated by surgical excision, depending on the site and extent. Recurrence may occur due to the surgical excision, and multiple recurrences have been associated with malignant transformation.

CONCLUSIONS

Neurofibromas may occasionally manifest as solitary swellings in intraoral region, as primary lesion. Appropriate diagnostic and surgical approach should be carried out in a case of neurofibroma, as done in the present case.

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