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**Case Report**

# NEUROFIBROMA MIMICKING NEURO VASCULAR HAMARTOMA –A CASE REPORT

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**A R T I C L E I N F O**

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**A B S T R A C T**

Neurofibroma is a benign neoplasm of non-odontogenic origin arising from peripheral nerves.They may present as a solitary lesions or in association with syndromic conditions. The solitary neurofibromas of the oral cavity are rare ranging from 2%-7%. Neurofibroma along with numerous feeding capillaries is an unusual presentation in the oral cavity which mimicks the Neurovascular hamartoma. We hereby present a case of solitary nodular palatal swelling of neurofibroma mimicking neurovascular hamartoma in a 13 year old girl. Histological investigation showed proliferation of spindle shaped cells with wavy nuclei along with numerous large and small proliferating capillaries. Toluidine blue staining was done to demonstrate Mast cells. Immunohistochemistry showed positivity for S- 100 and negativity for CD-34 based on which a final diagnosis of Neurofibroma was made.



## INTRODUCTION

Neurofibroma (NF) is a benign neural tumor which originates from the peripheral nerve sheath and it is composed of a variable mixture of Schwann cells, perineural cells, and fibroblasts**.** It can present either as a localized lesion or as part of generalized syndrome known as Neur of ibromatosis type1 (NF-1) orVonRecklinghausen's disease.[1] They typically arise in the second or third decade of life as asymptomatic, slow growing pedunculated or sessile nodular growth. Neurofibroma may involve any part of oral cavity, most common site being the tongue followed by buccal mucosa, floor of mouth, lips, and gingiva.[2]

Neurovascular hamartoma refers to a benign malformation, do not develop as part of an inflammatory orneoplastic process and have a self limiting proliferation. They present as smooth, painless pinkish nodular or pedunculated masses.The histology consists of proliferationof the nerve bundles along with numerous proliferatingcapillaries. Oralneurovascular hamartoma is rare because of proliferation of neural tissue solely or incombination with vascular elements.[3]

Neurofibroma along with numerous feeding capillaries is an unusual presentation in the oral cavity which mimicks the Neurovascular hamartoma.We report a rare case of solitary NF of the palate with numerous feeding capillaries mimicking Neurovascular hamartoma in a 13 year old girl.

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### *Case Presentation*

A 13year old girl presented with the chief complaint of a painless swelling in her right palatal region for the past one year.On intraoral examination there was a pinkish solitary nodular swelling measuring 2x1 cm in diameter on her right palatal region. On palpation it was firm and non-tender. The mucosa over the swelling was intact and nonulcerated (Fig 1).There was no history of trauma and the medical and family history was insignificant. Fine Needle Aspiration (FNA) was negative. A provisional diagnosis of Fibroma was made and surgical excision of the lesion was done. The excised specimen was sent for histopathological examination (Fig 2).

The H&E stained histopathologicalsections showed myxoid connective tissue stroma interspersedwith numerousspindle shaped cells having wavy nuclei along withlarge and small proliferating blood capillaries.The spindle shaped cells were seen proliferating in the form of bundles as well as in diffuse pattern(Fig 3,4,5,6). The stroma was collagenous with mild inflammatory infiltrate. Based on the histopathological evaluation, a differential diagnosis of "neurofibroma"and "neurovascular hamartoma" was considered.

To rule out Neurovascular hamartoma, Toulidine blue staining was done to demonstrate Mast cells (Fig 7). Further IHC for S-100 showed strong positivity along the nerve fibers (Fig 8) andCD-34 was negative (Fig 9), which ruled out neurovascular hamartoma. Based on the above findings the final diagnosis of Neurofibroma was made.After a follow up of 6 months patient revealed no recurrence.

