Case Report

Solitary Non Syndromic Oral Plexiform Neurofibroma: a Case Report and Review of Literature

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KEY WORDS
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ABSTRACT

In oral cavity neurogenic tumors are rare, especially if they are malignant. Neurofibromas are the benign tumors of nerve sheath origin involving multiple nerve fascicles of the smaller branches of peripheral nerves. They may present as solitary lesions or as a part of generalized syndrome of neurofibromatosis also known as von Recklinghausen's disease (VRD) or very rarely as multiple neurofibromas without VRD. Oral solitary neurofibromas are not uncommon, but histologically plexiform variant is a rare form. This article describes a case of oral solitary plexiform neurofibroma with a review of literature.

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Introduction

Neurofibroma is a benign nerve sheath neoplasm of heterogeneous origin. They are mostly seen as a part of neurofibromatosis type I (NF-1) and the presence as a solitary condition is uncommon. The World Health Organization (WHO) has subdivided neurofibromas into 2 broad categories: dermal and plexiform. Dermal neurofibromas arise from a single peripheral nerve, while plexiform neurofibromas are associated with multiple nerve bundles. Other clinicopathological subtypes include localized neurofibroma (sporadic neurofibroma), diffuse neurofibroma, plexiform neurofibroma, and epithelioid neurofibroma. [1] Past literature reports that the frequency of solitary neurofibromas in the oral cavity remains 6.5% especially in lesions not associated with NF-1. [1] Also, the presence of a plexiform neurofibroma in the oral cavity is rare. [1] Neurofibromas are benign tumors that are composed of neuromesenchymal cells, including Schwann cells, perineurial cells, fibroblast and mast cells. Multiple neurofibromas are found as part of NF-1, also known as von Recklinghausen's disease and neurofibromas are evenly distributed over the body surface. [2] Localized (solitary) neurofibromas most often occur as sporadic lesions in patients without NF-1. In general, sporadic neurofibromas are histologically identical to those seen in NF-1. They are clinically characterized by slow growth, lack of pain, and a superficial location. Histologically, these tumors are unencapsulated and comprise a mixture of Schwann cells, perineurial cells, and endoneurial fibroblasts, and are classified into major and minor variants based on their morphological features. [3] Major variants include plexiform, diffuse, and pacinian neurofibromas; while minor variants include epithelioid, cellular, myxoid, glandular, xanthomatized, and other neurofibromas. [3] The diagnosis can be confirmed by histological examination. Neurofibromas are immunopositive for the S-100 protein in 85 to 100% of the cases, indicating its neural origin. [4] Neurofibromas may exhibit sarcomatous alteration in 3%–15% of cases; especially in multiple neurofibromatosis. Occasionally, the malignant transformation of plexiform neurofibroma is reported. These

have poor prognosis and are designated as a malignant peripheral nerve sheath tumor. [1] This article reports a case of solitary plexiform neurofibroma of maxilla in a female patient without any other manifestations or family history of NF-1.

Case Report

A 37-year-old female patient, apparently healthy individual referred to the Department of Oral Pathology complaining of a solitary, painless swelling in the right upper labial mucosa since 4 years that gradually grew in size. No other symptoms were reported before the onset of the swelling. There was no history of any sort of trauma or spontaneous bleeding. The patient did not give history of substance abuse. Extra orally a single, diffuse swelling was present extending from ala of the nose to the corner of mouth (Figure 1).

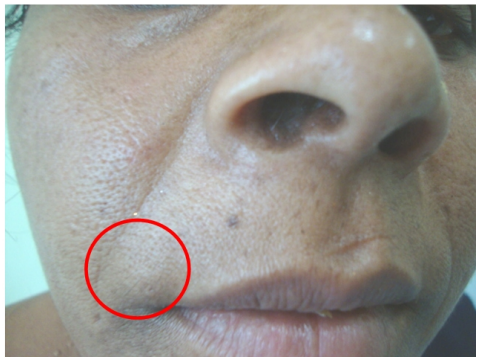

Figure 1: Extraoral photograph showing diffuse swelling obliterating nasolabial fold, elevating the right nasal floor.

