NON INVOLUTING CAVERNOUS HEMANGIOMA OF BUCCAL MUCOSA: A REPORT OF TWO CASES

Amit Mhapuskar[1], Prachi Shende[2,*]

[1]Prof. & Head, [2]P.G. Student, Dept. of Oral Medicine & Radiology,
Sinhgad Dental College & Hospital, Pune, India.

***Corresponding Author:**
E-mail: shende.prachi@gmail.com

ABSTRACT

Hemangioma is a benign vascular abnormality characterized by rapid growth of endothelial cells. It arises during first 8 weeks of life, rapidly proliferates during 6-12 months of life and gradually involutes by 9-12 years. Hemangiomas occur more in females with male to female ratio being 1:3 to 1:5. Almost 80 to 90% of hemangiomas involute completely by the age of 9 years. The remaining 10-20 % involute incompletely and require post adolescent ablative treatment or other modes of management. 50-60% of hemangiomas occur in head and neck region. Lips, buccal mucosa and tongue are the most common sites of involvement intra orally. Here, we report 2 cases of non involuting type of hemangioma located on buccal mucosa which were diagnosed histopathologically as cavernous hemangioma. Radiotherapy, cryotherapy, laser therapy, injection of sclerosing agent, selective embolisation are the various treatment modalities currently available for managing hemangiomas. Considering the location, size, accessibility and nature of lesion, surgical excision is the treatment of choice for smaller lesions.

Key Words: *Cavernous haemangioma, Intraoral, Phleboliths, Surgical excision.*

INTRODUCTION

Hemangioma is a benign vascular abnormality characterized by rapid endothelial cell proliferation. It is not present at birth but can be seen within the first month of life.[1] Hemangioma exist in three phases viz rapid proliferative phase, involuting phase, nonexistent phase.[2] 65% of haemangiomas are located in the head and neck region. Intra orally lips, tongue, buccal mucosa and palate are common sites. Presence of phleboliths was also reported in some cases.[3] Hemangiomas are known to bleed excessively on manipulation. Hence, it is important for the clinician to diagnose such lesions. This paper report two cases of buccal cavernous hemangiomas, diagnosed on the basis of clinical, radiographic and histopathological findings and were treated surgically.

CASE REPORT

Case1: A 50 year old female patient reported to the department of Oral Medicine with a chief complaint of swelling intra orally on left buccal mucosa. According to the patient, the swelling has been present since childhood with very minimal increase in size over the years. Past medical, dental, family histories were non contributory. On physical examination she appeared to be healthy with all her vital signs under normal limits. Intra oral examination revealed a solitary bluish purple lesion on the left buccal mucosa. The size of the lesion was about 2x1cm in diameter. Surface of the lesion appeared irregular and was well-delineated from surrounding mucosa which appeared normal. (Fig 1) On palpation the swelling blanched under pressure. Depending on the clinical features a provisional diagnosis of hemangioma was made. A modified soft tissue buccal mucosa radiograph with reduced exposure was made with an intra oral periapical film. It revealed the presence of radiopacities having a concentric arrangement indicative of phleboliths. (Fig 2)

Considering the small size (2x1cm) and the site (buccal mucosa) of the lesion, surgical excision was done with control of haemorrhage. Histopathological examination revealed presence of stratified squamous epithelium. The basement membrane was intact all over. Scanty fibro cellular stroma with numerous blood filled capillaries was seen throughout the connective tissue. Some areas showed extravasated blood elements and few foci of chronic inflammatory cells in underlying connective tissue. The overall picture was suggestive of cavernous hemangioma.

Case 2: A 25 year old female patient reported with a chief complaint of intra oral swelling on the right buccal mucosa. Patient became aware of the swelling 10 years back and since then the swelling has gradually increased to the present size. On intra oral examination, a single, well defined purplish blue, nodular lesion was seen on the right buccal mucosa, measuring approximately 2 x 2 cm in diameter. The overlying surface was irregular with normal surrounding area. (Fig 3) On palpation, the lesion was soft and compressible, non tender, non pulsatile, non mobile and non adherent to underlying structures.

