CASE REPORT

Neurovascular Hamartoma of Palate—A Rare Case Report

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Abstract

The oral hamartoma is a unique presentation of head and neck tumors with nonspecific etiology and atypical histopathological presentation. Oral hamartomas generally present as smooth, painless, pinkish masses, histologically comprising neural and vascular components intermingled in connective tissue stroma. The presence of a neurovascular hamartoma (NVH) within the oral cavity is truly a rare entity, scarcely reported in the literature. These tumors are difficult to diagnose and are most commonly benign with good prognosis and clinical outcome. Here, we present a case of oral NVH of the palate in a pediatric patient.

Keywords: Case report, Hamartoma, Lesion, Palate.

International Journal of Clinical Pediatric Dentistry (2024): 10.5005/jp-journals-10005-2888

Introduction

The term hamartoma was first used by Albrecht (1904), describing a benign tumor-like developmental malformation composed of proliferative tissue elements endogenous to its anatomic location of occurrence.[1] Hamartomas are commonly seen externally in many parts of the body, such as the skin, and also in visceral organs such as the lung, pancreas, spleen, liver, and kidney. Their occurrence is quite rare in the orofacial region.[2] Within the oral cavity, the most commonly implicated sites are the tongue, buccal mucosa, and lower lip.[3] They may arise from both odontogenic and nonodontogenic embryonic tissues.[3] An oral neurovascular hamartoma (NVH) is very rare, with the proliferation of neural tissue solely or in combination with vascular elements as the histological presentation.[1] We present a case report of NVH of the palate, which presented as a painless swelling associated with minor erosion of the bone.

Case Description

A 10-year-old male patient came to the Department of Pedodontics and Preventive Dentistry, complaining of a painless swelling on the left side of the palate. Medical history was insignificant. The swelling started as a small nodule 6 months back, which was asymptomatic and slow-growing. Swelling is approximately 1 × 2 cm, extending from the left maxillary canine to the mesial side of the left maxillary first molar (Fig. 1). The lesion extended medially to the midpalate. The swelling was soft at the center and firm at the periphery on palpation. A provisional diagnosis of odontogenic cyst or benign soft tissue tumor was put forward, and fine needle aspiration was negative for any content.

Three-dimensional cone-beam computed tomography (3D CBCT) showed mild erosion of the palatal cortical plate, with no major abnormality detected. Both axial and coronal sections did not reveal any perforation of the bone segment nor any radiolucency inside the bone, ruling out the possibility of lesion to be central in origin (Fig. 2). Hence, the case was posted for surgical excision and evaluation. After necessary investigations, the surgical procedure for excision was performed. Under local anesthesia, full-thickness mucoperiosteal flap was elevated with a relieving incision in the anterior palate (Fig. 3). Intraoperatively, there was

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How to cite this article: Sandeep V, Vidhyadhari P, Rajesh N, *et al.* Neurovascular Hamartoma of Palate—A Rare Case Report. Int J Clin Pediatr Dent 2024;17(7):826–828.

Source of support: Nil

Conflict of interest: None

Patient consent statement: The author(s) have obtained written informed consent from the patient's parents/legal guardians for publication of the case report details and related images.

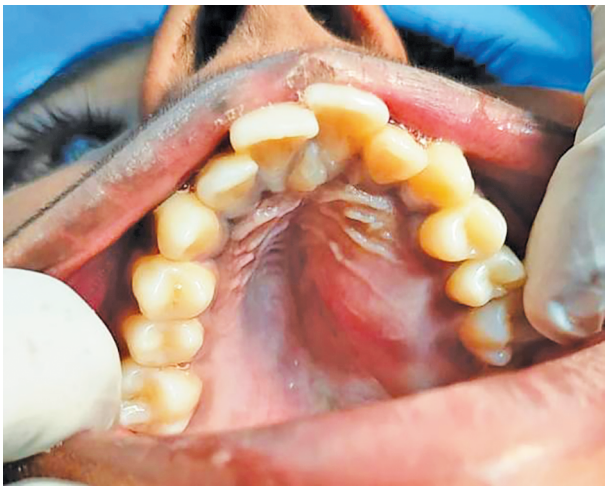

Fig. 1: Preoperative picture showing palatal swelling

no specific delineation or encapsulation of the lesion (Fig. 4); hence, with adequate margin, the entire lesion was curetted out (Fig. 5), followed by primary closure with 3-0 silk. A palatal obturator (Fig. 6) was given to prevent untoward bleeding, and prophylactic antibiotics and analgesics were prescribed. Healing was uneventful after 7 days, and sutures were removed.

