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A Solitary *Plexiform Neurofibroma* Mimicking *Parotid Tumor*: A Rare Case Report

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ABSTRACT

Our case report describes an uncommon occurrence of a solitary plexiform neurofibroma in the salivary gland, which is generally associated with neurofibromatosis-1 (NF-1) but, in this case, arose independently. A 6-year-old girl presented with a painless, progressively enlarging mass in the right parotid region over a span of five years. Histopathological analysis revealed a variety of cellular elements, such as Schwann cells, perineurial cells, axons, and fibroblasts, with S100 immunohistochemistry confirming the diagnosis. This case highlights the importance of identifying solitary plexiform neurofibromas in the salivary gland, even without NF-1, and stresses the need for prompt diagnosis and treatment for optimal management.

Keywords: Head and neck-cancer; Plexiform neurofibroma; Parotid malignancy; Neurofibromatosis type 1

NOMENCLATURE

PN : Plexiform neurofibroma
IHC : Immunohistochemistry

1. INTRODUCTION

Neural-origin tumors in salivary glands are extremely rare, making up only 0.401 % of all salivary gland tumors. Plexiform Neurofibromas (PN), benign tumors from Schwann cells, are rarely found in salivary glands. They are more commonly observed in regions such as the ocular area, neck, back, and inguinal zones. These tumors usually present as single or multiple lesions and are often linked to neurofibromatosis-1 (NF-1).Due to their diffuse and infiltrative growth pattern that affects multiple nerve bundles and adjacent tissues, the complete surgical removal of plexiform neurofibromas is notably challenging [2].

Although PN is a rare manifestation of NF-1 and is pathognomonic of this condition, its isolated occurrence is even less common[3]. Here, we describe a case of a PN presenting as a parotid swelling in a 6-year-old female, who showed no other features of NF-1.

2. CASE REPORT

A 6-year-old female presented to the ENT OPD with swelling in the right infraauricular region, slowing growing over the course of 5 years. Onexamination, a right-sided infraauricular swelling measuring 7x 8x 5 cm was observed.

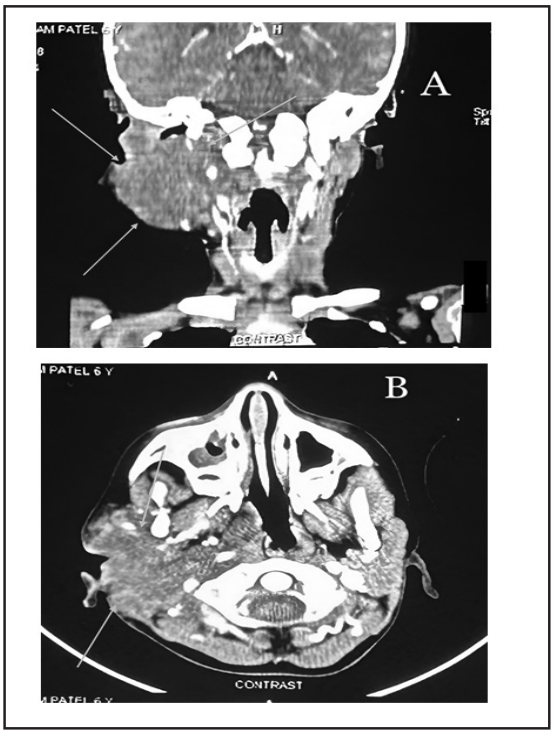

Figure 1. Contrast enhanced computed tomography scan coronal (A) and axial (B) cut - ill-defined heterogeneously enhancing mass arising in the Rparotid gland without any bony involvement or intracranial extension, anteriorly extending in the mastercator space; posteriorly till the right stemocleidomastoid; lateral to right external audiotory canal; medially reaching till the right parapharyngeal space.(as depicted via the arrows in yellow).

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The swelling was non-tender, Rubbery in consistency, and the surrounding skin appeared non adherent with no scars or sinuses. There was no family history of neurofibromatosis or any other genetic condition. The CECT scan demonstrated heterogeneously enhancing mass lesion, measuring 7 x 8 x 6.1 cm, with distinct margins originating within the right parotid gland, involving both the superficial and deep lobes without any intracranial extension or bony involvement (Fig. 1).

Fine-needle aspiration was inconclusive. After informed consent was obtained, the patient underwent a right total parotidectomy. Intraoperatively, a well-defined, pinkish-purple mass was found closely adhering to the lower trunk of the facial nerve. The mass was carefully dissected and removed with all necessary precautions.(Fig. 2).

