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Dentistry

NEUROVASCULAR HAMARTOMA: A RARE INTRAORAL ENTITY

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ABSTRACT Neurovascular Hamartoma is a very rare pathological lesion found in oral cavity. Due to rare intraoral presence of lesion very few literatures are available to identify, diagnose, treatment plan and histopathological impression. Here we report about a 44-year-old female who came with the complaint of painless palatal swelling. patient has history of Neurofibromatosis and was provisionally diagnosed with Neurofibroma. Histopathological examination confirmed the diagnosis with Neurovascular Hamartoma. Neurovascular hamartoma of oral cavity being a benign lesion creates diagnostic dilemma for the surgeon mimicking malignancy.

KEYWORDS : Neurofibroma; Neurofibromatosis; Neurovascular Hamartoma; Painless Swelling; Palate.

INTRODUCTION

The term hamartoma is derived from the Greek word 'Hamartia' which was first introduced by German pathologist Eugen Albrecht in 1904, which literally means "character flaw" [1]. It refers to a benign malformation in different areas of the body where growth occurs. The histology consists of an abnormal mixture of cells and tissues localised at the particular site secondary to a developmental error which are endogenous to the site [2,3]. They do not develop as part of an inflammatory or neoplastic process and have a self-limiting proliferation [2]. According to the predominance of the tissue, hamartomas can be classified as vascular angiomatous, lipomatous, chondroid, osseous or neurogenic [3]. An oral neurovascular hamartoma is rare because of proliferation of neural tissue solely or in combination with vascular elements [2]. Blood vessels, nerves, lymphatic vessels, skeletal muscles, adipose tissues, salivary gland components and epithelium are the endogenous components which may cause hamartoma with in the oral cavity [4,5].

Case Report

A 44-year-old female patient reported with the complaint of painless swelling in upper right jaw. Patient first noted this swelling 14 years back which was size of a pea. Swelling gradually increased and covered whole side of the right hard palate. Patient is having a habit of chew mawa (tobacco) for 5 years. Patient was also observed with Neurofibromatosis over skin which was first noticed 15 years back.

General examination revealed diffuse multiple brownish black pigmented nodular masses of various sizes spreading all over the body. On palpation, these noninflammatory nodules varied in consistency, from soft to firm.

On intraoral examination swelling was present on right side palate extending from teeth with relation to 11 to 18 which was covering complete right side hard palate and some part of soft palate not crossing midline. On palpation swelling was soft, sessile, non-tender, non-purulent. Patient had poor oral hygiene with chronic generalized gingivitis and periodontitis. Generalized mobility of teeth was present. Patient was provisionally diagnosed with the neurofibroma of hard palate.

For the further investigation patient was referred to department of dermatology and ophthalmology where patient was provisionally diagnosed with neurofibromatosis type-I over the skin and lisch nodule in bilateral pupil.

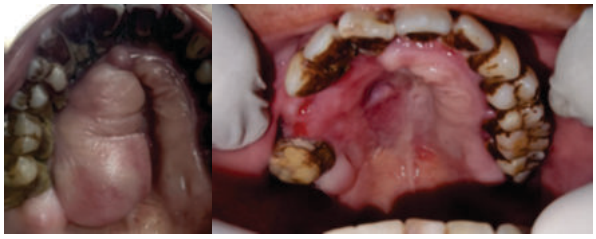
Investigation

The multi slice CT-Scan of maxilla shows a well-defined soft tissue density lesion of size 40(AP)x 14(CC)x 38(TR) mm is noted in hard palate on right side. The lesion shows minimal enhancement on post contrast study. The lesion is extending in upper alveolus on right side with minimal erosion of floor of right maxillary sinus was noted. The lesion is crossing midline and minimally extending to left side. Lesion is causing thinning with scalloping of adjacent upper alveolar arch and hard palate.

Based on the clinical and radiographic presentation patient was provisionally diagnosed with Neurofibroma.

