NON INVOLUTING HEMANGIOMA WITH PERIVASCULAR FIBROSIS: AN UNCOMMON PRESENTATION

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ABSTRACT: Hemangiomas are benign vascular lesions characterized by rapid growth of endothelial cells. They first arise during first 8 weeks of life, then proliferate and after that undergo involution by puberty. About 85 to 95% of hemangiomas involute completely by the age of puberty. The remaining 5 to 15% involute incompletely and require the management. Intraorally, they commonly affect lips, buccal mucosa and tongue. Hemangiomas, though are benign, requires accurate diagnosis and precise management. This case report presents a case of cavernous hemangioma of left buccal mucosa in a 21-year-old male.

KEYWORDS: Hemangioma, Vascular Malformations.

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INTRODUCTION: A number of terms have been used to describe vascular lesions, which are classified either as hemangiomas or vascular malformations. The classification developed by Mulliken and Glowacki in 1982 is based on the cellular kinetics of anomalous vessels, providing a diagnostic and therapeutic approach based on the biologic behavior of the lesion. In this classification, two entities exist: 1) Hemangiomas and 2) Vascular malformations.[1,2]

Hemangiomas are considered to be benign tumors of infancy that are characterized by a rapid growth phase with endothelial cell proliferation. On the other hand, vascular malformations are structural anomalies of blood vessels without endothelial proliferation.[1,2]

Hemangioma is further sub-classified based on their histological appearance as: (1) Capillary Lesions; (2) Cavernous Lesions; and (3) Mixed Lesions.[1,2] A sclerosing variety also occurs that tends to undergo spontaneous fibrosis.

Although, hemangioma is considered one of the most common soft tissue tumors of the head and neck, it is relatively rare in the oral cavity and uncommonly encountered by the clinicians. We are presenting a case of cavernous hemangioma of buccal mucosa.

CASE REPORT: A 21 yr male visited with chief complaint of a reddish purple swelling with cheek mucosa near upper left back teeth region. Past medical history, past dental history and family history were not significant. No abnormality was detected during a general and extraoral examination. (Figure 1).

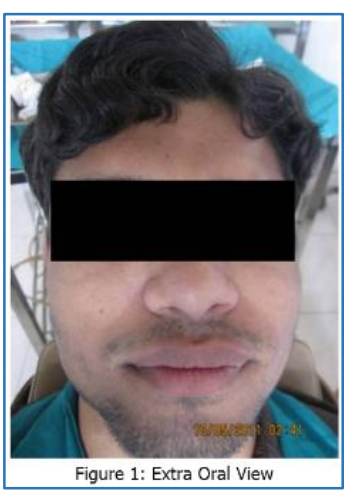

Figure 1: Extra Oral View

Intraoral examination revealed a single, oval swelling on buccal mucosa of left side in relation with 25, 26, and 27. On inspection, the size of swelling was 1.5cm x 1cm approx., brownish purple in color. The surface of the swelling was irregular, pebbly. Margins were well defined. On palpation consistency of the swelling was soft. It was compressible, nonfluctuant, nonreducible and nontender. (Figure 2) Surrounding mucosa was normal. The clinical differential diagnoses considered were hemangioma, varicosities, arterio-venous malformation. The lesion was surgically excised under local anesthesia (Figure 3) and sent for histopathological examination.

Gross examination of the specimen revealed a single soft tissue bit of 1.5cm X 1cm dimensions, oval in shape, soft in consistency, and grayish white in color is received. (Figure 4).

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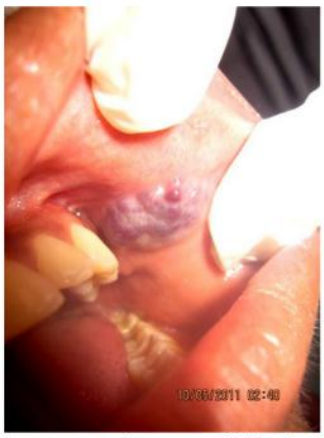

Figure 2: Intra Oral View: A single bluish-purple colored swelling with lobulated surface