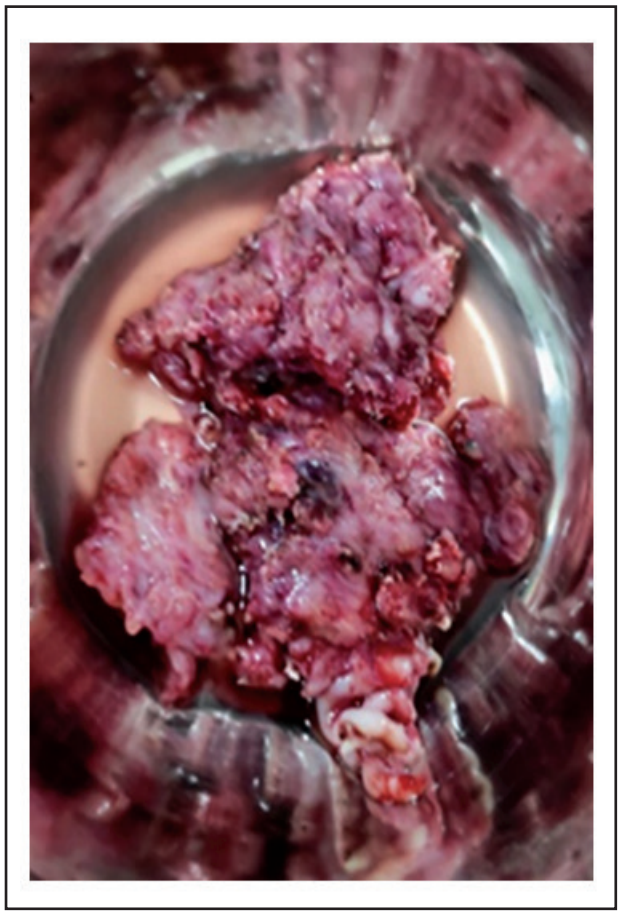

Figure 2. Intraoperative specimen following total parotidectomy.

Gross examination revealed a reddish brown-colored, firm mass measuring 7 × 8 × 6 cm with irregular, tumor nodules within which convoluted nerve bundles are present. Histopathological analysis at low magnification revealed multiple thickened nerve bundles interspersed with normal serous salivary gland tissue. Higher resolution and magnification revealed convoluted nodules showing peripheral spindle shaped cells with wavy comma shaped nuclei and eosinophilic cytoplasm and show a central myxoid stroma with collagen fibrils and palisading nerve fibres (Fig. 3).

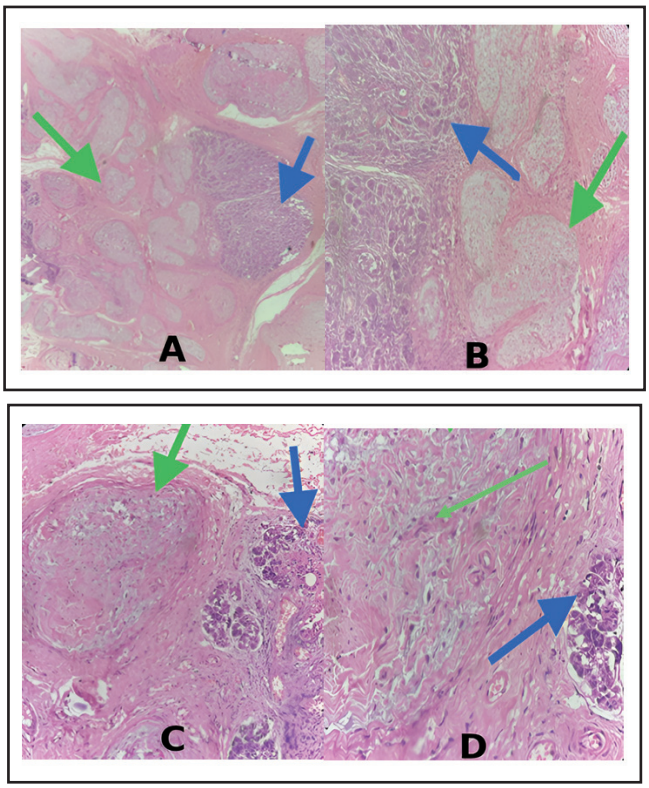

Figure 3. Haematoxylin and eosin-stainedslides, scanner view (A) (4x magnification) show multiple nodules (green arrow) of varying sizes and are seen enclosing lobules of salivary gland (blue arrow) tissue, a lymph node and within peripheral adipose tissue. (B) At 10x magnification showing better delineation of nerve bundles arranged in plexiform pattern (green arrow) surrounded by normal salivary glands(blue arrow). (C) at 20x magnification and (D) at 40x magnification these nodules show peripheral spindle shaped cells with wavy comma shaped nuclei eosinophilic cytoplasm and show central myxoid stroma with collagen fibrils and palisading nerve fibres (green arrow). salivary gland (blue arrow)shows lymphoplasmacytic infiltrates.

