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Solitary Neurofibroma of the Hard Palate. A Case Report

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I. Introduction

Neurofibroma is the most common type of benign, slow-growing, peripheral nerve neoplasm, which arises from a mixture of cell types such as Schwann cells and perineural fibroblasts.[1] It can arise as a solitary tumor or be a component of Neurofibromatosis and von Recklinghausen's disease. Solitary neurofibromas are not frequently seen in the oral cavity, and those in the palate are reported in less numbers.[2] 4–7% of patients affected by neurofibromatosis display oral manifestations [3]. The frequency of isolated neurofibromas unassociated with neurofibromatosis in the oral cavity is uncertain [4].

There have only been 7 published cases of solitary neurofibroma of the hard palate. The mean patient age in these cases was 40±15 years, and there were 5 women (71%), suggesting a higher incidence among women [5].

This report describes a rare case of solitary neurofibroma in the hard palate.

II. Case Report

A 50-year-old female referred to the Department of Oral and Cranio-Maxillofacial Surgery, Hamad Medical Corporation, in 2017 with a chief complaint of swelling in the palate, with no associated pain but mostly discomfort while eating. The patient first noticed the swelling two years ago, and since then it had grown slowly to attain the present size which is around 2cm in diameter. patient was known to have rheumatoid arthritis, anemia and had hysterectomy for endometrial polyps in 2016. There was no family history of neurofibromatosis involved.

Oral examination showed pedunculated nodular round mass of 2 cm diameter overlying the mid portion of the hard palate, with normal mucosal color and no ulcerations, presence of overlying capillaries seen Fig 1. On palpation, consistency was firm, non-tender, non-mobile.

Initial panoramic view did not show any significant bony changes, CT was done and showed a well-defined rounded soft tissue lesion at the submucosal central hard palate location, measuring about 14.5 x 16 x 13 mm in maximum AP, transverse and cranio-caudal dimensions. The lesion showed minimal enhancement with well-defined surrounding fatty plane, with mild underlying bony remodeling without erosion. Fig. 2

Our initial differential diagnosis was pleomorphic adenoma, nerve sheath tumor, fibroma.

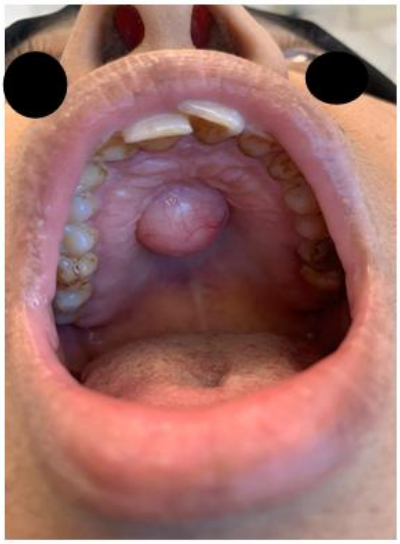

Fig 1. Clinical photo of the palatal tumor on presentation.

