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Neurofibroma of tongue – A rare case report

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Abstract
The benign tumours of the tongue are far less common than the malignant ones. Among bening tumours, Papilloma, fibroma, adenoma, mucous cyst, lymphangioma are the tumours most frequently encountered. Neurofibroma of the tongue is rare. A review of the available literature records only 37 cases of tumours of Nerve sheath origin of tongue. The rarity of lingual benign tumours and their distinctive histopathological features have prompted us to report this case.

Keywords: Neurofibroma, Tongue, Histopathological.

Introduction
Neurofibromas are rare in Head & Neck region, but are most common among neural lesion in the region [1]. Up to 10% of these lesions are associated with Neurofibromatosis, an autosomal dominant disorder. Neurofibroma can be of three subtypes- localised, diffuse and plexiform [2, 3]. Plexiform neurofibroma is least common and is pathognomic of von Recklinghausen disease, seen in 17 – 30% of patient, caused by mutation of NF1 gene in chromosome 17. The most common sites in Head and Neck are temple, back of the neck and tongue. Oral manifestations are described in only 4-7% of patient, and tongue is the most common site. [2, 4, 5] Approximately 5-10% of Plexiform neurofibroma undergo 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 [3, 5, 6-8]. Here, we present a case of an isolated plexiform neurofibroma of the base of tongue.

Case Report
A 22 year old male patient reported with the chief complaint of swelling in the tongue since 1 year. It was not associated with sensitivity or pain. Patient had visited a dentist for the treatment of a swelling in the tongue a few months. The patient revealed that the swelling had been present for the last 1 year. It was insidious in onset, it was initially small in size and it has gradually increased to the present size. There has been no significant increase in size in the last 6 months. The swelling was asymptomatic, did not interfere with swallowing, speech or movements of the tongue. Patient does not give any history of altered sensation and any other secondary changes such as ulceration or discharge. Patient was conscious, co-operative with moderate built, normal gait and posture. There were no positive signs of pallor, icterus, cyanosis or clubbing. No abnormality was detected on extra-oral examination (figure 1). A dome shaped swelling was seen on the left dorsal aspect of the tongue in the posterior part of anterior one-third. It measured approximately 25x 30mm. The surface of the swelling appeared normal with unaltered filiform and fungiform papillae (figure 2). No pulsations were evident. 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 labial or lingual gingiva. Fine needle aspiration cytology (FNAC) of the lesion was done. Frank blood was obtained on aspiration. A well-defined hypoechoic lesion measuring 26x28 mm seen on the dorsal aspect of tongue on the left side. The lesion was seen reaching up to the midline (figure 3). On Color Doppler study, no internal vascularity was noted. Incisional biopsy was done and the stained sections revealed lesional tissue composed of delicate spindle cells with thin wavy nuclei along with fine collagen fibrils (figure 4). Myxoid and cellular areas were seen. Mast cells were scattered throughout the lesion. The patient was given empirical treatment with oral propanolol

and local sclerosant (phenol) injection. However, there was no decrease in size of the lesion so patient was advised for surgical treatment. Patient didn't agree for the surgery and never reported back.