The lesion blanched on pressure. Depending on the clinical features a provisional diagnosis of hemangioma was made. Routine blood investigations were normal. On surgical excision, histopathlogical

examination revealed cavernous hemangioma with stratified squamous keratinized epithelium. The connective tissue stroma showed large dilated, endothelial lined vascular spaces engorged with red blood cells. Deeper connective tissue showed adipose tissue, muscle and mild chronic inflammatory cell infiltrate. (Fig 4)

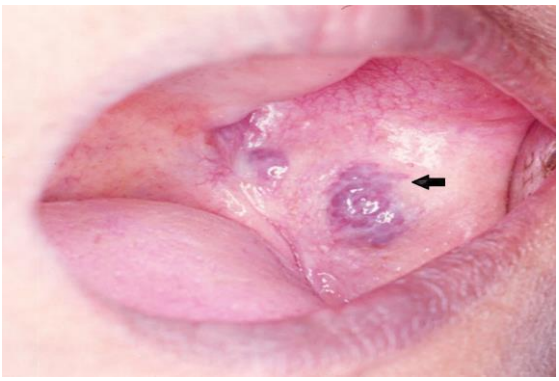

Fig. 1: Intra oral view showing bluish purple lesion on left buccal mucosa (black arrow)

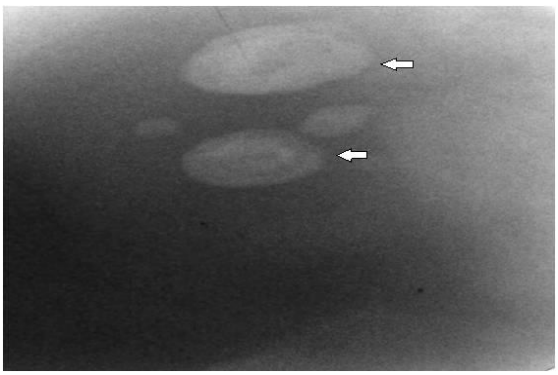

Fig. 2: A modified soft tissue intra oral buccal mucosa radiograph showing concentric arrangement of radiopacities (white arrow)

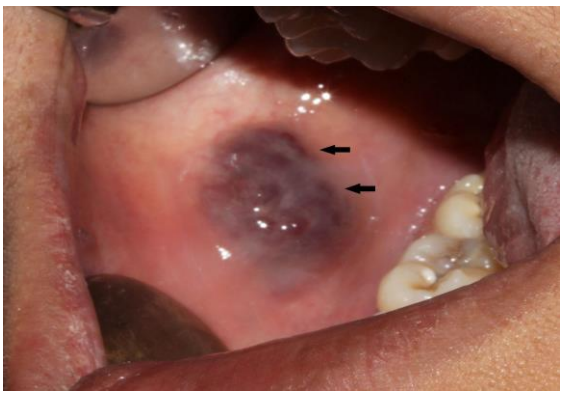

Fig. 3: Intra oral view showing bluish purple lesion on right buccal mucosa (black arrow)

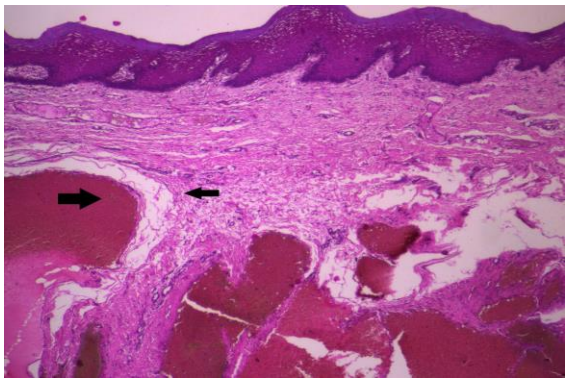

Fig. 4: Hematoxylin and Eosin section (10X) stain showing large dilated endothelial lined vascular spaces (black arrow)

DISCUSSION

Hemangiomas are the most common soft tissue lesions characterized by abnormal rapid proliferation of the endothelial cells.[1] The estimated prevalence of hemangiomas are 2-3% in neonates, 10-12% under age of 1 year and 22-30% in premature babies weighing less than1000g at the time of birth.[2, 3] Hemangiomas have female predilection with a ratio of 3:1 to 5:1.[2]