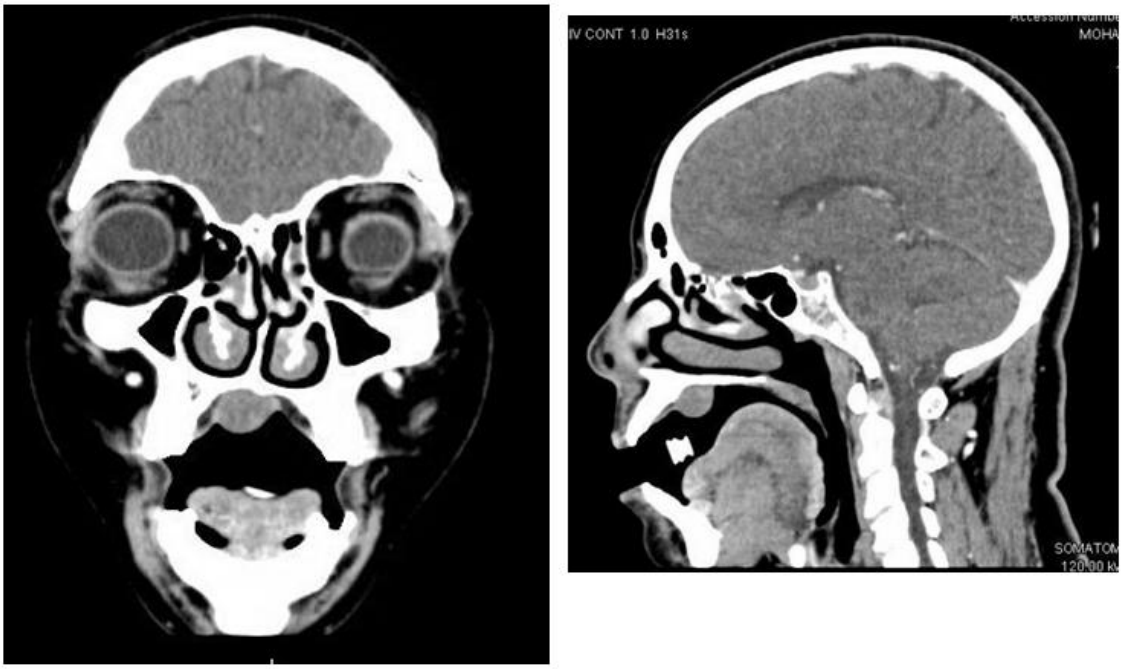

Fig 2. CT scan, Coronal and sagittal view.

Treatment

Fine needle aspiration biopsy was performed, spread on sheets and sent for cytology evaluation. The result came inconclusive.

The patient was planned for excision of the tumor under general anesthesia and consented for surgery. An impression was taken and splint was made pre-surgery. Then, under general anesthesia, with nasal endotracheal intubation, the tumor was excised completely using a monopolar diathermy with a margin of around 2-3mm all around, down to the underlying hard palatal bone. The mass was pedunculated and a groove of 2mm around it could be probed, Hemostasis achieved, and splint placed.

The post-operative period was uneventful, and the patient was discharged on day 2 post-op on soft diet, antibiotics, analgesics and antiseptic mouthwash.

On following up the patient, the wound was granulating well after 10 days, so the splint was trimmed from the palatal aspect and worn by the patient for another 5 days Fig. 3. By next appointment after 5 days, the splint was removed, she had no complaint while eating. The bed of the wound looked healthy and granulating well.

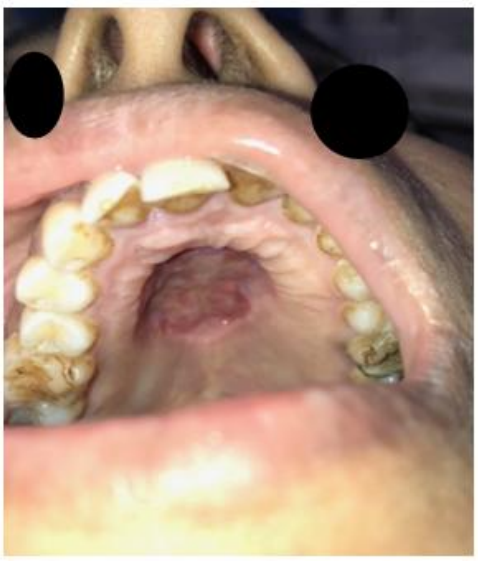

Fig 3. Clinical photo, 10 days post tumor excision

Histopathology

Sections show a well-circumscribed unencapsulated spindle cell tumor characterized by short fascicles of spindle cells with bland nuclear features and tapered ends. Mitotic rate is 1/10 HPF. Occasional atypical, degenerated nuclei are seen. By IHC the tumor cells are strongly positive with S100, CD34, SOX-10 and EMA while negative with CK 7, SMA. Ki-67 is 1%.

All controls Show appropriate reactivity. So Diagnosis of neurofibroma was made.

III. Discussion

Neurofibromatosis (NF) was first described in 1882 by the German anatomo-pathologist von Recklinghausen. Two clinically and genetically distinct subtypes were identified and have been designated as NF-1 and NF-2. The most common form is NF-1 which constitutes 80–95% of the cases. Neurofibroma is defined as a well-demarcated intraneural or diffusely infiltrative extra-neural tumor consisting of a mixture of cell types including Schwann cells, perineurial-like cells, and fibroblasts [6].