S100 immunostaining revealed both nuclear and cytoplasmic positivity, confirming the tumor's neural origin. Consequently, the diagnosis of PN was confirmed through histopathological examination, including IHC(immunohistochemistry).

Following surgery, patient had a smooth recovery and was discharged in good condition, with only mild marginal mandibular nerve palsy. The patient was monitored for six months post-surgery, with no signs of recurrence.

3. DISCUSSION

Neurofibromas are benign nerve tumors that can appear as solitary or multiple growths, occurring sporadically or with neurofibromatosis types I or II. The WHO classifies them as grade I tumors, either dermal, affecting a single nerve, or plexiform, involving multiple nerve bundles[3].

PN account for 15 % of benign mesenchymal tumors and 11 % of nonepithelial salivary gland tumors. They typically grow slowly, infiltrate surrounding tissues,

and are generally painless[5]. Clinical symptoms vary based on the tumor's location and can include pain and neurological deficits. Plexiform neurofibromas in the parotid gland are extremely rare, with benign tumors like pleomorphic adenomas being more frequently encountered in this region[5].

As a significant feature of neurofibromatosis 1 syndrome, plexiform neurofibromas require careful monitoring for specific clinical signs indicative of the condition. Histopathologically, plexiform neurofibromas are characterised by a diffuse tubular enlargement of nerve fascicles, hypocellularity, and a matrix containing Schwann cells and other components. Imaging techniques such as computed tomography can assist in diagnosing these tumors, which appear as non-encapsulated, tortuous nodular growths along nerve branches[6-7].

Surgical excision is the primary treatment for plexiform neurofibromas. Recurrence rates are approximately 20 % after complete resection and up to 44 % following subtotal removal[8]. It is crucial to monitor for signs of malignant transformation, such as rapid growth or changes in consistency, as there is a 2–5 % risk of malignancy in plexiform neurofibromas. Histopathological examination, including IHC for S100, is vital for confirming the diagnosis. Understanding the clinical and pathological characteristics of plexiform neurofibromas is essential for effective management and ensuring optimal patient outcomes[9,10,11].

4. CONCLUSION

In conclusion, the presence of plexiform neurofibromas in atypical locations such as the salivary glands underscores the importance of thorough evaluation and consideration of less common aetiologies in cases of isolated glandular swellings. The association of these tumours with neurofibromatosis type 1 (NF-1) emphasizes the need for comprehensive genetic investigations in affected individuals. Early detection and vigilant monitoring are crucial in patients with NF-1-associated PN, as they face an elevated mortality risk. Long-term follow-up is essential to track disease progression, detect recurrences, and identify potential malignant transformations. Maintaining a proactive surveillance approach enables healthcare providers to deliver personalized care, optimize outcomes, and ensure timely interventions for individuals with plexiform neurofibromas.

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CONTRIBUTORS

Dr. Ashfaque A. Ansari serves as the Professor and Head of the Department of ENT at MGM Medical College. With a distinguished career marked by numerous accomplishments, His expertise and leadership were pivotal in the execution

of this study. His extensive experience in ENT surgeries and patient management provided critical guidance throughout the research process.
Dr. Ansari also played a key role in overseeing the surgical procedures and ensuring adherence to the highest standards of medical practice.

Dr. Mahendra Katre is an Associate Professor in the Department of ENT at MGM Medical College. With a solid foundation in head and neck surgery, Dr. Katre brought invaluable expertise to the study. He served as a primary operating surgeon alongside Dr. Ansari and was instrumental in the successful completion of the surgical aspects of the research.
Dr. Katre's contributions were essential in refining the surgical techniques and providing mentorship to junior researchers involved in the project.

Dr. Lovee Gupta is currently a Junior Resident Doctor in the Department of ENT at MGM Medical College. Dr. Gupta was responsible for the meticulous preparation of the manuscript. Her role encompassed compiling data, conducting comprehensive analysis, and drafting the initial and subsequent versions of the manuscript. Dr. Gupta's dedication to detail and thorough understanding of the subject matter were crucial in presenting the findings accurately.
She coordinated closely with the other authors to ensure the manuscript's integrity and quality.