Histopathological examination showed keratinized stratified squamous epithelium with underlying fibrovascular connective tissue stroma. Under higher magnification (4×), the epithelium was parakeratinized with six to seven layers of thickness. The connective tissue showed numerous proliferating blood capillaries (Fig. 7). Few large capillaries with extravasated red blood cells were seen in the loosely arranged collagen fibers. The transverse and longitudinal sections of nerve bundles were seen surrounded by small blood capillaries. Focal areas of adipose tissue were evident at the periphery of the section, along with mucous acini (Fig. 8).

Discussion

The epithelial and mesenchymal hamartomas involving the oral cavity are quite rare, and the neurovascular component in a hamartoma is even more infrequent.[4] Few cases of NVH have been reported in the literature, which are associated with syndromes and cleft lip and palate.[5] The present case is a nonsyndromic NVH in a 10-year-old patient. Palatal NVH has histologically many components derived from the embryonic derivatives, such as smooth muscle and adipose tissue, but not numerous proliferating blood capillaries along with neural tissues, as in our case.[6] NVH of the palate poses radiological challenges in finding out the extension of the lesion.[7] So, more advanced aids like CBCT can give clear extension of the lesion to determine its course, whether malignant or benign. CBCT, in this particular case, does show little bony erosion. Few cases were reported in the literature with palatal bone erosion similar to our case.[6]

Allon et al. first described the microscopic characteristics of NVH, which usually present as nonencapsulated lesions, poorly circumscribed masses comprising nerve bundles closely intertwined with small- to medium-sized blood capillaries within a loose connective tissue matrix.[8] The case we present here showed longitudinal and transverse sections of nerve bundles that are dispersed in highly vascular loose connective tissue stroma with numerous blood capillaries. The microscopic features coincide with those in literature as described by Allen et al., by which diagnosis was concluded as NVH.

The clinical course of most of the oral hamartomas is benign, and there is no reported reoccurrence after surgical excision.

