International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Isolated Neurofibroma of the Soft Palate-An Uncommon Case Study

Dr Anjali Vinod[1], Dr. Suchit Roy B R[2]

[1]Senior Resident, Department of ENT, Government Medical College, Thiruvananthapuram, Kerala, India

[2]Professor, Department of ENT, Government Medical College, Thiruvananthapuram, Kerala, India

Abstract: ***This paper presents a unique case of an isolated neurofibroma in the soft palate, a rare occurrence not associated with systemic pathologies such as Von Recklinghausens disease or Multiple Endocrine Neoplasia. This case, the sixth of its kind reported in English literature, involve a 42-year old male patient who underwent successful surgical excision of the tumour. The patient has remained stable post-surgery, with no signs of recurrence or systemic involvement. This case study contributes to the limited body of knowledge on isolated neurofibroma of the soft palate and underscored the importance of considering such rare diagnoses.***

Keywords: Isolated Neurofibroma, Soft Palate, Surgical excision, Rare Tumours

1. Introduction

Neurofibromas are relatively common benign tumors that are composed of neuromesenchymal cells, including schwann cells, perineurial cells, fibroblast and mast cells. Multiple neurofibromas are found as part of neurofibromatosis type I (von Recklinghausen's disease), an autosomal dominant condition characterised by multiple neurofibromata, cafe´-au-lait patches and lisch spots. Neurofibromas are evenly distributed over the body surface[1]. Neurofibromas can be broadly classified into localised neurofibroma, which arise from a single nerve fascicle and plexiform neurofibroma which are associated with multiple nerve bundles, commonly growing within the trunk of the nerve. Less commonly, multiple neurofibroma can be restricted to a single body part, known as segmental neurofibromatosis. The solitary neurofibroma is a benign, slowly growing, relatively circumscribed but non-encapsulated tumour and it is usually diagnosed by the absence of the other features of neurofibromatosis and is common in young adults without gender preference. Although neurofibroma has predilection for head and trunk isolated cases of solitary palmar neurofibroma has also been reported [2].

Approximately, 25% of all neurofibromas are found in the head and neck region of this 6.5% occur in the oral cavity according to literature reports[3]. In oral cavity the preferential site is tongue, followed by buccal mucosa and lips, the other affected sites are gingiva, palate, major salivary glands.

Here, we report an interesting case of a 42 year old male with solitary neurofibroma in the soft palate which was clinically diagnosed provisionally as pleomorphic adenoma. The patient had no systemic manifestations of neurofibromatosis.

2. Case Report

A 42 year old male patient presented in ENT OPD with a swelling in the oral cavity of 3 months duration, associated with muffled voice. There was no respiratory difficulty, difficulty in swallowing. There was no history of similar illness in his family

Examination of oral cavity showed a 3 x 2 cm pale pinkish swelling on the left side of soft palate. It was firm in consistency, non tender, non fluctuant and non pulsatile swelling. General examination was done and no other swellings suspicious of systemic involvement were noted.

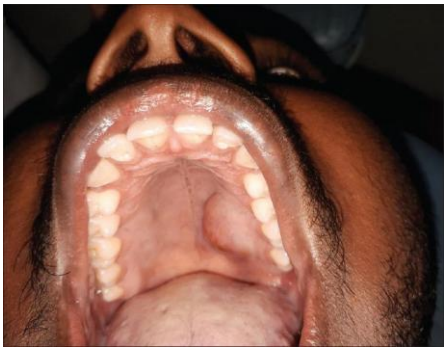


Figure 1: 3 x 2 cm swelling on the left side of soft palate

The routine laboratory tests were normal. A contrast enhanced computed tomography of paranasal sinus was taken and it showed a well defined homogenous fluid density cyst in the left side arising from anterior portion of soft palate with no erosion of hard palate. Differential diagnosis were 1) epithelial cyst 2) epidermoid cyst of soft palate

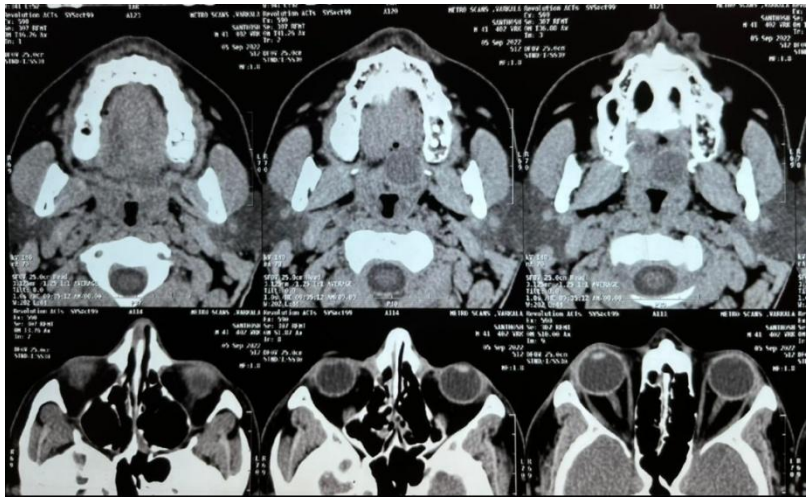


Figure 2: Axial section of CT PNS showing well defined homogenous fluid density cyst of 1.6 x 1.8 x 1.5 cm (CC x transverse x AP) in left side arising from anterior portion of soft palate with rim enhancement

A Fine needle aspiration cytology of the swelling was done which showed a few epithelial cell clusters in a background of mucinous material and stromal fragments with possibility of a benign salivary gland tumour possibly pleomorphic adenoma.

