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Case Report

Neurofibroma of Tongue - A Case Report

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Abstract:

Neurofibroma is a benign tumor of nerve sheath of peripheral nervous system. It frequently involves the skin and rarely the oral mucosa. The pattern of the disease has been recognized as hereditary with an autosomal dominant trait with variable penetrance. The most common variant is neurofibromatosis type I (NF-I). The oral lesions occur as discrete, nonulcerated nodules, which tend to be of the same color as the surrounding mucosa. Usually occurs on the buccal mucosa, palate, alveolar mucosa, vestibule and the tongue.

A case of asymptomatic swelling of the right side of the tongue near the tip since 02 years is presented. The swelling was gradual in onset and slowly increased in size. It was a solitary diffuse, smooth, firm and non-tender swelling of the right side of the tongue near the tip. Excision was done under lingual nerve block by local anesthetics (2% Lidocain with Adrenalin) and biopsy was performed and histological examination revealed spindle cells having serpentine nuclei, arranged in bundle and fascicles is a fibrillary background.

Keywords: *Solitary lingual neurofibroma, nonmyelinating Schwann cells, NF1genes, spindle cells, serpentine nuclei, sporadic neurofibroma, Lingual nerve.*

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Introduction:

Neurofibroma is a rare benign tumor of the oral cavity derived from the cells that constitute the nerve sheath. Neurofibroma appears either as a solitary lesion or as part of the generalized syndrome of neurofibromatosis. Usually neurofibromatosis type 1 (NF-1), also known as von Recklinghausen disease of the skin. NF-I is estimated to occur in one in every 3000 births with no sex predominance[1]. Most of the time it is tested by genetic method, only 2/3rd of cases are

detected but cannot determine the severity of disease. Oral cavity involvement by a solitary and peripheral plexiform neurofibroma in patients with no other signs of neurofibromatosis is not common. Sporadic cases have been reported in the submandibular gland, tongue and on the periosteum at the mental foramen[2]. This sporadic syndromic occurrence has also been seen in the cutaneous region. Several authors have suggested that these isolated neurofibromas may represent with a hamartomatous growth. The World Health Organization (WHO) has subdivided neurofibromas into 2 broad categories: dermal and plexiform. Dermal neurofibromas arise from a single peripheral nerve on the other hand plexiform neurofibromas are associated with multiple nerve bundles. Other clinicopathologic subtypes include localized neurofibroma (sporadic neurofibroma), diffuse neurofibroma, plexiform neurofibroma, and epithelioid neurofibroma. Localized or solitary neurofibroma is found most commonly and develops along a peripheral nerve as a focal mass with well-defined margins but not encapsulated. Localized or solitary neurofibroma is uncommon in infancy and typically appears in late childhood or during teenage years. Most of the isolated or solitary neurofibromas are sporadic, and a small numbers may be associated with the NF-1 syndrome. Most of these arise in the third to fourth decades of life. Neurofibroma involving a major nerve, especially those encased in bone. Soft tissue growths are noted when smaller peripheral nerves are involved. The cause of solitary neurofibroma is still unknown. However, neurofibromatosis is inherited as an autosomal dominant trait with a high degree of penetrance but variable expressivity. Two subsets have been defined: one is associated with the NF-1 (NF1) gene, and the other is associated with the NF2 gene. An estimated 5% of patients with NF1 have an intraoral manifestation of the disease. Discrete neurofibromas may involve the tongue or the larynx[3,4,5].

Case Description:

A female patient aged 27 years has been admitted in the department of Otolaryngology and Head-Neck Surgery, Sher-e-Bangla Medical College Barishal in 6th October 2024 with the complaint of asymptomatic swelling of the right side of anterior half of the tongue measuring 1.5cm x 3.0cm with mild difficulty in speech and swallowing since 02 years. On examination the swelling was oval shaped, solitary, smooth, firm and non-tender. The swelling was gradual in onset and slowly increased in size since 02 years. Patient did not have any symptoms of altered sensation in the region. There was no associated medical illness. Patient doesn't has any history of drug allergy. There was no known evidence of hereditary disease in the family and none of the other relatives had a known history of this disease. A FNAC was done before excision and revealed spindle cell neoplasm. The excised mass was found lobulated and grape like, pale-pink in color[Fig.-03] and it was taken for histopathology and stained with hematoxylin and eosin (H and E). Histological examination revealed a benign neoplasm composed of spindle cells having serpentine nuclei, arranged in bundle and fascicles is a fibrillary background[Fig:4]. With the biopsy we came to conclusion as the neurofibroma of tongue. Probably it arised from Lingual nerve [Fig:02].