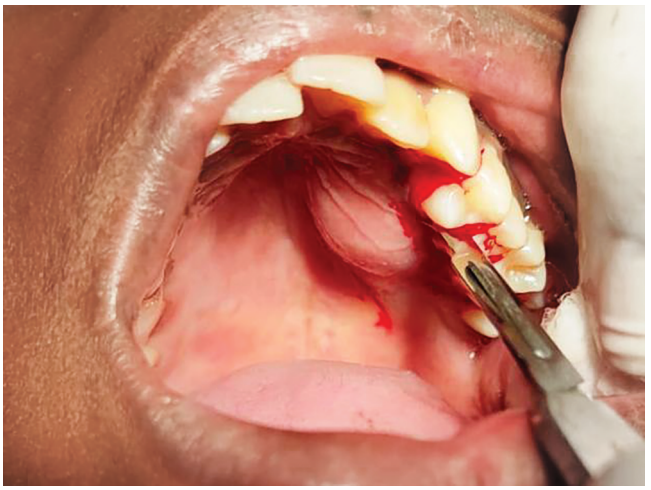

Fig. 3: Full-thickness mucoperiosteal flap raised with sulcular incision

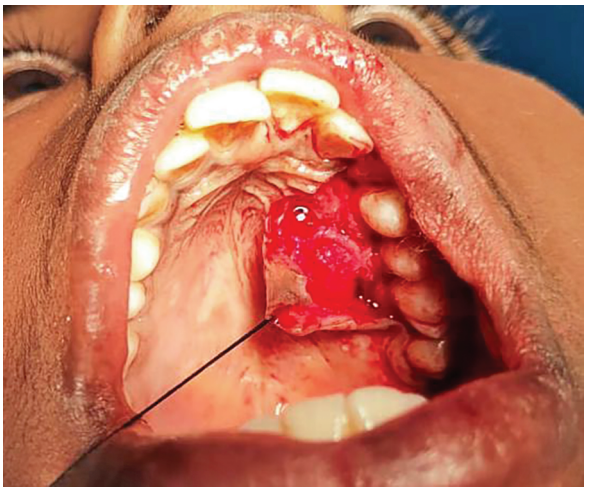

Fig. 4: Lesion showing nonencapsulated mass

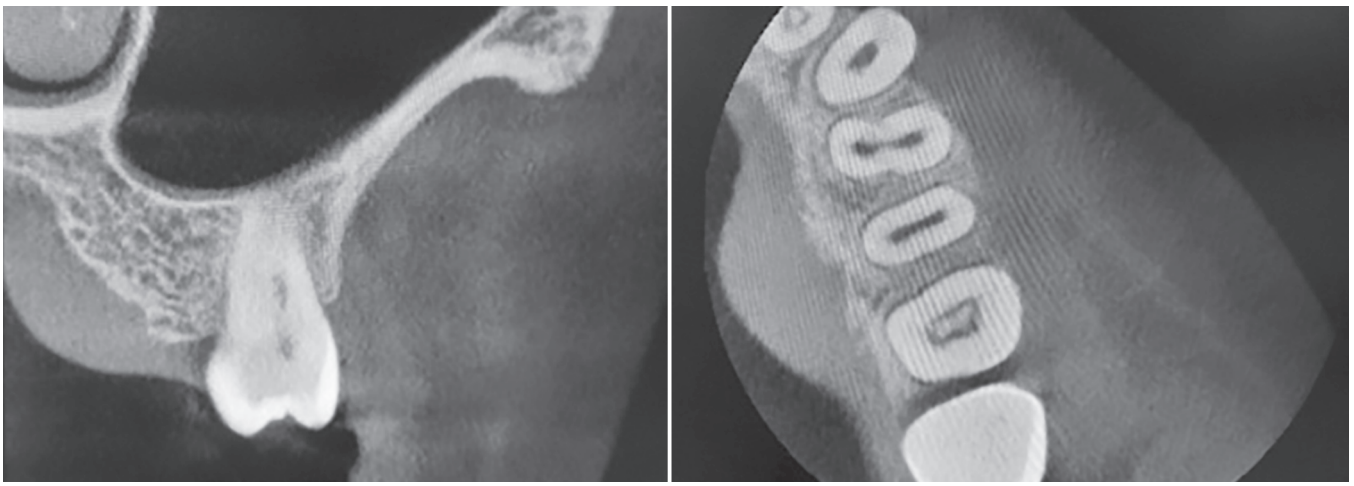

Fig. 2: Cone-beam computed tomography showing no bony involvement with minor erosion

