ORIGINAL RESEARCH PAPER
Volume - 12 | Issue - 04 | April - 2023 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

SPORADIC NEUROFIBROMA TONGUE- A CASE REPORT

ENT

Dr.P.V.S.Sai Karthik Junior Resident, ENT, Andhra Medical College

Dr.Ch.Ramesh Kumar Assistant Professor, ENT, Andhra Medical College.

Dr.K.Jeevan Pradeep Professor, ENT, Andhra Medical College.

ABSTRACT

Neurofibromas are the most common benign peripheral nerve sheath tumours.They arise from endoneurium and the connective tissues of peripheral nerve sheaths.Here we present a case of 22year old male with history of a slow growing mass in the left lateral border of tongue.Preop FNAC and CECT were done. Mass excision was done under general anesthesia .Post op histopathological examination confirmed it to be a neurofibroma.Neurofibromas of tongue are a rare occurence1.They may be sporadic or associated with neurofibromatosis syndromes.Along with schwannoma ,fibroma, neurofibroma also forms an important differential diagnosis in the swellings of tongue.

KEYWORDS

neurofibroma, tongue

INTRODUCTION

Neurofibroma is a benign tumor of peripheral nerve sheath phenotype with mixed cellular components including Schwann cells,perineural hybrid cells,and intraneural fibroblasts. Neurofibromas may be sporadic or associated with neurofibromatosis syndromes.Intraorally 6.5% of neurofibromas are sporadic,with tongue being the most common site

CASE DESCRIPTION

A 22year old male patient presented to our OPD with history of a swelling in the left lateral border of tongue which was first observed 4months ago.There was no history of pain,ulceration or bleeding from the site.No other swellings were found anywhere else on the patients body.The patient did not complain of any difficulty in swallowing, chewing, tongue movement, speech, and breathing.There was no history of substance abuse.On examination a single spherical swelling of size 2*2 cm was seen in the tongue along the left lateral border.It was firm and non tender with well defined borders. Mucosa over the swelling was normal.Rest of the oral cavity and oropharynx were normal.There were no palpable lymph nodes in the neck

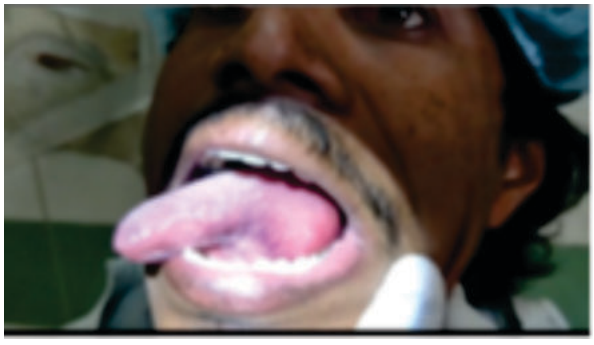

Fig 1:clinical Picture Showing Swelling On Left Lateral Border Of Tongue

On contrast enhanced CT examination a lobulated soft tissue lesion in the left side of tongue measuring 23*21 mm with mild enhancement in post contrast study was noted with possible differential diagnosis as benign salivary neoplasm and neurofibroma. FNAC of the swelling revealed spindle cells with wavy nuclei.There were no mitotic figures.

The mass was excised under general anesthesia and sent for histopathological examination.Post operative histopathology showed randomly arranged spindle cells in a collagenous to myxoid stroma with wavy nuclei-suggestive of neurofibroma.Tissue histochemistry was strongly positive for S-100 protein.The patient was thoroughly examined and ophthalmologist opinion was taken before confirming the diagnosis of solitary neurofibroma.In 6 months of followup there was no recurrence and no other new swellings elsewhere.

