SOLITARY NEUROFIBROMA ON THE FLOOR OF THE MOUTH: A RARE CASE REPORT AND REVIEW OF LITERATURE

ABSTRACT

Neurofibroma is an uncommon benign tumor of oral cavity derived from cells that constitute nerve sheath. The cases of oral cavity that involves neurofibroma with no other signs of neurofibromatosis is rare. Neurofibromas may present either as a solitary lesions or as a part of the generalised syndrome of neurofibromatosis or Von Recklinghausen's disease of the skin. Clinically, oral neurofibromas usually appear as a pedunculated or as sessile nodules, with slow growth and mostly without pain. The diagnosis can be confirmed by histological examination. Neurofibromas are immunopositive for the S-100 protein, indicating its neural origin. Treatment is surgical and the prognosis is excellent. Neurofibroma arising from the floor of the mouth is extremely rare. Here we present an unusual case of neurofibroma of the floor of the mouth.

Key words: solitary neurofibroma, neurofibromatosis, immunohistochemistry.

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Introduction

Localized or solitary neurofibroma develops along a peripheral nerve as a focal mass with well-defined margins but is never encapsulated. It is seen either as a solitary lesion or as a part of the generalized syndrome of neurofibromatosis called as Von Recklinghausen disease of the skin.[1] Since the first description of solitary neurofibroma (neurilemmoma, schwannoma) of the oral cavity in 1954 by Bruce, only few cases have been reported in the literature.[2] The oral lesions seldom transform into sarcoma but may become large enough to interfere with the proper functioning of the tongue, which may be a hindrance to the patient.

Clinically, oral neurofibromas appear as pedunculated or sessile nodule, with slow growth. They are usually painless, but pain or paresthesia may occur due to nerve compression. Skin is the frequent location of neurofibromas but lesions in oral cavity are also not uncommon. The most frequent location is the tongue, although they may occur at any site, especially on the palate, buccal mucosa and floor of the mouth. On rare occasion the tumor can arise centrally within the bone. [3]

Case report

A 38 year old female patient reported in the department of oral medicine in Annoor dental college, Muvattupuzha with a complaint of a painless swelling in the floor of the oral cavity. The swelling had started about two years back and gradually to attain its present size. On examination, a well circumscribed 2.5x3cm lesion was seen on the floor of the oral cavity lingual to mandibular left premolars.(fig:1) The mucosa over the lesion was normal with no redness or draining sinuses seen over it. On palpation, the swelling was non- tender, firm in consistency, nodular and fixed to the underlying tissues. The lesion was non-pulsatile and non reducible. Careful examination was done to rule out Lisch's nodules and Crowe's sign. The patient was moderately built and nourished and did not have any other swellings palpable elsewhere in the body. A provisional diagnosis of a benign tumour was made and an excisional biopsy was planned under local anesthesia. The excised specimen was almost 4cm in length. It was sent for histopathological examination. The excised specimen appeared as a firm whitish mass which had a shiny surface.

Hematoxylin and eosin stained soft tissue section showed hyperkeratinized hyperplastic stratified squamous epithelium with evidence of basilar hyperplasia. The underlying connective tissue was composed of dense bundles of spindle shaped cells with wavy nuclei suggestive of nerve cells with collagen bundles. Numerous mast cells and blood capillaries were also evident. On corelating clinical and histopathological evidence the lesion is suggestive of neurofibroma. (fig:2,3)

Discussion

Neurofibroma of the oral cavity is a rare, benign, non-odontogenictumor.[4] Neurofibromatosis (Von Recklinghausen's disease) is an autosomal dominant disease which affects the neural crest cells that give rise to ectodermal and mesodermal derivatives. This genetic disorder affects 1 in every 3000 of the population and has the highest mutation rate among genetic disorders.[5]

Von Recklinghausen's disease which occurs as a result of an abnormality of chromosome 17 and have the characteristic features of Café au lait spots, multiple neurofibromas, Lisch nodules (hamartomas of the iris) and Crowe's sign (axillary and inguinal freckling). Solitary neurofibroma, by definition is seen in those patients who do not have neurofibromatosis.[6,7] Only few cases of solitary neurofibroma have been reported in the literature. [8]