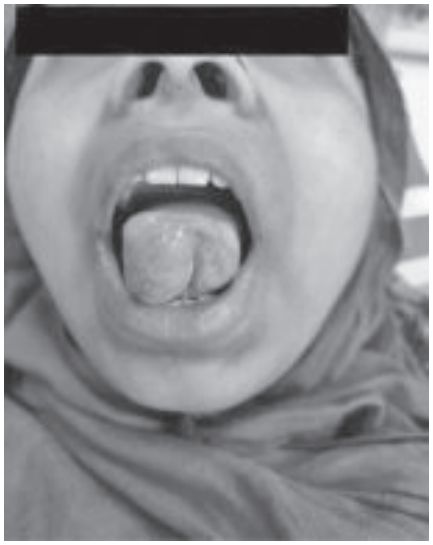

Figure 1: *Close-up view of Tongue Neurofibroma*

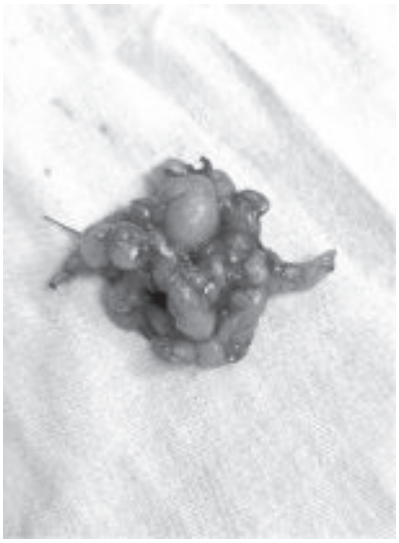

Figure 3: *Specimen (NF of tongue) for histopathology*

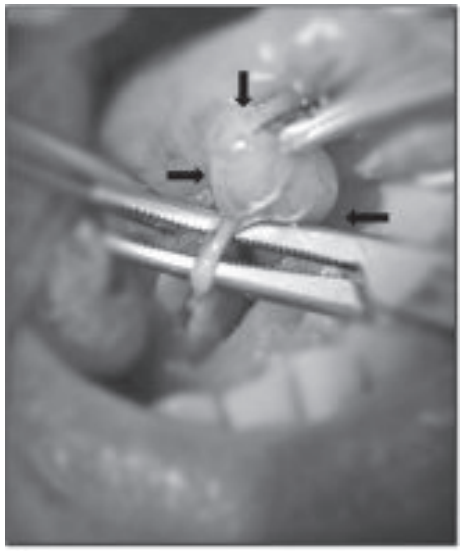

Figure 2: *Surgically excised neurofibroma (oval shape)*

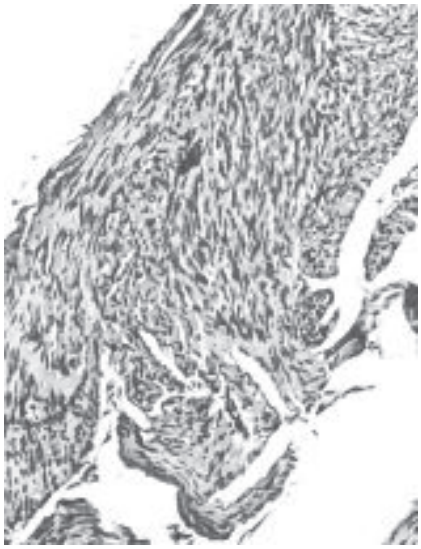

Figure 4: *Haematoxilin and eosin staining of tongue Neurofibroma*

Differential Diagnosis:

In general, diagnostic consideration for tongue lipoma, lymphatic malformation, Mucous retention cyst of tongue. Lipoma is a benign tumor of fat cells. It is usually soft, smooth surfaced nodular masses that can be sessile or pedunculated. These are asymptomatic and often noted for months to years. Clinically they can appear as yellow and deeper lesions may appear pink. Common intraoral sites are buccal mucosa and vestibule; most of the patients are older

than 40 years. Lymphatic malformations are normal lymphatic cell rests that happen to be formed in excess amounts at certain anatomical location during embryonic development. They can be in isolation or in combination with other vascular components. Lymphatic malformation of the tongue is described discretely. As the lesion only grows with the body, radical excision sacrificing healthy tissues is not necessary. Treatment is required when bulk causes mechanical or pressure effects and/or for cosmetic purposes. Mucous cyst are small fluid filled thin sac that usually developed inside the mouth or on the fingers and toes. They are usually smooth or shiny in appearance and bluish-pink in color. It is not associated with any symptoms other than the presence of cyst itself[6,7].