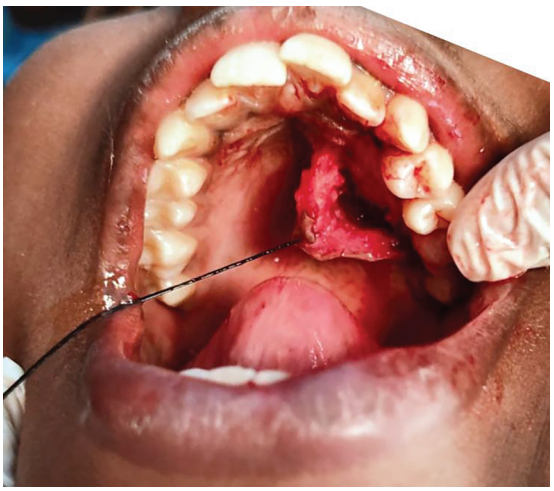

Fig. 5: Postoperative picture showing excised lesion

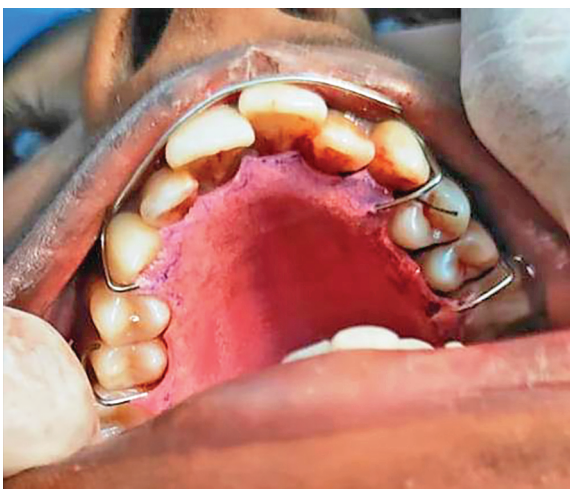

Fig. 6: Postoperative palatal obturator

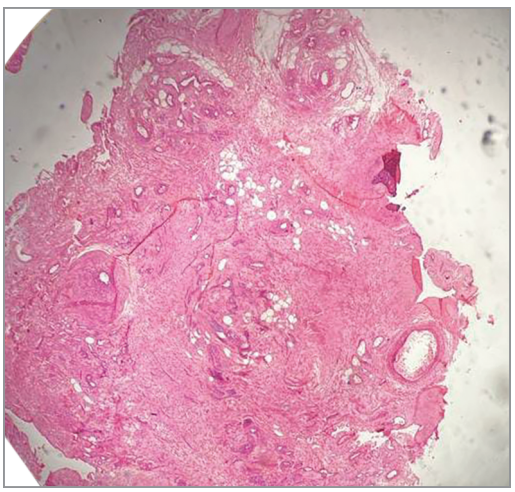

Fig. 7: 4× hematoxylin and eosin (H&E) section showing parakeratinized epithelium with underlying fibrovascular connective tissue

A recent review of head and neck hamartomas has shown that these benign lesions may expand and recur, and chronic inflammation may increase the risk of malignant transformation.[9] Harmartoma

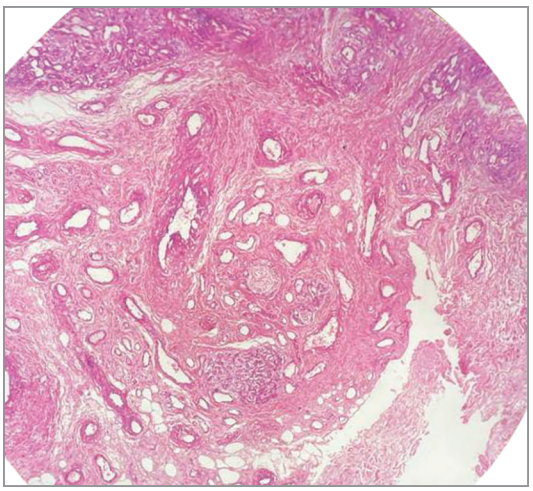

Fig. 8: 10× H&E section showing fibrovascular connective tissue with transverse section of nerve bundle and numerous proliferating blood capillaries

can be associated with skin lesions as well, with a possible association with more aggressive and often fatal rhabdoid tumor, as reported by Lee et al.[10] Hence, we suggest surgical excision with wide margins as the treatment of choice, followed by patient review at regular intervals to check for any recurrence of the lesion. In the present case, an obturator was given to the patient to compress the palatal mucosal swelling and aid in faster healing. Recall follow-up of the patient showed no recurrence after 6 months.

Why this paper is important to pediatric dentists:

- This paper highlights the importance of considering hamartoma as a differential diagnosis in palatal painless swellings.
- Clinical presentation of histopathologically confirmed NVH of the palate is mostly benign and has good prognosis.
- Confirmed NVH should be carefully followed if the patient has associated dermal rhabdoid tumors.

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