Swelling was soft to firm in consistency. Temperature of the overlying skin was normal. Routine hematological examinations were carried out which revealed normal red blood cell and white blood cell counts. Platelet count was also within normal limits. The HIV status of the patient was negative. All routine investigations were done along with HIV status before planning for incisional biopsy. An incisional biopsy of swelling was performed. Histopathological examination of the haematoxylene and eosin stained section showed parakeratinized stratified squamous epithelium and connective tissue. The connective tissue showed proliferation of delicate spindle cells with wavy nuclei intermingled with connective tissue fibrils (Figures 2a and b). Abundant nerve tissue fascicles and collagenous stroma arranged in an orderly fashion was noted. Connective tissue capsule was evident. Toluidine blue stain revealed presence of numerous mast cells 5-6/high power field (Figure 2c).

The present case was diagnosed as plexiform neurofibroma, solitary type. Surgical excision of the lesion was done and patient was followed up for a period of 15 months. There was no recurrence of the excised lesion or any new lesion and no other clinical manifestations developed related to NF-1. Based on this, we ruled out syndromic neurofibromatosis and no genetic studies were carried out.

Discussion

Plexiform neurofibroma (PN) is an unusual histological variant of neurofibroma and is defined as a benign tumor of nerve sheath origin involving multiple nerve fascicles of the smaller branches of the peripheral nerves. [5] PNs develop frequently in individuals with NF1. Physical examination can detect superficial PNs in about 27% of NF1 patients by internal PNs can only be detected by imaging studies such as magnetic resonance imaging. [6] PNs exhibit a bizarre histopathologic picture. Tumor mass is generally well circumscribed, locally invasive and leads to great deal of deformity.

Blending of myxoid and collagenous stromal elements is usually seen. These lesions produce a typical

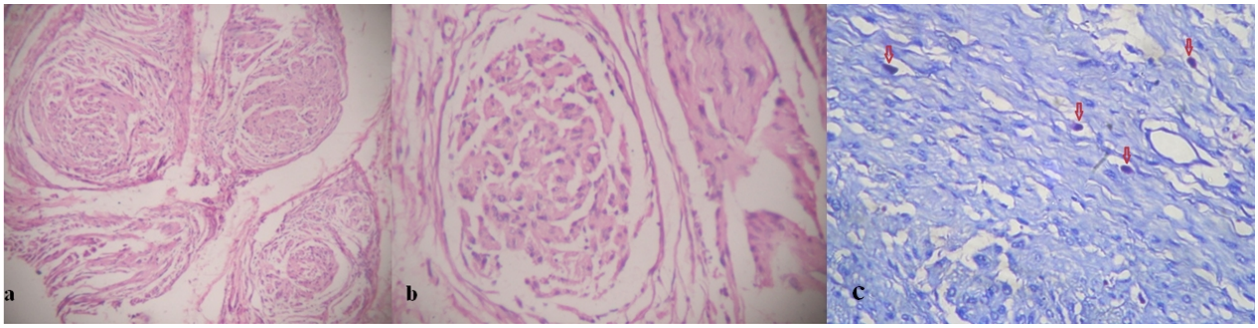

Figure 2a: Microphotograph showing proliferating spindle shaped cells forming fascicles or microfascicles that are coursed in different directions. (H&E, 100X), **b:** High power view showing Schwann cells with elongated to round nuclei predominantly seen with crosses sectioned fascicles (H&E, 400X), **c:** Mast cells with pink granules in the cytoplasm (arrows). (Toluidine blue, 100X)

"bag of worms" appearance. Plexiform neurofibromas are considered to be pathognomonic for NF-1. Their occurrence as a solitary form in the oral cavity is exceedingly rare. They are usually diagnosed in children and occurrence after adolescence is uncommon. [1]

We reviewed the literature for the non-syndromic cases of plexiform solitary neurofibroma of oral cavity [1, 3-5, 7-14] and found 11 cases (including the present case) reported which predominantly occurred in the age group of 30-70 years. 81.8% of cases showed female predilection. There was wide variation in the site of occurrence. 36.3% occurred in buccal mucosa followed by mandibular gingiva (18%), lower lip (18%), and floor of the mouth (9%), maxillary gingiva (9%), tongue (9%) and labial mucosa (9%). [1, 3-5, 7-14]

PN can involve superficial tissue or deeper tissue and the most common site is trunk (43%), head and neck area (42%) and limbs (15%). [5] It is important to distinguish between isolated neurofibromas and those associated with NF-1 because the treatment and prognosis differ greatly. Neurofibromas associated with NF-1 are more likely to recur or undergo malignant transformation. [6] PNs are known to transform into malignant peripheral nerve sheath tumors, which occur in about 10% of NF-1 patients. [7]

Our case is unique as the lesion is sporadic, without any familial history was recorded. A thorough clinical examination of the patient ruled out NF-1. Surgical excision of the lesion was done and patient was kept under follow for a period of 15 months which showed no clinical manifestations of NF-1.