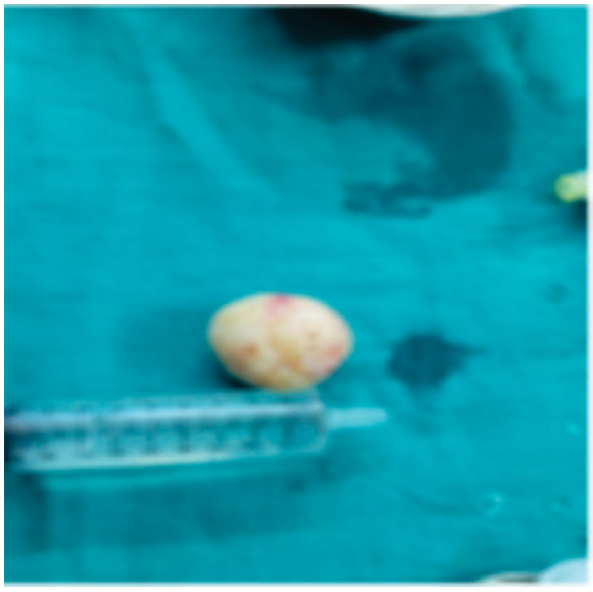

Fig 2:excised Mass

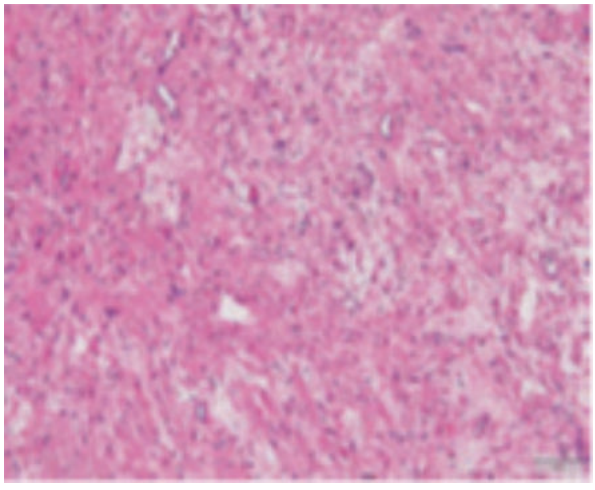

Fig 3: Post Op Histopathology Showing Spindle Cells

DISCUSSION

Neurofibromas present as either a solitary mass or as part of neurofibromatosis syndrome. 25% of all neurofibromas occur in head and neck region.Oral cavity neurofibromas are characteristically associated with multiple lesions elsewhere in the body and are manifestations of neurofibromatosis type 1 (von Recklinghausen syndrome) or neurofibromatosis type 2 (central neurofibromatosis) or Multiple Endocrine Neoplasia Type 3(MEN type III). NF type 1 involves multiple café au lait macules,intertriginous freckling and neurofibromas of skin.There may be associated behaviour and learning abnormalities.About half of patients with NF type 1 have plexiform neurofibromas.The presentation of patients and range of symptoms depend on the site of origin and surrounding vital structures involved.Optic nerve gliomas and sphenoid wing dysplasia are manifestations of NF type 1 commonly seen in head and neck region.Neurofibromatosis type 2 is characterised by schwannomas including bilateral vestibular schwannoma, retinal hamartoma, multiple meningiomas, cataract and mononeuropathy in childhood.Diagnosis of neurofibromatosis depends on characteristic array of symptoms and DNA sequencing for genetic abnormalities.In multiple endocrine neoplasia type 3 features include multiple neuromas, medullary carcinoma thyroid and pheochromocytoma.

Solitary intraoral neurofibroma not associated with NF-I is very rare in the oral cavity[2]. The pathogenesis of solitary neurofibroma not associated with NF-I is poorly understood.Somatic inactivation of NF-1 gene located on chromosome 17 is implicated.Studies suggest that Solitary neurofibroma is a hyperplastic hamartomatous malformations rather than a neoplastic disease[3].

Solitary neurofibroma of oral cavity are slow growing pedunculated or sessile masses.Clinically, when the tumor on tongue is large in size it can cause difficulty in swallowing, mastication, chewing and respiratory obstruction. Impingement on nerve may lead to pain and paresthesia .Intraoral lesions of neural tissues mainly originate from the branches of fifth, seventh, and rarely ninth cranial nerves

Treatment of solitary neurofibroma involves excision with margins free of disease.Neurofibroma is usually unencapsulated,hence leaving tissue during excision may lead to recurrence.During surgery, the surgeon should be wary of the anatomy and location of lingual artery in relation to the mass,as injury to lingual artery may lead to troublesome bleeding.Solitary neurofibroma may rarely , if ever turn malignant.

CONCLUSION

Lesions on the tongue pose a diagnostic challenge as clinically they can mimic a variety of neoplasms.Differential diagnosis include Schwannoma,Fibroma,Neurofibroma,Lipoma,leiomyoma etc[4].Histopathological examination and immunohistochemistry with S-100 protein remains mainstay of diagnosis[5].Complete excision is the treatment of choice.Neurofibromatosis syndromes like Neurofibromatosis 1 (von Recklinghausen's disease) &Neurofibromatosis 2 must be ruled out as they have higher potential to become malignant.

REFERENCES

1. Roy P, Chakraborty S, Das S, Roy A. Solitary Neurofibroma at the Base of the Tongue: A Rare Presentation. Indian J Dermatol. 2015 Sep-Oct;60(5):497-9.
2. S. S, Shashikumar P, H.S. S, G. SK.Solitary Plexiform Neurofibroma of the Gingiva: Unique Presentation in the Oral Cavity.J Clin of Diagn Res.2013; 7(9):2090-2092.
3. Marocchio LS, Oliveira DT, Pereira MC, Soares CT, Fleury RN. Sporadic and multiple neurofibromas in the head and neck region: a retrospective study of 33 years.
4. Lahoz Zamarro MT, Galve Royo A. Neurofibroma de lengua [Neurofibroma of the tongue]. An Otorrinolaringol Ibero Am
5. Pathology of the Head and Neck,springer