CASE REPORT-SOLITARY NEUROFIBROMA OF THE TONGUE

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ABSTRACT

Neurofibroma is an uncommon benign tumor of the oral cavity derived from the cells that constitute the nerve sheath. The oral cavity involvement by a solitary and peripheral plexiform neurofibroma in patients with no other signs of neurofibromatosis is uncommon. Amongst the histological variants, plexiform types are considered exclusive. These are poorly circumscribed, locally invasive and may exhibit sarcomatous potential. Plexiform neurofibromas are key features of Neurofibromatosis – 1 and their solitary intra-oral presentation is uncommon.

The following case report describes isolated solitary plexiform neurofibroma of tongue in a middle aged female patient.

Key words: Neurofibroma, Plexiform, Tongue

INTRODUCTION

Neurofibromas are benign tumors arising from peripheral nerve sheaths that affect the head and neck region only rarely,¹ and consist of Schwann cells, perineural cells, and a variable amount of mature collagen.² They are classified as solitary when not associated with any syndrome or multiple when associated with autosomal dominant neurofibromatosis syndrome or type III multiple neoplastic syndrome;³ They can also be classified as myxomatous (solid, central, diffuse) or plexiform (peripheral).⁴

Localized or solitary neurofibroma is the most frequent manifestation and develops along a peripheral nerve as a focal mass with well defined margins but is never encapsulated. Localized or solitary neurofibroma is rare in infancy and typically appears in late childhood or during teenage years. Solitary plexiform neurofibromas are exceedingly rare in oral cavity. Most common sites are buccal region, palate, tongue and lips⁵-¹⁰

Past literature reports that the frequency of solitary neurifibromas in the oral cavity remains 6.5% especially in lesions not associated with neurofibromatosis type -1.¹¹ Also, the presence of a plexiform neurofibroma in the oral cavity is rare.³
Morphologically, the tumor is composed of small axons among the proliferating Schwann and perineural cells, supported by collagen matrix. The treatment of choice is complete excision. The nerve from which the tumors originates may have to be resected in cases where the lesions cannot be dissected. Recurrence is rare.

CASE REPORT

A 35 years old female patient (Fig.1) reported to the outpatient department with a chief complaint of increased mass at the right lateral border of tongue for the past 3 yrs. (Fig.2)

Since then, there had been gradual increase in size of the growth to its present size. The lesion was asymptomatic as there was no history of associated pain, bleeding or any impairment of function.

On physical examination the patient was otherwise healthy. Medical history of the patient was insignificant. Also, no pertinent family history of the similar growths was reported.

On inspection solitary swelling was seen extending 1cm behind from tip of tongue in horizontal direction to right lateral posterior surface of tongue measuring about 3.5 cm (Fig.3). In vertical direction it was extending from dorsal surface to the ventral surface of tongue measuring about 2.2 cm (Fig.4). Swelling was sessile and irregular in shape with well defined edges and smooth surface. Overlying mucosa was normal in color.

On palpation, the swelling was normal in temperature, non tender, firm in consistency, non fluctuant and non compressible with no evident discharge. It had gradually increased to present size within a year of approx 2.5 yr.

No anaesthesia or paresthesia of the lingual nerve distribution was found. No deviation of tongue on protrusion.

Fig.1. Preoperative front view

Fig.2. Preoperative view showing tongue growth
Considering the clinical presentation and localization of the lesion, we included, reactive lesions, such as giant cell fibroma or focal fibrous hyperplasia, neurilemmoma, granular cell tumor, neurofibroma in the differential diagnosis.

On the basis of the diagnostic possibilities and considering the probable benign nature of the lesion, excisional biopsy was planned.

The growth was surgically excised under local anaesthesia using 2% lignocaine with 1: 200,000 Adrenaline along with healthy margins. Primary closure was done with 3-0 Vicryl Suture. The patient was put under analgesics and antibiotics for three days. Post operative healing was uneventful.

The excised mass (Fig.5) was washed with 0.9 % normal saline, stored in 10 % formalin and sent to the department of Pathology for routine histopathological evaluation.

On histopathologic examination the H and E stained sections showed superficial stratified squamous parakeratinized epithelium. Connective tissue showed presence of tumour mass consisting of cells arranged in fascicles. The cells were elongated and spindle shaped consisting of thin wavy nucleus and scanty cytoplasm. Presences of numerous nerve bundles were noted. The surrounding stroma consisted of connective tissue fibrils, fibroblasts and few blood vessels (Fig.6.) Based on these findings the definitive diagnosis of Plexiform Neurofibroma was given.

During the follow up period of approx 1 year no signs of recurrence were noted.
DISCUSSION

Neurofibromatosis is a hereditary condition resulting in a widespread developmental defect of the nerve sheath. Although, the disease became widely recognized as a pathological in the late 19th century: it was only recently, that its two subsets have been defined. They are associated with Neurofibromatosis type–1 and Neurofibromatosis type–2. The former is more common and accounts for about 90% of the cases.

Neurofibromas are benign tumours of nerve cell origin. They are mostly seen as a part of neurofibromatosis–I 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 clinicopathologic subtypes include localized neurofibroma (sporadic neurofibroma), diffuse neurofibroma, plexiform neurofibroma, and epithelioid neurofibroma.

Solitary neurofibroma is a benign, slowly growing, relatively circumscribed, but non-encapsulated tumor diagnosed by absence of other features of the associated systemic disease. Oral lesions are associated with as much as 72% of the multiple form in patients with neurofibromatosis. Intra-oral presentation is extremely rare in case of the solitary type of neurofibromas.

It is believed that the frequency of oral solitary neurofibromas not associated with neurofibromatosis-I is low. They are slow-growing, nodular, sessile, and mobile tumors, usually painless, although pain or numbness may occur due to nerve compression. Cherrick and Eversole observed a higher incidence in females. Chen and Miller reported that oral neurofibromas affect individuals between 9 and 72 years of age.

Although some lesions require imaging tests to determine possible extension, conventional histological analysis is conclusive. Microscopically the tumor is composed of an irregular pattern of proliferative spindle cells. The stroma is composed of collagen fibers and mucoid masses. Small axons all over the tumoral tissue are demonstrated with silver staining. Neurofibromas are immunopositive for the S-100 protein in 85 to 100% of the cases, indicating its neural origin.

Treatment of choice is surgical excision of the solitary lesions, trying to conserve the nerve from which the tumor originates. 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.

The present case is unique as the presentation of lesion was sporadic and no associated family history was reported. A thorough examination of the patient was performed for the various manifestations of neurofibromatosis–I (like cafe au lait spots, lisch nodules, axillary freckling etc). The disease was ruled out due to the absence of the same. The patient had reported with no complications over the follow up period of 12 months and is kept under observation.

References


