CASE REPORT

Plexiform Neurofibroma: A Rare Tumor of Submandibular Salivary Gland

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Received on: 14 July 2025; **Accepted on:** 06 December 2025; **Published on:** 15 January 2026

ABSTRACT

Plexiform neurofibroma is more commonly seen in the orbit, neck, back, and inguinal region. It is extremely rare in the submandibular gland. These lesions rarely transform into malignancy but are locally infiltrative and can lead to haemorrhage. Therefore, plexiform neurofibroma should always be considered during differential diagnosis while excising a submandibular gland mass. We present here a case of plexiform neurofibroma in the submandibular gland and diffuse neurofibroma in the floor of the mouth in a 15-year-old male, not associated with neurofibromatosis-1.

Keywords: mouth, neurofibroma, Plexiform, submandibular gland.

INTRODUCTION

Neurofibroma is a disease of the peripheral nervous system and occurs most commonly in the extremities. Several forms have been described: cutaneous neurofibromas (both localised and diffuse types), intraneural neurofibromas (localised and plexiform), massive soft tissue neurofibromas (solitary or multiple), and sporadic neurofibromas or those associated with neurofibromatosis-1 (NF-1).[1,2] Neurofibromas constitute only 0.4% of all salivary neoplasms. Plexiform neurofibroma is usually recognised as a pathognomonic criterion of NF-1 (or Von Recklinghausen's disease); it may also occur as a solitary lesion arising in a nerve root.[3] Plexiform neurofibromas of the salivary glands are rare, often presenting in the parotid gland. They are very rare in submandibular salivary gland. Till date, very few cases have been reported in the literature, and much fewer which are not associated with NF-1.[4,5] We are presenting this case because of its rarity of presentation.

CASE REPORT

15-year-old boy presented with a swelling in the left submandibular region and floor of the mouth with duration of 3 months which gradually increased in size. There was no history of pain or increase in size of swelling while taking meal. On physical examination, there was a non-tender mass, firm in consistency, not attached to the skin, measuring 5 × 4 cm in the left submandibular region (Figure 1) and 2.5 × 1 cm in the floor of the mouth (Figure 2). No nerve involvement was observed. It was bidigitally palpable. On systemic examination, there was no swelling or café-au-lait spots all over the body. A family history of NF was absent. Laboratory investigations were within normal limits. An ultrasound of both masses revealed heterogenous echogenecity with ill-defined borders. FNAC was suggestive of Chronic Sialadenitis.

CECT Neck had revealed a large lobulated heterogenous lesion measuring approx. 7.1cm *(CC)* 2.8cm * (AP) * 3.5 (TR) with multiple hypodense non enhancing cystic spaces within noted in left sublingual space, extending to left submandibular gland and floor of the mouth. On post contrast study the lesion showed heterogenous enhancement through which vessels were noted to pass with possibility of venolymphatic malformation. CE -MRI neck with oral cavity had revealed a lobulated soft-tissue lesion of 8cm *3cm*3.8cm involving the left sublingual space, left submandibular gland and the

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Source of funding: None

Conflict of interest: None declared

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How to cite this article: Sharma A, Sharma U, Deka D, Kakoti K Sharma A, Sharma U, Deka D, Kakoti K. Plexiform Neurofibroma: A Rare Tumor of Submandibular Salivary Gland. Journal of Otolaryngology and Head & Neck Surgery. 2026; 1(3):18-20

floor of the mouth. Medially involving the left genioglossus and myelohyoid muscle. Laterally invaded the masticator space closely abutting left parotid gland and encased the left pterygoid muscle, extending into subcutaneous tissue layer (Figure 3, 4). After completion of routine investigations and required imaging we had planned for excision of the lesion under General Anesthesia.

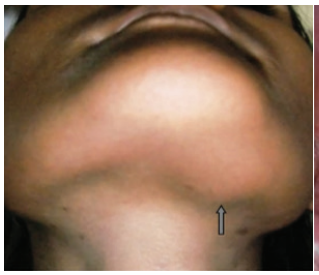

Fig. 1: Left Submandibular Swelling

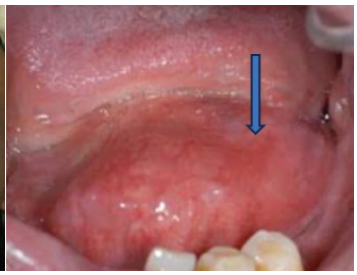

Fig. 2: Intraoral swelling in the floor of the mouth

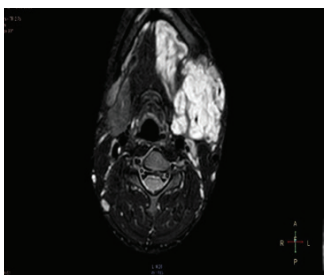

Fig. 3: CE-MRI Neck with oral cavity Image (axial cut)

