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Neurofibroma of Tongue – A Rare Case Report

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Abstract- Neurogenic tumors are rare in the oral cavity. Neurofibroma is a rare benign nonodontogenic tumor which may present either as a solitary lesion or as part of the generalized syndrome of neurofibromatosis or von Recklinghausen's disease (VRD) of the skin. The rarity of benign lingual tumors and their distinctive histopathological features have prompted us to report this case.

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I. Introduction

Neurofibromas are rare in head & neck region but are the most common among neural lesion [1]. 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. Up to 10% of these lesions are associated with neurofibromatosis, an autosomal dominant disorder [1]. Neurofibroma can be of three subtypes, localized, diffuse and plexiform [2, 3].

Approximately 5-10% of plexiform neurofibroma under go 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 [3-8]. Here, we present a case of a plexiform neurofibroma of the base of the tongue.

II. Case Report

An eighteen-years-old man presented to our department with a history of a slowly growing painless mass involving the tongue which was present for two years. Oral cavity examination revealed a pinkish red swelling with an irregular surface involving the right side of the mobile tongue. The swelling, which was evident on inspection, was approximatively 4x5 cm in size and was soft, nonreducible and nonpulsatile on palpation.

Complete blood count was normal. Magnetic resonance imaging (MRI) showed a hypervascularized mass in the tongue which was consistent (figure 1). Histopathological examination of the biopsy revealed lesional tissue composed of delicate spindle cells with thin wavy nuclei along with fine collagen fibrils (figure 2). Myxoid and cellular areas were seen. The patient underwent surgical removal of the mass under general anesthesia which was non-encapsulated and infiltrating deep into the intrinsic tongue muscles with a poor cleavage plane between the lesion and these muscles (figure 3).

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Neither a cranial nerve trunk nor a main vascular structure was seen in the surgical field. The postoperative period was uneventful. The motor functions and sensation of the tongue were normal as well.

The patient was evaluated clinically for features of the syndrome. Multiple café au lait macules were subsequently discovered on his trunk, the patient was referred to a cancer center for further management and follow-up.

III. Discussion

Neurofibromas of the large nerves, which appear clinically as soft, drooping and doughy masses, are benign tumors composed of neurites, Schwann's cells, and fibroblasts within a collagenous or myxoid matrix [9,10]. In contrast to shcwannomas, they are nonencapsulated and engulf the nerve of origin. Plexiformneurofibromas, forming tortuous cords along the segments and branches of a nerve with a tendency to grow centripetally, 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 [2].

Differential diagnosis of such a tongue mass must include neurofibroma, schwannoma (neurilemoma), lymphangioma, hemangioma, hamartoma, teratoma, pyogenic granuloma, nerve sheath myxoma, and cystic lesions such as mucoid cysts and dermoid cysts.

The standard treatment for neurofibromas has been surgical excision and the diagnosis can only be confirmed by histological examination. Neurofibromas have extensive vascularity and tend to bleed during surgery. Therefore, excessive bleeding should be kept in mind while attempting surgical removal [3]. Early diagnosis in such a patient is very important and these patients need regular follow-up during their lifetime to detect recurrences. Fortunately, there were no signs of recurrence or other manifestationsduring the follow-up period of our patient untill that date.

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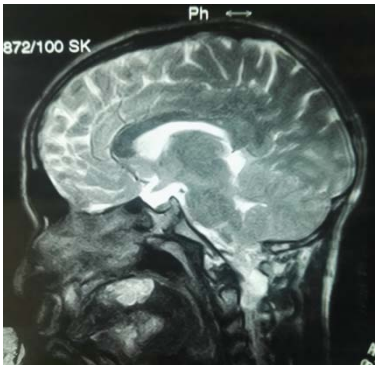


Figure 1: Preoperative-contrast T1-weighted magnetic resonance images of the patient

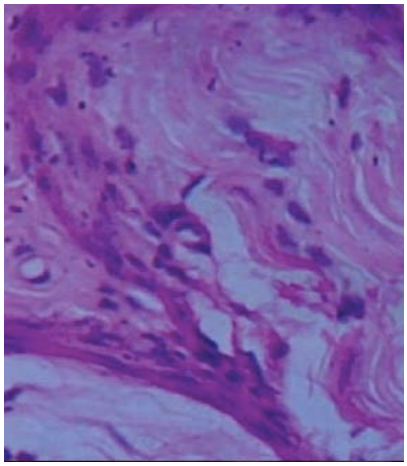


Figure 2: Showing histopathology picture of tissue taken

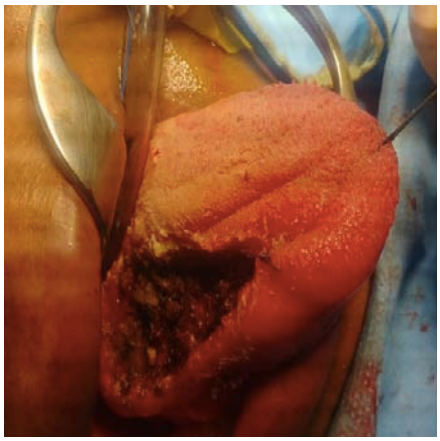


Figure 3: Image of the tongue after surgical excision

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