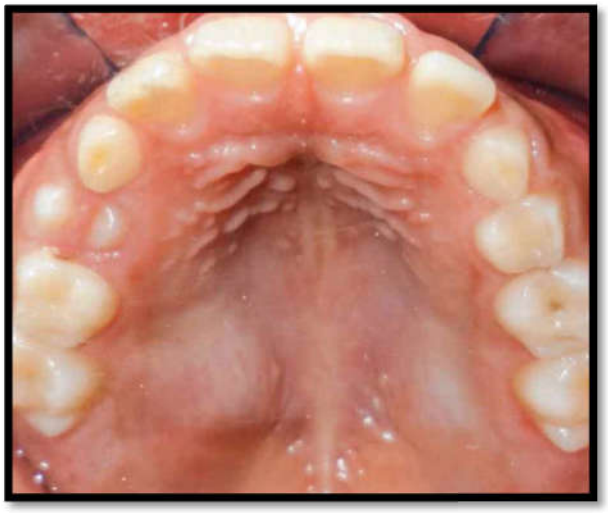

**Fig 1**

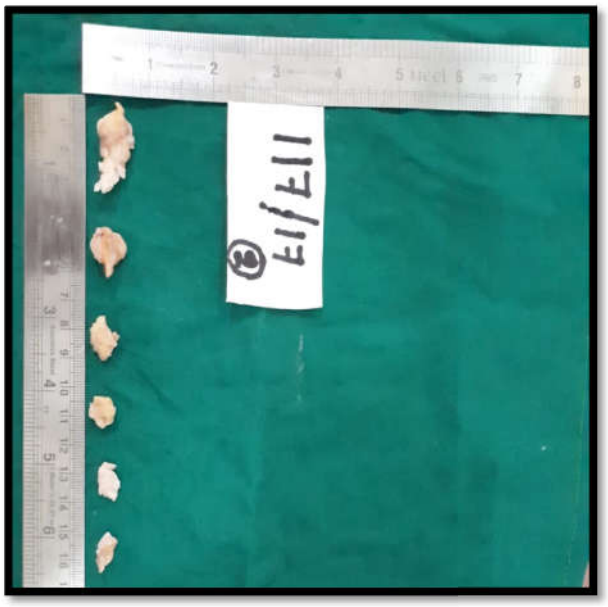

**Fig2**

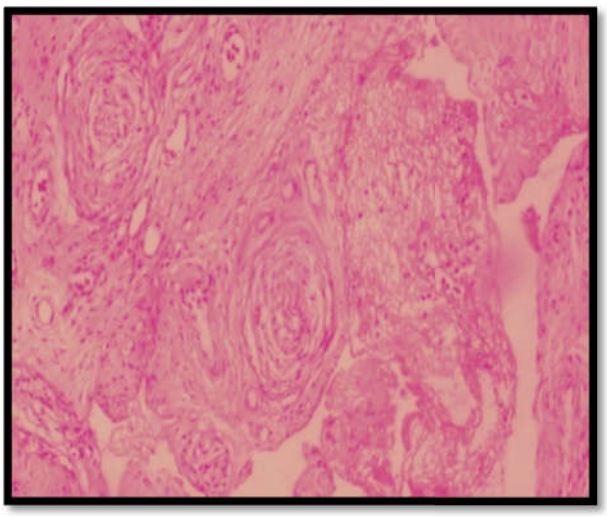

**Fig 3**

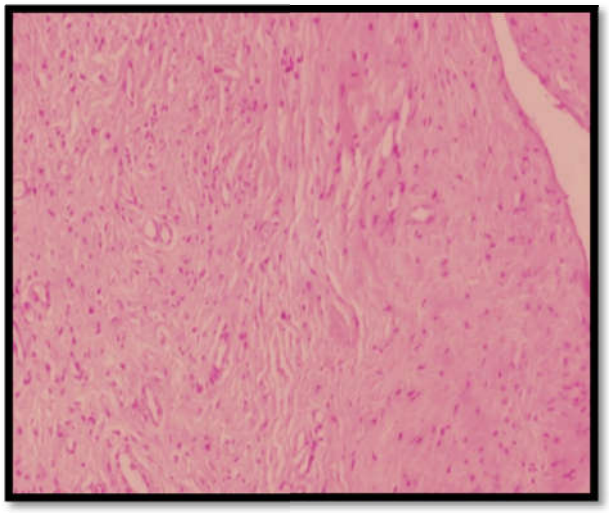

**Fig 4**

**Fig 1** Intraoral picture of the lesion**, Fig 2** Gross image of the lesion, **Fig 3 and 4** histopathological picture showing the connective tissue stroma interspersed with numerous spindle shaped cells having wavy nuclei along with large and small proliferating blood capillaries (H &E, x 400)

