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CASE REPORT

Neurofibroma the Benign Tumor of Oral Cavity: A Case Report

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Abstract

The malignant tumours of the tongue are far common than the benign ones. Papilloma, fibroma, adenoma, mucous cyst, lymphangioma are the most commonly reported benign tumours. Neurofibroma of the tongue is also a rare one. In this case report, lingual benign tumours and their distinctive histopathological features of a 22 years old male patient has been reported with.The chief complaint of swelling in the tongue since one year. Incision biopsy was done.Histopathology showed spindle cells with thin wavy nuclei along with fine collagen fibrils. Histopathological and immunohistochemical studies confirmed the diagnosis of a myxoid predominant intramural solitary neurofibroma.The purpose of this case report is to apprehendthe uncommon presentation of neurofibroma and to document the successful management of such a lesion using an intraoral approach.

Keywords: *Neurofibroma, Tongue, Histopathological, Radiology, Oncology*

1. Introduction

Cases of Neurofibromas are very rare in head and neck region. Up to 10% of these lesions are associated with neurofibromatosis, an autosomal dominant disorder. Neurofibroma can be of three subtypes- localized, diffuse and plexiform1; 2. Plexiform neurofibroma is least common and is path gnomic of von Recklinghausen disease, seen in 17-30% of patient, caused by mutation of NF1 gene in chromosome. Oral manifestations are described in only 4-7% of patient, and tongue is the most common site1; 3; 4. Approximately 5-10% of plexiform neurofibroma undergoes malignant transformation and their rate of growth is inversely proportional to age. The growth of plexiform neurofibroma is usually ill-defined, and there is a risk of recurrence(2; 4; 5; 6). Here, we present a case of an isolated plexiform neurofibroma of the bottom of tongue.

2. Case

A 22 year old male patient reported with the chief complaint of painless swelling in the tongue since one year. Patient had visited a dentist for the treatment of the swelling in the tongue a few months back. The patient revealed that the swelling had been present for the last one year. The swelling was asymptomatic, did not interfere with swallowing, speech or movements of the tongue. No history of altered sensation and any ulceration or discharge from the swelling. Patient was conscious, co-operative with moderate built, normal gait and posture. There were no positive signs of pallor, icterus, cyanosis or clubbing. No systemic abnormality was detected. A dome shaped swelling was seen on the left dorsal aspect in the posterior part of tongue. It measured approximately 25x30mm. The surface of the swelling appeared normal with unaltered filiform and fungi form papillae. No pulsation was found. There was no restriction of the tongue movements. No abnormality was detected in any other soft tissue components of the oral cavity including the palate, buccal mucosa, floor of the mouth and lips etc. Fine Needle Aspiration Cytology (FNAC) of the lesion was done.

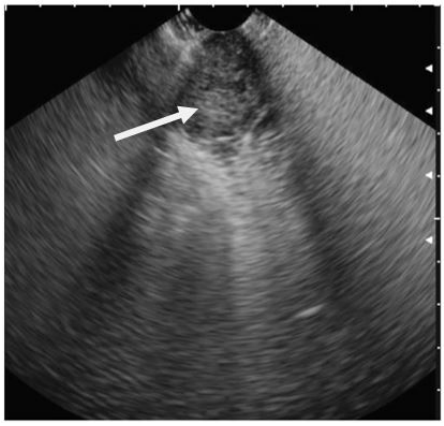

Figure 1: Ultrasound showing hypo echoic lesion on the dorsal aspect of tongue on the left side

Frank blood was obtained on aspiration. On ultrasound, a well-defined hypo echoic lesion measuring 26x28 mm was seen on the dorsal aspect of tongue on the left side(Figure 1). The lesion was seen reaching up to the midline. On Color Doppler study, no internal vascularity was noted. Incision biopsy was done and the stained sections revealed lesion tissue composed of delicate spindle cells with thin wavy nuclei along with fine collagen fibrils. Myxoid and cellular areas were seen. Mast cells were scattered throughout the lesion. The patient was given empirical treatment with oral propanolo and local sclerosant (phenol) injection. However, there was no decrease in size of the lesion so patient was advised for surgical treatment. Patient did not agree for the surgery and never reported back.