The present case report try to highlight that PN can occur in the oral cavity as a benign, isolated and superficial tumor in patients with no family history or other features of NF-1.

The pathogenesis of isolated solitary plexiform neurofibroma is still unclear due to less number of cases reported. Studies with large number of cases can help in depicting the exact nature of the disease process.

Conflict of Interest

The authors of this manuscript certify no financial or other competing interest regarding this article.

References

[1] S S, Shashikumar P, H S S, Kumar GS. Solitary plexiform neurofibroma of the gingiva: unique presentation in the oral cavity. J Clin Diagn Res. 2013; 7: 2090-2092.

[2] Lee YB, Lee JI, Park HJ, Cho BK. Solitary neurofibromas: does an uncommon site exist? Ann Dermatol. 2012; 24: 101-102.

[3] Ohno J, Iwahashi T, Ozasa R, Okamura K, Taniguchi K. Solitary neurofibroma of the gingiva with prominent differentiation of Meissner bodies: a case report. Diagn Pathol. 2010; 5: 61.

[4] Depprich R, Singh DD, Reinecke P, Kübler NR, Handschel J. Solitary submucous neurofibroma of the mandible: review of the literature and report of a rare case. Head Face Med. 2009; 5: 24.

[5] Zwane NP, Noffke CE, Raubenheimer EJ. Solitary oral plexiform neurofibroma: review of literature and report of a case. Oral Oncol. 2011; 47: 449-451.

[6] Nguyen R, Dombi E, Widemann BC, Solomon J, Fuensterer C, Kluwe L, et al. Growth dynamics of plexiform neurofibromas: a retrospective cohort study of 201 patients with neurofibromatosis 1. Orphanet J Rare Dis. 2012; 7: 75.

[7] Marocchio LS, Pereira MC, Soares CT, Oliveira DT. Oral plexiform neurofibroma not associated with neurofibromatosis type I: case report. J Oral Sci. 2006; 48: 157-160.

[8] Kınış V, Özbay M, Bakır S, Keleş AN. Solitary plexiform neurofibroma of the buccal region unassociated withneurofibromatosis type 1. Kulak Burun Bogaz Ihtis Derg. 2013; 23: 242-245.

[9] Borges AH, Correia Rde M, Borba AM, Guedes OA, Estrela CR, Bandeca MC. Unusual solitary neurofibroma on the lower lip of a child. Contemp Clin Dent. 2013; 4: 512-514.

[10] Gómez-Oliveira G, Fernández-Alba Luengo J, Martín-Sastre R, Patiño-Seijas B, López-Cedrún-Cembranos JL. Plexiform neurofibroma of the cheek mucosa. A case report. Med Oral. 2004; 9: 263-267.

[11] Alatli C, Oner B, Unür M, Erseven G. Solitary plexiform neurofibroma of the oral cavity A case report. Int J Oral Maxillofac Surg. 1996; 25: 379-380.

[12] Aduana V, Mohammadi H, Vaiana J, Ghosh L. Hemangiopericytoma arising in a solitary plexiform neurofibroma: report of a case and review of the literature. J Oral Maxillofac Surg. 1988; 46: 1106-1109.

[13] Güneri EA, Akoğlu E, Sütay S, Ceryan K, Sağol O, Pabuçcuoğlu U. Plexiform neurofibroma of the tongue: a

case report of a child. Turk J Pediatr. 2006; 48: 155-158.

[14] Kamra HT, Dantkale SS, Birla K, Sakinlawar PW, Bharia PH. Plexiform neurofibroma in the submandibular gland along with small diffuse neurofibroma in the floor of the mouth but without neurofibromatosis-1: a rare case report. Ecancermedicalscience. 2013; 7: 313.