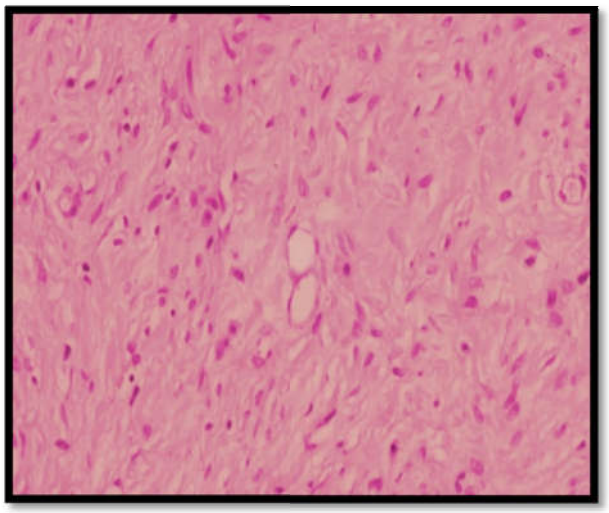

**Fig 5**

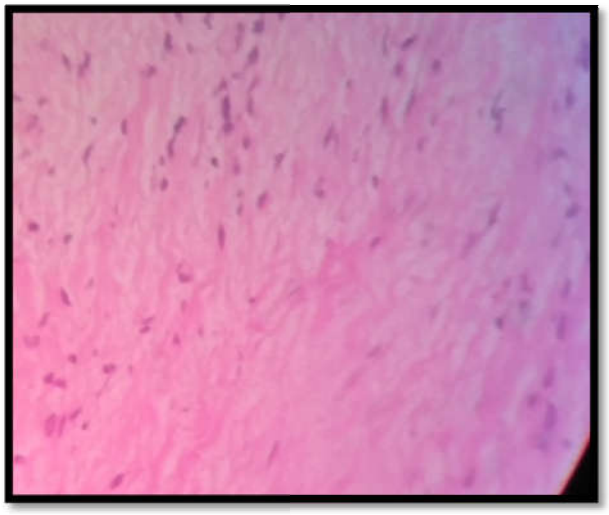

**Fig 6**

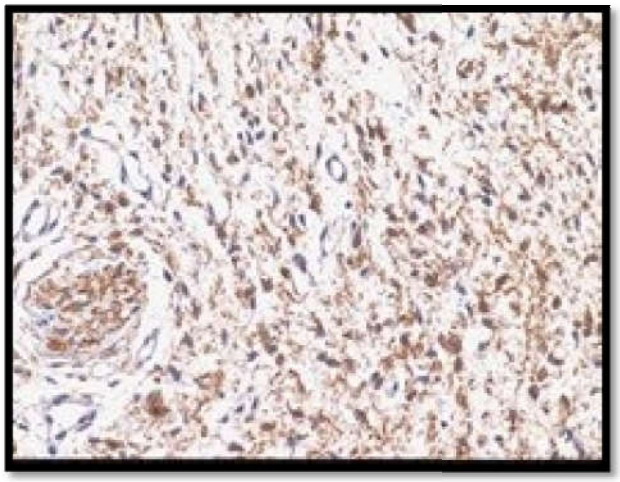

**Fig 7**

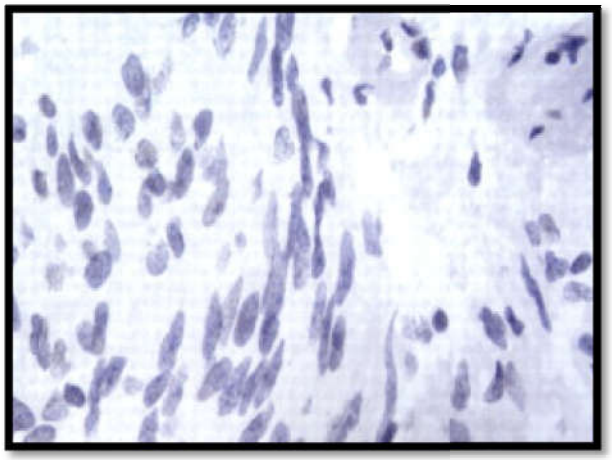

**Fig 5 and 6**: Histopathologic picture showing spindle shaped cells proliferating in the form of bundles as well as in diffuse pattern(x 400).
**Fig 7 and 8:** IHC showing strong positivity for S-100 and Negativity for CD-34. (x 400)

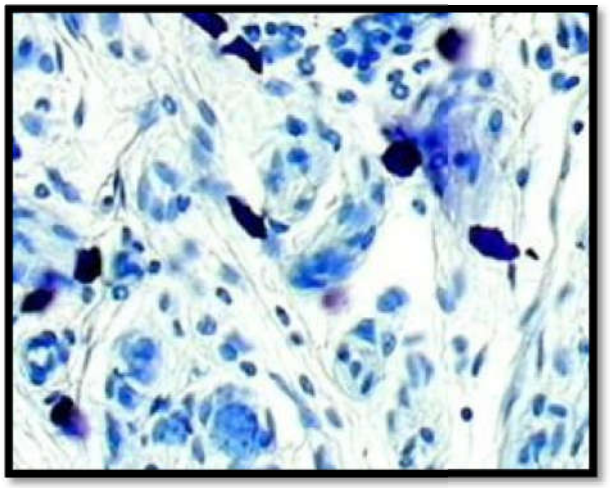

**Fig 9** Toluidine blue stain demonstrating mast cells. (x 400)

## DISCUSSION

Neurofibroma is a benign nervetissue tumor, with the reported incidence of 37% in the head and neck region. Neurofibromas affecting oral cavity arerare, ranging from 2% to 7%. Most of the cases are associated to be a part of multiple neurofibromatosis.Solitary NF is a rare variant of NF, they are distinct entities as they lack the systemic and hereditary components.[4] This type of NFs occurring in the palate are extremely rare with only 6 cases reported so far in the literature.We present a case of solitary neurofibroma of the palate.