3. Discussion

Neurofibromas of the large nerves, which appear clinically as soft, drooping and doughy masses[7], are benign neoplasm's composed of neuritis, Schwann's cells, and fibroblasts within a collagenous or myxoid matrix(7). In contrast to shwannomas, they are nonencapsulated and engulf the nerve of origin. Plexiform neurofibromas, forming tortuous cords along the segments and branches of a nerve with a tendency to grow centripetally, are poorly circumscribed tumors(7). This tumor is claimed to be indicative of VRD albeit it's going to be the sole manifestation of the disease(8; 9). Neurofibromas, usually related to VRD(Von Recklinghausen's Disease), are generally encountered as multiple lesions, and sometime it may occur as a solitary tumor, as in the case. However, the plexiform neurofibroma is never encountered in type-2 and rapid climb of a plexiform neurofibroma usually suggests transformation into a neurofibrosarcoma [7]. Despite their occurrence within the head and neck region, neural sheath tumors are rarely seen within the mouth. Only 4-7% of patients affected by neurofibromatosis display oral manifestations(10). The mobile tongue is the most commonly involved site followed by buccal mucosa, floor of the mouth, palate, lips and gingival(11). Reportavailable on occurrence of a series of neural tumors of the tongue in which there were two neurofibromas, and one malignant and five benign shwannomas(12). The base of the tongue may be a relatively rare location and tumors during this region may cause an upper airway obstruction(13). Respiratory failure and autopsy revealed laryngeal sub mucous plexiform neurofibromatosis nodules as well as extensive plexiform neurofibromas involving the vagal, recurrent laryngeal and phrenic nerves. We didn't encounter with any lesion within the upper airway in the present case. Differential diagnosis of such a tongue mass in childhood must include neurofibroma, shwannomas(neurilemoma), lymphangioma(14), cavernous hemangioma, hematoma, teratoma(15)lipoma(16),myofibroma and myofibromatosis(17),leiomyoma(18),cyst-adenoma(19), pyogenicgranuloma(20), nerve sheath myxoma(21), congenital granular cell tumor(22), and cystic lesions such as mucoid cysts(23), dermoid cysts(24), and cysts of foregut origin(25).

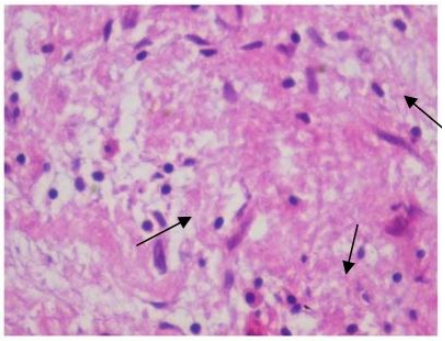

Figure 2: Histopathology shows the tissues of Neurofibroma

The treatment of such lesions is usually surgical and therefore the diagnosis can only be confirmed after histological examination(Figure 2). It is important to differentiate a neurofibroma from a shwannomas histopathologicaly; since the Von Recklinghausen's neurofibromatosis associated with neurofibroma has greater potential for malignant transformation (5-16% is reported)(26). Neurofibromas have extensive vascularity and tend to bleed during surgery. Early diagnosis of Neurofibroma is extremely important and regular follow up of these cases is essential to detect recurrences.

4. Conclusion

The diagnosis of neurofibroma was confirmed by histopathological evaluation and immunohistochemicalstudies. Localized (solitary) neurofibromas most often occur as sporadic lesions, however, diagnosis of a solitary neurofibroma prompts clinical evaluation to exclude the remote possibility of neurofibromatosis. The purpose of this case report is to apprehend the uncommon presentation of neurofibroma and to document the successful management criteria of such a lesion using an intraoral approach.

Conflict of Interest Authors do not have any conflict of interest.

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