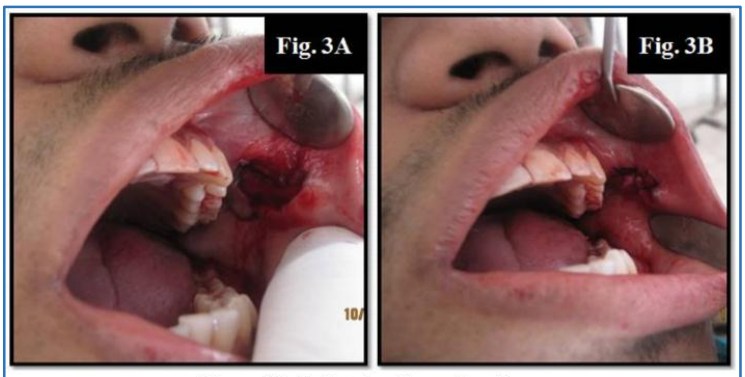

Figure 3A & B: Intra Operative View

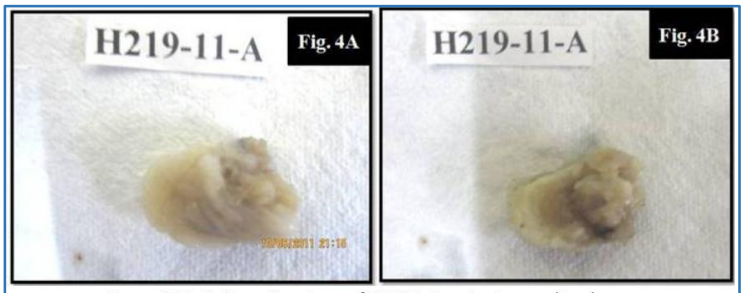

Figure 4A & B: Gross Specimen of 1.5 X 1.5 cm in size, oval in shape, grayish white in color, firm in consistency

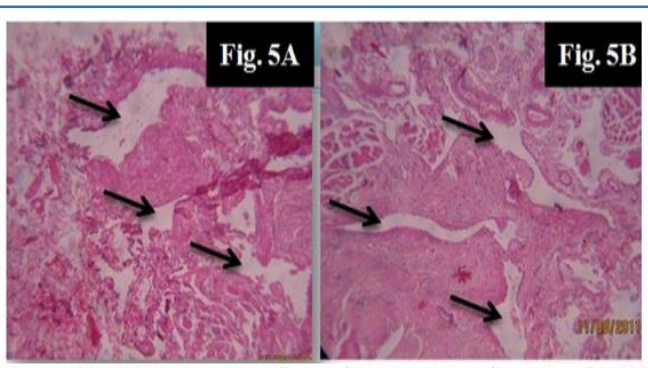

Figure 5A & 5B: Hematoxylin and Eosin stained section (100X) showing large dilated endothelial lined vascular spaces (Black arrow)

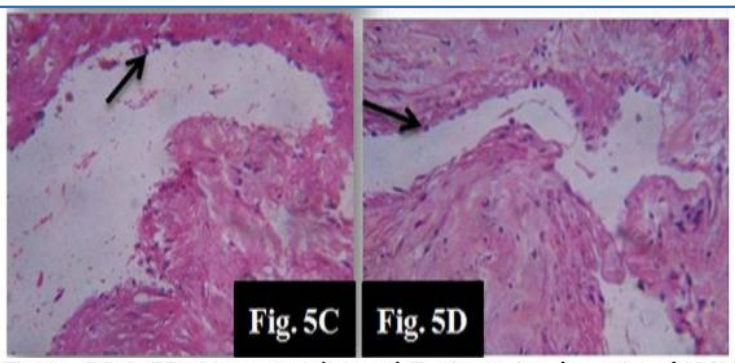

Figure 5C & 5D: Hematoxylin and Eosin stained section (400X) showing large dilated vascular spaces lined by plump endothelial cells (Black arrow)

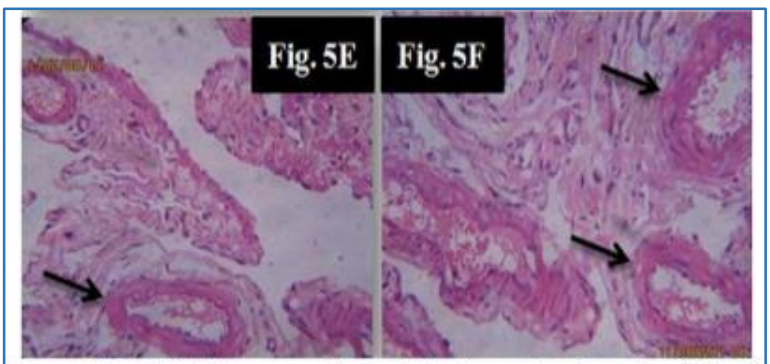

Figure 5E & 5F: Hematoxylin and Eosin stained section (400X) showing endothelial lined capillaries with perivascular fibrosis

Histopathological examination of the specimen with H and E staining showed lesional connective tissue with multiple irregularly shaped vascular spaces lined by the endothelial cells. (Figure 5A, 5B, 5C, 5D). It also shows the few endothelial lined blood vessels. The blood vessels show some perivasular condensation of the collagen fibers. (Figure 5E and 5F) It also shows some bundles of skeletal muscle fibers and some serous salivary gland acini. Overlying parakeratinized stratified squamous epithelium is also noted.

From all the above clinicopathological examination, final diagnosis of Cavernous Hemangioma with peri-vascular fibrosis was concluded.

Hemangiomas are the most common tumor of infancy, occurring in 5% to 10% of 1 year old children. They are much common in females than males (ratio 3:1).[3] In contrast to this, the present case was observed in a 21 year old male. Eighty percent of hemangiomas occur as single lesion, but 20% of affected patients will have multiple tumors.[3] The present case was also having a single tumor.