A solitary neurofibroma must be differentiated from a schwannoma. A schwannoma is encapsulated, eccentric to the nerve and composed of Schwann cells. A neurofibroma on the other hand, incorporates the nerve (which may or may not be identifiable) and it is composed of Schwann cells, perineural-like cells, fibroblasts and transitional cells. [4]

Neurofibromas occur in people of all age groups, but they are most commonly diagnosed in young adults. Neurofibromas demonstrated 10 variants: classic,

cellular, myxoid, hyalinized, epithelioid, plexiform, diffuse, pigmented, granular cell, and pacinian.[9] Subsequently, new variants were incorporated, as dendritic cell neurofibroma with pseudorosettes, lipomatous, and hybrid tumors.[10] The most commonly affected site is the tongue.[11] Here we reported a case of neurofibroma which is located in the floor of the oral cavity, which is not a common location.

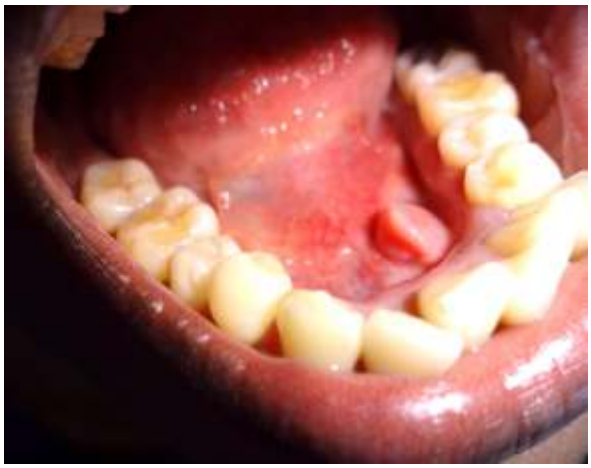
Over time, neurofibromas have propensity for progression into neurofibromatosis. Although these are originally benign lesions they also have tendency for malignant transformation. Almost 6-29% of malignant transformation has been reported.[12,13] Surgical excision by conserving the nerve of origin is the treatment of choice. [14]

Conclusion

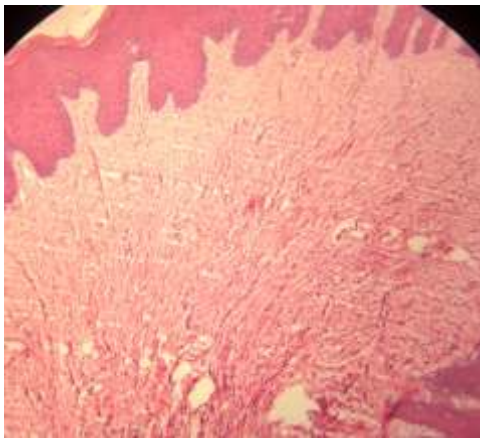
Neuroibromas are benign tumors and have a good prognosis, since they have a propensity for malignant transformation and also chances of progressing into neurofibromatosis, these lesions must be monitored carefully and treated meticulously. The diagnosis of the lesion was made based on the presence of clinical findings and histopathology. Even though neurofibromas are rare in the oral cavity, solitary neurofibromas must be considered in the list for differential diagnoses in cases of intraoral swellings and intraosseous lesions of the jaws.

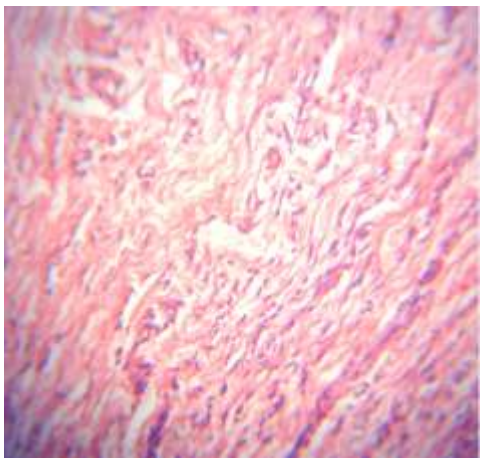
Conflict of Interests

The authors declare that there is no conflict of interests regarding the publication of this paper.



Soft, pedunculated, non-tender, palepink, mobile single swelling estimating from the junction of lingual attached gingiva /floor of the mouth. (fig.1)





Histopathology shows dense bundles of spindle shaped cells with wavy nuclei suggestive of nerve cells with collagen bundles. Corelating clinically and histopathologically suggestive of neurofibroma. (fig:2,3)

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