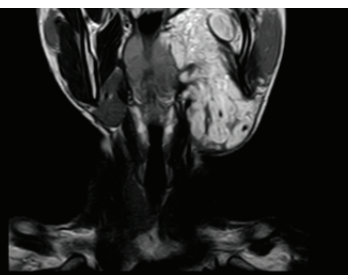

Fig. 4: CE-MRI Neck with oral cavity Image (coronal cut)

Intraoperatively, mass was arising from submandibular salivary gland. A tubular growth was extending from the mass into the adjacent tissue. Gross specimen was gelatinous and vaguely nodular, showing greyish white to grey yellow areas (Figure 5). Microscopic picture showed cellular and nodular area of tumor tissue. Central nerve fiber bundles surrounded by neurofibroma tissue were seen in an abundant mucoid matrix and collagen background. (Figure 6) . Adjacent salivary gland also showed thickened nodular nerve bundles between salivary lobules. No atypia was present. Features were suggestive of plexiform neurofibroma of the submandibular salivary gland.

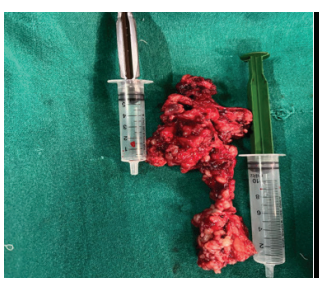

Fig. 5: Gross appearance of the tumor

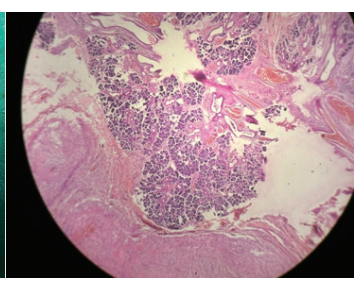

Fig. 6: Microscopic picture of tumor showing central nerve bundles.

DISCUSSION

Neurofibromas are benign nerve sheath tumors which are present in the following three forms: local discrete, generalized neurofibromatosis, and plexiform neurofibromas. Common sites of occurrence of plexiform neurofibroma are fifth cranial nerve and extremities. Neurofibromas constitute only 0.4% of all salivary neoplasms. Plexiform neurofibromas of the salivary glands are rare, usually presenting in the parotid gland. They are very rare in submandibular salivary gland.

Plexiform neurofibromas are diffuse enlargements of multiple fascicles of the nerves and its branches, leading to thickening of nerves. They grow along nerves extending into the surrounding tissue. They are slow growing and locally infiltrating benign tumors. When located deeply, they may have greater chance of malignancy. Large plexiform neurofibromas of submandibular salivary gland may be associated with pressure symptoms on trachea and pharynx. Sometimes they may extend into spinal canal and compress the spinal cord. They grow rapidly in children, during puberty, and in pregnancy. Plexiform neurofibromas are frequently seen in patients with Type I neurofibromatosis and undergo malignant changes in 2% of the cases. Patient with plexiform neurofibroma without family history or without features of Type I neurofibromatosis require genetic work up. In such cases, tumor may present as a result of local somatic mutation and patient may not transmit the disease to their offspring.[5] Neurofibromas have been reported in parotid glands.[6]

Of 300 neurofibromas in the minor salivary gland tumors, two were found to be plexiform neurofibromas.[7] Plexiform neurofibromas are exceedingly rare in submandibular and sublingual salivary glands.[8] A search in the literature revealed only few cases of plexiform neurofibromas of submandibular salivary gland.

If we consider FNAC to diagnose plexiform neurofibroma, its diagnostic yield appears to be low. A plexiform neurofibroma may be suspected in the appropriate setting with the help of cross sectional imaging. Computed tomography (CT) imaging shows conglomerate multilobulated masses that may appear as a 'bag of worms.' There is hypoattenuation with heterogenous contrast enhancement. A characteristic 'branching' hypoattenuated mass on CT, with branching tubular masses extending into adjacent tissue, could virtually be diagnostic.[9] Histopathologically, there is diffuse cylindrical enlargement of multiple fascicles of the nerve and its branches. There is a myxoid matrix containing Schwann cells, nerve fibers, mast cells and perineurial, endoneurial fibroblasts. Surgical excision is the treatment of choice. Recurrence may occur. It is reported in 20% of the patients with plexiform neurofibroma after complete resection and increases to 44% with incomplete resection (where there is involvement of vital structures.[3]

CONCLUSION

The diagnostic yield of fine-needle aspiration of plexiform neurofibroma of the submandibular gland is low, hence diagnosis is based on histopathological examination. In a solitary neurofibroma occurring without stigmata of NF-1, the tumor probably represents the segmental form of NF-1 caused by a later somatic mutation. In adults, plexiform neurofibroma should be considered in differential diagnosis of isolated swellings, although it is of neurogenic rather than salivary gland origin. Follow-up of the patients is necessary due to the possibility of recurrence and malignant transformation. Genetic counselling and evaluation of other family members should be performed for those suspected to be affected by the syndrome.

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