Approximately 85% of childhood onset hemangiomas spontaneously regress after puberty, whereas a varix arises in older individuals and once formed does not regress.[4] The present case was of an hemangioma which was not regressed even after puberty.

Congenital hemangioma, the lesion starts developing at 8th week of intrauterine life. They show three distinct developmental phases: Proliferation, quiescence and involution. During proliferation phase, hemangiomas rapidly proliferate within 12-18 months. This phase may also be accompanied by ischemia, necrosis, ulceration and bleeding. After proliferation phase, haemangiomas enter a slower or no growth phase, known as quiescence. The last and unique phase of haemangioma is known as involution. Graying of the overlying skin and shrinking of the deeper components are important clinical features of involution phase; 90% of infantile hemangiomas slowly involute by the age of 9 years.[5] But the hemangioma presented in this case has not involuted after puberty of the patient.

The pathogenesis of hemangiomas still remains unclear. Currently, there are two proposed theories. The first theory suggests that the endothelial cells of haemangiomas arise from disrupted placental tissue embedded in the foetal soft tissues during gestation or birth. Markers of haemangiomas have been shown to coincide with those found in placental tissue.[5] The second theory arose from the discovery of endothelial progenitor and stem cells in the circulation of patients with haemangioma. The development of haemangioma in animals from stem cells isolated from human specimens support this theory.[5]

Cytokines, such as basic fibroblast growth factor (bFGF) and vascular endothelial growth factor (VEGF) are known to stimulate angiogenesis. Excesses of these angiogenic factors or decreases of angiogenesis inhibitors (eg,

gamma-interferon, tumor necrosis factor–beta, transforming growth factor–beta) have been implicated in the development of hemangiomas.[6]

The clinical appearance of this tumor is similar to that of many other vascular tumors. The hemangioma appears as soft mass, smooth or lobulated, and sessile or pedunculated and may vary in size from a few millimeters to several centimeters. Cavernous hemangiomas in adults do not regress and have a chronic course with slow progressive growth. They are usually deep red and may blanch on the application of pressure and if large in size, might interfere with mastication.[6] Our case was clinically similar to those above described.

Congenital hemangiomas have been classified microscopically as capillary hemangiomas or cavernous hemangiomas, depending on whether the microscopic size of the capillaries is small or large, respectively. The vascular spaces are lined by endothelium without muscular support. Clinically, no significant difference is noted between capillary and cavernous hemangiomas. Hemangiomas are differentiated from vascular malformations based on the fact that malformations consist not only of capillaries but also of venous, arteriolar, and lymphatic channels. Direct arteriovenous communications are typical. Lesions may be of purely one type of vessel, or they may be combinations of two or more.[2]

Mucocele, ranula, superficial cysts, varicosities and arteriovenous shunts are considered as differential of hemangiomas. Mucocele, ranula and superficial cysts show mostly bluish color, and found most commonly on lower lip or the floor of the mouth. On palpation they are soft, fluctuant, freely mobile and cannot be emptied by digital pressure. Varicosities are of dilation of superficial vein, appear as elongated enlargements and are typically observed on the ventral surface of the tongue. Arteriovenous shunts are rubbery, nonfluctuant, throbbing and pulsatile.[7]

Syndromes associated with cavernous hemangiomas are Rendu-Osler-Weber syndrome, Sturge-Weber-Dimitri syndrome, Kasabach-Merritt syndrome, Maffucci syndrome, von Hippel-Lindau syndrome, Klippel-Trenaunay-Weber syndrome, PHACE (posterior fossa brain malformations, haemangioma of the face, arterial cerebrovascular anomalies, cardiovascular anomalies, eye anomalies, and sterna defects or supraumbilical raphe) syndrome.[8]

Hemangiomas are highly vascular soft tissue lesions, so biopsy and fine needle aspiration cytology is contraindicated. Magnetic Resonance Imaging is used for volumetric analysis by T1 and T2 weighted images. Gradient Recalled Echo images are used to decide whether the anomaly is a high-flow lesion or slow-flow lesion.[9] Color Doppler ultrasound is also used to identify the feeding vessel, which is then ligated during the surgical procedure.[9]

Most of the true hemangiomas undergo spontaneous regression at an early age. Only 10-20% requires treatment.[1] Different therapeutic procedures available are microembolization, radiation, cryotherapy, sclerosing agents, corticosteroids and laser therapy. Of them, complete surgical excision of these lesions, if possible provides the best chance of cure. In the present case, the treatment comprised of complete surgical excision of the lesion with removal of entire base of lesion. The prognosis of hemangioma, in general, is excellent since it does not tend to recur or undergo malignant transformation following adequate treatment. In the case presented here, the patient was recalled at regular intervals and no sign of recurrence was reported till one year follow-up.

CONCLUSION: Intraorally, hemangiomas most commonly affect buccal mucosa. Although most of hemangiomas involute, few of isolated hemangiomas do not involute and require treatment. These types of lesions should be properly differentiated from other bluish red lesions. Proper care should be taken during management of such lesions as there are chances of heavy bleeding during treatment which could be fatal.

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