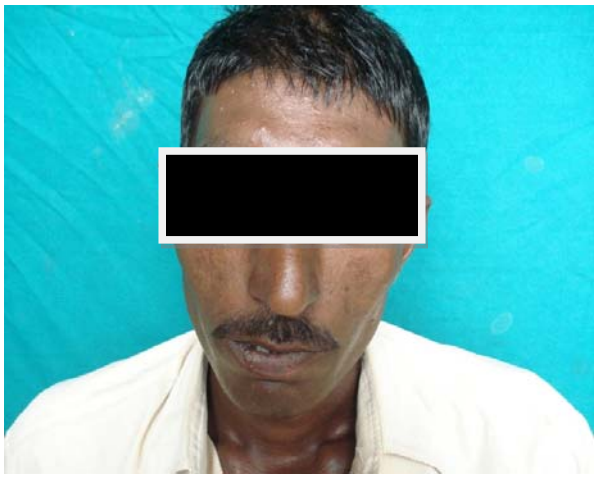

Fig 1: Showing extraoral profile of patient

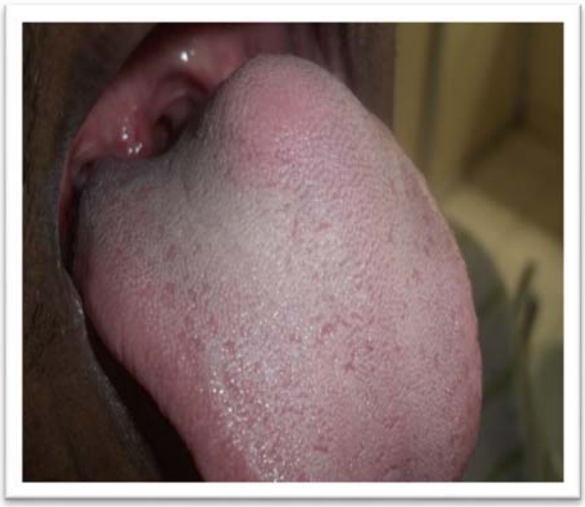

Fig 2: Showing intraoral swelling on the tongue

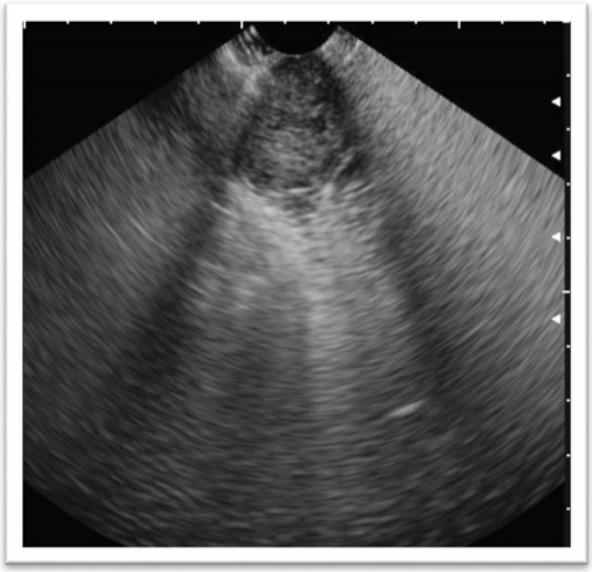

Fig 3: Showing ultrasound

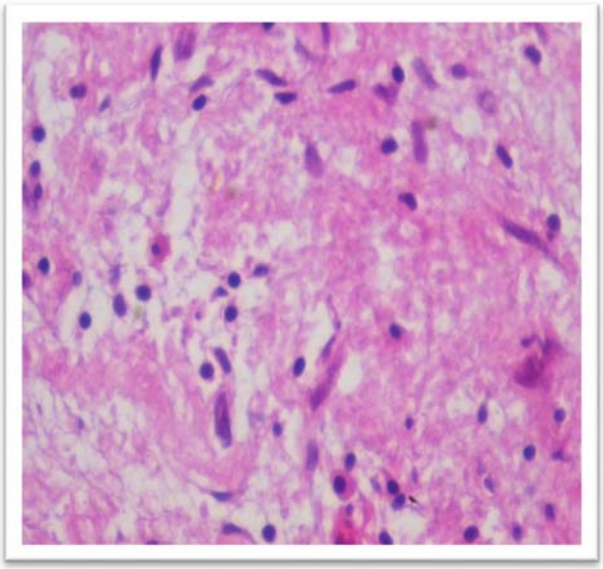

Fig 4: Showing histopathology picture of tissue taken

Discussion

Neurofibromas of the large nerves, which appear clinically as soft, drooping and doughy masses [9], are benign neoplasms composed of neurites, Schwann's cells, and fibroblasts within a collagenous or myxoid matrix [10]. 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 centripedally, are poorly circumscribed tumors [10]. This tumor is said to be indicative of VRD even though it may be the only manifestation of the disease [11,12]. Neurofibromas, usually associated with VRD, are generally encountered as multiple lesions, and rarely occur as a solitary tumor, as in our case. However, the plexiform neurofibroma is rarely encountered in NF-2 and rapid growth of a plexiform neurofibroma usually suggests transformation into a neurofibrosarcoma [9]. Despite their occurrence in the head and neck region, neural sheath tumors are rarely seen in the oral cavity. Only 4-7% of patients affected by neurofibromatosis display oral Manifestations [13]. The mobile tongue is the most commonly involved site followed by buccal mucosa, floor of the mouth, palate, lips and gingiva [14]. Peron *et al.* [15] reported a series of 13 neural tumors of the tongue in which there were two neurofibromas, and one malignant and five benign schwannomas. The base of the tongue is a relatively rare location and tumors in this region may lead to an upper airway obstruction [16]. Since neurofibromas are usually multiple lesions, the whole body must be investigated as well as larynx and trachea in such a patient with oral neurofibroma, since lesions in the upper airway may cause respiratory obstruction. Yamada et al [17]. reported a seven-monthold infant who presented with respiratory distress and a sublingual mass. The patient died due to respiratory failure and autopsy revealed laryngeal submucous plexiform neurofibromatous nodules as well as extensive plexiform neurofibromas involving the vagal, recurrent laryngeal, and phrenic nerves. In our patient, we did not encounter any lesion in the upper airway. Differential diagnosis of such a tongue mass in childhood must include neurofibroma, schwannoma (neurilemoma), lymphangioma [18], cavernous hemangioma, hamartoma, teratoma [19] lipoma [20], myofibroma and myofibromatosis [21] leiomyoma [22] cystadenoma [23] pyogenic Granuloma [24], nerve sheath myxoma [25], congenital granular

cell tumor [26], and cystic lesions such as mucoid cysts [27], dermoid cysts [28] , and cysts of foregut origin [29] .The treatment of such lesions is generally surgical and the diagnosis can only be confirmed after histological examination 1. The family members of the index case should also be examined, since intragenic microsatellite markers were reported to be highly informative for familial neurofibromatosis in Turkish families [30]. It is important to differentiate a neurofibroma from a schwannoma histopathologically, since von Recklinghausen's neurofibromatosis associated neurofibroma has greater potential for malignant transformation, which was reported to be between 5-16% Neurofibromas have extensive vascularity and tend to bleed during surgery. Therefore, excessive bleeding should be kept in mind while attempting surgical removal. Early diagnosis in such a patient is very important and these patients need regular follow up during their lifetime to detect recurrences.

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