Solitary neurofibroma can present at any age ranging from 10 months to 70 years, but most commonly occur during the 3rd to 4th decade of life, no racial or gender predilection has been described.Exact etiology of these lesions are unknown.[5]

The oral NFs usually present as a slow growing painless swelling which are either sessile or pedunculated with nonulcerated mucosa. The color of the overlying mucosa varies from normal mucosal color to red or sometimes yellow. The differential diagnosis of these lesions when they occur in hard palate includes fibroma, schwannoma, traumatic neuroma, and salivary gland tumor.[6]

Histopathologically they are mostly unencapsulated and show diffuse proliferation of spindle shaped cells with wavy and serpentine nuclei separated by abundant fine collagen fibres.[7] Mastcells are typically found and they are believed to be the inciting factor contributing to fibroblastic proliferation and growth of neurofibroma.[8]

In our case due to the presence of numerous large and small proliferating blood capillaries along with the spindle shaped cells, it mimicked neurovascular hamartoma, hence IHC was done which showed strong positivity for S-100 and CD-34 was negative, which helped to rule out neurovascular hamartoma and arrive at a final diagnosis of Neurofibroma.

This case is unique as it occurred as a solitary lesion in the hard palate and the presentation of the lesion was sporadic without any associatedfamily history, the histopathology of the lesion also showed numerous proliferating capillaries which is an unusual feature of Neurofibroma.

The treatment for solitary NF is complete surgical excision. Recurrence is common due to nonencapsulated and infiltrative nature of the disease, 20% of the patients show recurrence after complete resection.[9]Malignant transformation is rare, which is estimated to occur in about 5% of cases.The most common associated malignancy is neurofibrosarcoma.[10] Therefore follow up after surgery is important, this patient was followed up for six months with no recurrence.

## CONCLUSION:

Solitary Neurofibroma is a rare tumor of oral cavity, which has to be differentiated from other neural lesions. In our case as the lesion had histopathologic similarity with neurovascular hamartoma, toluidine blue staining and immunohistochemistry was needed for the confirmatory diagnosis of neurofibroma. These lesions show good prognosis and follow up is required to look for recurrence.

## Reference

1. Priya M, Bakshi SS, Coumare VN, Vijayasundaram S, Latheef MN. Solitary extraosseous neurofibroma of hard palate: Report of a case with a review of literature. *J Dent Allied Sci* 2016; 5:95‑7.
2. Sahni P, Singhvi A, Nayakm.Solitary Neurofibroma of Tongue: A Rare Case Report.Dental Impact 2012; 4(1).
3. Kreiger PA, Ernst LM, Elden LM, *et al*. Hamartomatous tongue lesions in children. *Am J SurgPathol* 2007; 31:1186–90.

4. Thimmaiah AH, Santharam L, Gangaraj SS, Vijay S. Neurofibroma of soft palate: A rare case report. *OtorhinolaryngolClin* 2013; 5:109‐10.
5. Cherrick HM, Eversole LR. Benign neural sheath neoplasm of the oral cavity. Report of thirty‐seven cases. *Oral Surg Oral Med Oral Pathol* 1971; 32:900‐9.
6. Jangam SS, Ingole SN, Deshpande MD, Ranadive PA. Solitary intraosseous neurofibroma: Report of a unique case. *ContempClin Dent* 2014; 5:561‐3.
7. Mahmud A,,Chattaraj M,Solitary Encapsulated Neurofibroma Not Associated with Neurofibromatosis-1 Affecting Tongue in a 73-Year-Old Female,*J Case Reports in Dentistry* 2016, 34-38.
8. Viskochil HD, It takes two tango: mast cell and Schwann cell interactions in neurofibromas,*J Clin.Invest*, 2003;112:1791-1793.
9. Needle MN, Cnaan A, Dattilo J, Chatten J, Phillips PC, Shochat S, *et al*. Prognostic signs in the surgical management of plexiform neurofibroma: The Children's Hospital of Philadelphia experience, *J Pediatr* 1997;131:678‐82.
10. Hope DG, Mulvihill JJ. Malignancy in neurofibromatosis. *J Adv Neurology*, 1981;29:33‐56.

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