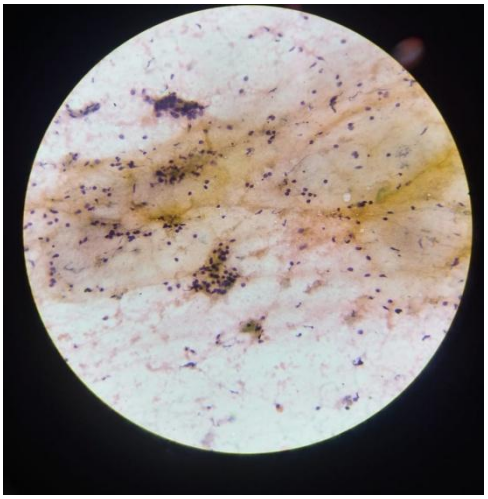


Figure 3: Epithelial cell clusters in background of mucinous material and stromal fragments

Under general anesthesia and orotracheal intubation surgery was done, with patient in Roses position mouth was opened using Boyle Davis mouth gag and the tumour was visualised. Vertical incision was put over the swelling and the mass was enucleated and removed in toto. The wound closed in layers using 3-0 vicryl. Post operative period was uneventful.

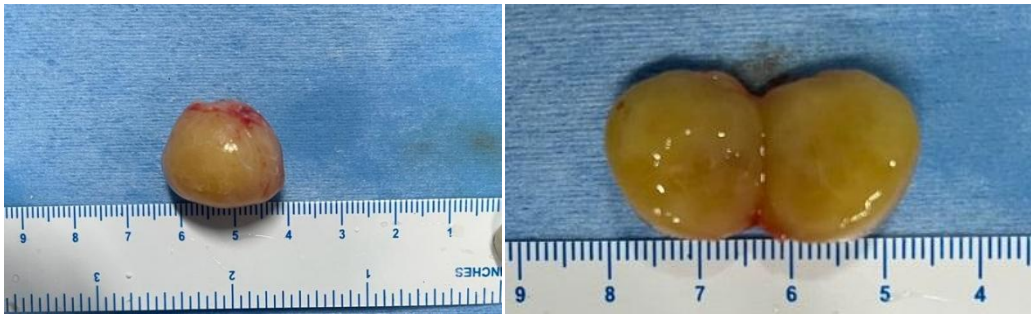
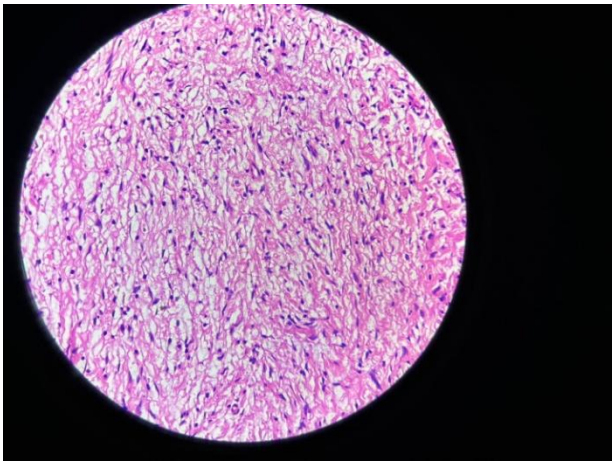


Figure 4 & 5: Gross surgical specimen after excision of approximatly2 cm and cut section of the same specimen

Histopathology report came as benign spindle cell neoplasm compatible with neurofibroma with IHC showing S100 positive. The patient was followed up for 6 months and there was no recurrence and also dermatology and medical neurology consultations were done to rule out systemic involvement and turned out to be negative.