Diagnosis and Treatment:

Lesion was surgical excised under Lingual nerve block by local anesthesia (2% Lidocain with Adrenalin) and specimen was sent for histopathological examination. A longitudinal incision was made over ventral surface of the tongue parallel to the midline. After exposure, the mass was found as grape like structure which invade the tongue musculature. Bipolar diathermy was used for dissection of the the mass. In posterior part it was free from lingual nerve. Histopathology reveals a benign neoplasm composed of spindle cells having serpentine nuclei, arranged in bundle and fascicles is a fibrillary background with no granuloma or malignancy was seen. Histopatho-logically it was diagnosed as neurofibroma. The wound was closed primarily after proper hemostasis. The patient has reported to our hospital after two weeks and six months duration for follow-up and there was no recurrence of the lesion but mild sensory deficit at tip of the tongue [Fig:05].

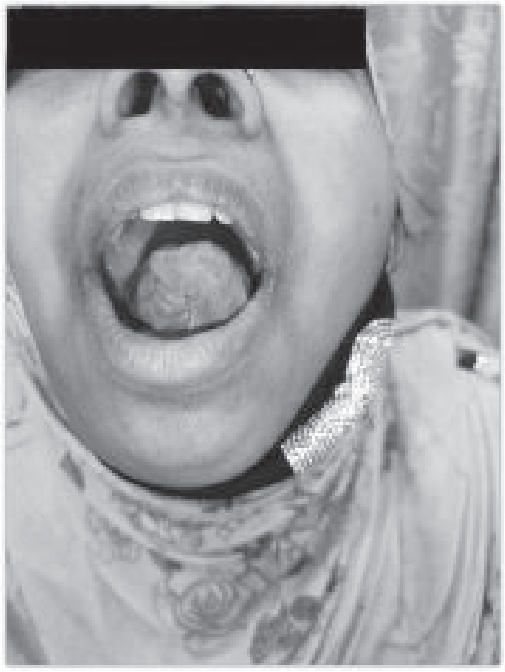

Figure 5: Close view of operated tongue during follow-up after two weeks.

Discussion:

Neurofibroma is a benign tumor of neural tissue origin. It was first described in the year of 1882 by Von Recklinghausen. It most frequently involves the skin and rarely the oral mucosa. The nature of the disease has been recognized as hereditary with an autosomal dominant trait with variable penetrance. Two variant of neurofibromas have been defined:

1. Neurofibromatosis type 1

2. Neurofibromatosis type 2

Incidence of type 1 : (2,500-3,000) births and its prevalence in population is 1 : 5,000 and type 2 incidence is 1 : (33,000 - 40,000) births and its prevalence in population is 1 : 2,10,000.Neurofibromas are derived from the Schwann cells, perineural fibroblasts. Skin lesions appear as soft, drooping and doughy masses. Only 4-7% of patients affected by neurofibromatosis shows oral manifestations. Oral lesions are discrete, nonulcerated nodules, which tend to be of same color of

surrounding mucosa[8,9]. Usually occurs on buccal mucosa, palate, alveolar ridge, vestibule and tongue; also reported in lips and gingival. Intraosseous lesions are quite rare, with the most common site being the posterior mandible. Females are more frequently affected, with the female-to-male ratio being 2:1. Most patients are younger than 45 years of age. In early stages central neurofibromas are asymptomatic, but, as they grow in size, they may cause expansion of the cortical plates of the mandible, with or without destruction, and pain and anesthesia or paresthesia of the lower lip. Radiologically, the tumor appears as a well-circumscribed or poorly demarcated radiolucent lesion, usually involving the mandibular canal. CT or MRI is important to specify the exact location and dimensions of the tumor. Malignant transformation of neurofibroma is reported to occur in 5% to 16% of patients with neurofibromatosis. Malignant transformation is very uncommon in case of Solitary neurofibromas. Neurofibromas have been reported to locally recur after treatment; because the absence of a capsule in neurofibroma, making complete eradication of the tumor more difficult. Even more radical surgery has been proposed to prevent local recurrence[10,11].

Conclusion:

Wound was healed spontaneously within 14 days. After 6 months follow-up there was no recurrence of the disease except mild sensory deficit at tip. Lingual nerve block provides minimum pain sensation, better comfort to patient during surgery. Neurofibroma usually presents with classical features like wide spread soft tissue nodules and skin pigmentations thus making it not so difficult to identify and diagnose.

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