National Institute of Health consensus Development Conference in 1988 proposed a diagnostic criterion for neurofibromatosis type 1, if a patient has two or more of the following findings:

(1) Six or more cafe au lait macules more than 5 mm in greatest dimeter in prepubertal persons and more than 15 mm in greatest diameter in post pubertal persons;
(2) Two or more neurofibromas of any type or one plexiform neurofibroma;
(3) Freckling in the axillary or inguinal regions;
(4) Optic glioma;
(5) Two or more Lisch nodules (iris hamartomas);
(6) Distinctive osseous lesion such as sphenoid dysplasia;
(7) Family history of first-degree relative with neurofibromatosis.

Among the neural lesions affecting the head and neck, Neurofibromas (NFs) are relatively common, with the reported incidence in the head and neck being 37%.[7] The head and neck is commonly involved because of rich innervations of this area. NFs of the oral cavity and oropharynx are extremely rare, ranging from 2% to 7%,[8] and are mostly associated with NF-I.

In the oral cavity, the tumor often involves the trigeminal and upper cervical nerves, with the most common site in the oral cavity being the tongue followed by buccal mucosa, the floor of mouth, lips, and gingiva.[7] Solitary NF is a rare variant of NF; it seldom affects the head and neck. They are distinct as unlike other NFs that they lack the systemic and hereditary components.

Distinguishing between isolated neurofibroma and those associated with NF-1 is important because of difference in clinical behavior, treatment, and prognosis. The current treatment of neurofibroma is complete resection. The recurrence is seen in as many as 20% of the patients with a neurofibroma after complete resection and increases to 44% with subtotal resection [9]. Recurrence can be avoided by providing a safety margin, including the periosteum, around the tumor at the point of resection [5].

In this report, the patient did not have any of the criteria mentioned above to suspect neurofibromatosis, she was seen by an ophthalmologist and cleared, full body x-rays were done to rule out any other bony lesions and found to be clear. In addition, she has no family history for the disease, and clinically did not show any freckling's or discoloration of the skin.

The excision in this case was complete with safe margins, and the patient was followed for 2 years with good healing and no recurrence.

Solitary neurofibromas of the hard palate, as in the present study, are extremely rare, and only 7 cases have been reported to date. There was no recurrence in any of the 7 previous reports.

In conclusion, solitary neurofibroma of the oral cavity is a rare benign pathology that should be kept in mind, more commonly associated with neurofibromatosis disease. It is important to look for other symptoms related to NFS, to avoid any delay of treatment of the disease.

Conflicts of interest

There are no conflicts of interest.

References

[1]. Neville BW, Damm DD, Allen CM, Jerry E. Bouquot. Oral and Maxillofacial Pathology. 3rd Edition. Philadelphia; WB Saunders Company 2013. P. 528.
[2]. Maruyama M, Fushiki H & Watanabe, Y. Solitary neurofibroma of the floor of the mouth: a case report. Case reports in otolaryngology 2011;2011:1-3.
[3]. A. Narwal, S. Saxena, V. Rathod, and P. Bansal, Intraoral solitary neurofibroma in an infant, Journal of Oral and Maxillo Facial Pathology, 2008;12:75-78.
[4]. L. S. Marcocchio, D. T. Oliveira, M. C. Periera, C. T. Soares, and R. N. Fleury, Sporadic and multiple neurofibromas in the head and neck region: a retrospective study of 33 years, Clinical oral investigations, 2007;11:165-169.

[5]. Taketomi T, Nakamura K, Teratani Y, Matsuo K, Kusukawa J. Solitary Neurofibroma of the Hard Palate: A Case Report and Literature Review. Am J Case Rep. 2021; 22;22:e929674.
[6]. J. M. Woodruff, H. P. Kourea, D. N. Luis, and B. W. Scheithauer, "Neurofibroma," in WHO classification: Tumours of the Nervous System, pp. 167-168, IARC Press, Lyon, France,1st edition, 1999.
[7]. Thimmaiah AH, Santharam L, Gangaraj SS, Vijay S. Neurofibroma of soft palate: A rare case report. Otorhinolaryngol Vlin 2013;5:109-10.
[8]. Behbehani EM, Al-Ramzi AH, Mohamed EA, Oral manifestations of neurofibromatosis: Case report. Dent News 1997;4:17-21.
[9]. M. N. Needle, A. Cnaan, J. Dattilo et al., Prognostic sings in the surgical management of plexiform neurofibroma: the children's hospital of Philadelphia experience, Journal of Pediatrics, 1997;131:678-682.

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