H

Figure 6: H & E staining showing a spindle cell tumour with a mucinous stroma containing numerous mast cells. Occasional foci of rudimentary nerve bundles seen, appearance consistent with neurofibroma

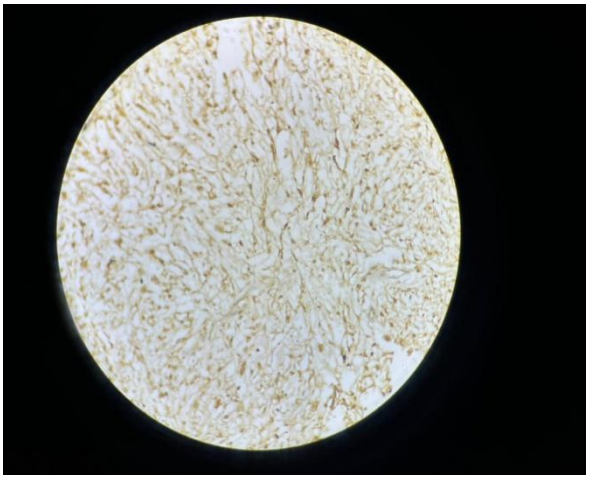


Figure 7: IHC showing S 100-positive

3. Discussion

Neurofibromas are benign neoplasms which originate from peripheral nerve sheaths. They are usually associated with Von Recklinghausen's disease as part of a generalized neurofibromatosis. The incidence of neurofibromas reported in the head and neck is approximately 25%, occurrence in oral cavity is 6.5%[3]. Neurofibromas of oropharynx are extremely rare.

There are only 5 reported cases of solitary neurofibroma of soft palate in English literature. This is the 6th case of solitary neurofibroma not associated with Von Recklinghausen's disease.

Sinha et al reported the first case of isolated neurofibroma of soft palate not associated with Von Recklinghausen's disease in 2002. The mass was 5x4x4 cm and a tracheostomy was done for this case as attempted oral and nasal intubation failed[4].

The second case was reported by Mazzoleni et al in 2009 in which the entire ulcerated mass was accompanied by the extraction of the upper left-hand second molar, which was attached to the mass through its palatal root[5].

The third case was reported by Choi et al in 2011[6]. The fourth case was by Smitha et al in 2013, which was diagnosed as neurofibroma with secondary changes[7]. The fifth case was reported in 2016 by Ramdurg et al [8].

Neurofibroma occurs in patients of any age and shows no sex predilection. Based on their site of occurrence and pattern of growth neurofibromas have been categorized into five distinct forms: localized and diffuse cutaneous neurofibromas, localized and plexiform intraneural neurofibromas, and a massive soft tissue variant. On gross examination they are seen as fusiform tumor originating from a nerve. Plexiform neurofibromas are elongated with beaded or bag of worm's appearance. Microscopically, neurofibromas are composed of spindle cells having an irregular, wavy nucleus, with varying cellularity. The nerve sheath origin of these tumours can be confirmed by the expression of S-100 protein, Ncam (CD56), and Leu 7 (CD57). Marked atypia and hypercellularity are indicators of malignant transformation[9]. Surgical excision is the treatment.

4. Conclusion

The successful identification and surgical excision of the isolated neurofibroma in the soft palate of the patient underscores the importance of considering rare diagnoses. The patient's post-surgery stability and absence of systemic involvement further highlight the potential for effective management of such rare cases. This case study contributes to the limited literature on isolated neurofibromas of the soft palate and emphasizes the need for continued research and documentation of such rare occurrences.

References

[1] Lee YB, Lee JI, Park HJ, Cho BK. Solitary Neurofibromas: Does an Uncommon Site Exist? Ann Dermatol.2012; 24 (1): 101.
[2] Pearl RA, O'Toole G. A case of large sporadic neurofibroma of the hand. Journal of Plastic, Reconstructive & Aesthetic Surgery.2010 Jul; 63 (7): e573–5.
[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. Clin Oral Invest.2007 Jun; 11 (2): 165–9.
[4] Sinha R, Paul R, Sen I, Sikdar B. A solitary huge neurofibroma of the soft palate. J Laryngol Otol.2002 Aug; 116 (8): 637–8.
[5] Solitary neurofibroma of the palate. A case report-Minerva Stomatologica 2009 September; 58 (9): 453-8 [Internet]. [cited 2023 Jul 9]. Available from: https: //www.minervamedica. it/en/journals/minerva-dental-and-oral science/article. php?cod=R18Y2009N09A0453
[6] Choi JS, Jang TJ. Solitary Myxoid Neurofibroma of the Soft Palate: Journal of Craniofacial Surgery.2011 Sep; 22 (5): 1901–3.
[7] Smitha S, Gangaraj SS, Shantharam L, Vijay S. Neurofibroma of Soft Palate: A Rare Case Report. An

International Journal of Otorhinolaryngology Clinics.2013; 5 (2): 19–20.
[8] Ramdurg P, Puranik SR, Dantu R, Shivanand R. Solitary Neurofibroma of the Soft Palate: A Rare Entity. Indian J Otolaryngol Head Neck Surg.2019 Oct; 71 (S1): 38–41.
[9] Neurofibroma-an overview | ScienceDirect Topics [Internet]. [cited 2023 Jul 9]. Available from: https: //www.sciencedirect. com/topics/veterinary-science-and-veterinary-medicine/neurofibroma