Treatment

After routine blood examination patient was planned for excision of lesion. The complete soft tissue growth was excised from right side hard palate under local anaesthesia along with extraction of tooth with relation to 14,15,16. Raw surface of palate was covered with paraffin gauge and fixed with prefabricated palatal splint using interdental wire. The patient was advised to take soft and semi liquid diet. After 15 days palatal splint was removed and wound was irrigated using normal saline and kept on follow up after 7 days. Specimen sent for histopathological examination.



(A) (B)

Figure 1(A) Oral Neurovascular Hamartoma on Right side Palate

Figure 1 (B) Post operative healing after four weeks

Histopathology

On histopathological examination submucosal diffuse proliferation of neural cells, fibroblast with entrapped medium size blood vessels and adipocytes was noted. Also noted hypertrophied nerve fascicles and skeletal muscle tissue. There was no capsular or circumscription and no evidence of malignancy. The lesion was concluded to be Neurovascular Hamartoma of oral cavity.

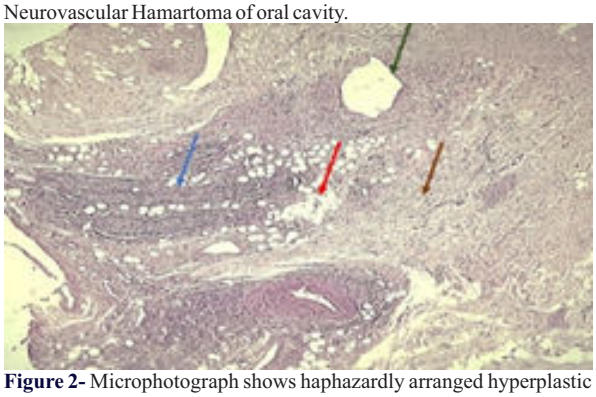


Figure 2- Microphotograph shows haphazardly arranged hyperplastic nerve fascicles (blue arrow), blood vessels (green arrow), adipocytes (red arrow) and a loose spindled stroma (brown arrow), H&E, 10x10X=100X.

Follow-up

Patient kept on follow-up on weekly basis. On patients first follow-up patient had no pain related to this and had some difficulties in having food. On second follow-up prefabricated splint was removed. Healing was satisfactory with no complain. Patient was advised to keep maintaining oral hygiene. Further weekly follow-up was also planned to observe healing. After 6 months there was no evidence of recurrence.

DISCUSSION

Hamartomas arising within the oral cavity shows a variety of clinical presentations, histological and growth patterns [2]. The biological behaviour is benign with reasons for development vague. The epithelial and mesenchymal hamartomas involving the oral cavity are rare and the presence of a neural or a neurovascular component in a hamartoma is even more infrequent[6]. Oral neurovascular hamartomas present as smooth surfaced exophytic lesions usually pedunculated or wide base and their colour may range from pink to yellow [2,7]. These lesions are usually painless and leading general discomfort to the patient. Head and neck hamartomas shows benign entities which may expand and may have little or no malignant transformation rate [1,8].

Histologically, lesions are non-encapsulated and poorly circumscribed from the adjacent tissues [5]. They are composed of hypocellular connective tissue containing aggregates of loose vessels closely packed with small to medium sized nerve bundles, they are not easily distinguishable from the surrounding non-hamartomatous tissues [2,5,8]. Oral neurovascular hamartomas resemble traumatic hamartomas and can be distinguished based on the fact that the blood vessels in the former are very closely entwined with the neural component.

CONCLUSION

Neurovascular Hamartoma is a rare intraoral entity. It should be considering as differential diagnosis of many oral lesions. Neurovascular hamartoma was initially diagnosed as neurofibroma, fibrous hyperplasia, pyogenic granuloma, peripheral giant cell granuloma, lipoma. Therefore, it is important to know the histopathological characteristics associated with the lesion.

Informed Consent

The patient consent has been obtained.

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Nil.

Conflicts Of Interest

There are no conflicts of interest.

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