They can be classified as Benign Vascular tumours under the classification given by Mulliken et al in 1982. International Society for the Study of Vascular Anomalies (ISSVA) in their first workshop held in Rome during June 1996 revised Mulliken et al classification and broadly classified vascular abnormalities as vascular tumours consisting of Infantile hemangioma, Congenital hemangioma and vascular malformations according to the flow as capillary and venous malformations for slow flow and arterial malformation for fast flow.[4]

Congenital hemangiomas are present at birth itself. They may rapidly involute over a very brief period in infancy or never involute. The rapidly involuting ones are known as rapidly involuting congenital haemangiomas (RICH) and those which never involute are known as noninvoluting congenital haemangiomas (NICH).[5]

In infantile haemangioma, the lesion starts appearing at 8th week of intrauterine life. Infantile haemangiomas follow a predictable course with three distinct developmental phases: proliferation, quiescence and involution. In the first phase, hemangiomas rapidly proliferate within 12-18 months. Proliferation phase may be associated with ischemia, necrosis, ulceration and bleeding. Following proliferation phase, haemangiomas enter a slower or no growth phase, known as quiescence. The final and unique phase of haemangioma is knows as involution. This phase is marked by graying of the overlying skin and shrinking of the deeper components. 90% of infantile hemangiomas slowly involute by the age of 9 years.[5]

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] Also, abnormal levels of matrix metalloproteinases (MMP-9) and proangiogenic factors like vascular endothelial growth factor (VEGF) and basic fibroblast growth factor (b-FGF) play a role in haemangioma pathogenesis.[6]

Clinically, hemangiomas are red to purplish blue in colour. They appear as erythematous small clusters of red papules at about 6 months of age. They increase in size and may change their colour to bluish red by 5-7 years of age. Cavernous hemangiomas in adults do not regress and have a chronic course with slow progressive growth. Size can vary from few millimetres to some centimetres. They blanch on the application of pressure. Haemorrhage may occur either spontaneously or after minor trauma. They are generally painless.[7]

Histologically, hemangioma can be classified as capillary hemangioma and cavernous hemangioma. Capillary hemangiomas are characterised by many small capillaries lined by a single layer of endothelial cells supported in a connective tissue stroma of varying density. Cavernous hemangiomas are formed by large; thin walled vessels, or sinusoids lined by epithelial cells separated by thin layer of connective tissue septa. [8]

Differential Diagnosis includes Mucocele, Ranula, Superficial cysts, Varicosities and arteriovenous shunts. Mucocele, Ranula and Superficial cysts are mostly bluish in colour, located on the 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 elongated enlargement of superficial vein and are located typically on the ventral surface of the tongue. Arteriovenous shunts are rubbery, non fluctuant, throbbing and pulsatile.[9]

Syndromes associated with cavernous hemangiomas are Sturge Weber syndrome, Kasabach Merritt 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.

Radiographic imaging shows phleboliths as small round radiopaque concentric structures. Phleboliths are formed due to calcification of thrombus produced by slowing of peripheral blood flow. The fibrinous component then undergoes secondary calcification and gets attached to primary core of calcification.[10]

Magnetic Resonance Imaging is used for volumetric analysis by T1 and T2 weighted images. They are used for evaluating the size, extension and location of the lesions. Gradient Recalled Echo images are used to decide whether the anomaly is a high-flow lesion or slow-flow lesion.[10]

Colour Doppler ultrasound is also used to identify the feeding vessel, which is then ligated during the surgical procedure.[10]

Treatment depends up on the stage of growth in which the haemangiomas are present. Small isolated lesions are indicated for wait and watch. These small lesions should be checked for growth in size, haemorrhage, infection or ulceration. Proliferative stage of hemangiomas, multiple hemangiomas and hemangiomas affecting vital organs can be treated by systemic corticosteroids, propranolol, alpha interferon and anticancer drugs. Isolated small lesions which do not respond to systemic therapy and are deeply seated or are difficulty to access surgically can be treated with sclerotheraphy.[2]

5% sodium morrhuate or 5% ethanolamine oleate or sodium tetradecyl sulphate can be used as sclerosing agents. They cause an inflammatory response which leads to endothelial fibrosis. Cryotherapy and laser therapy can also be used for managing haemangiomas. Laser therapy using argon laser, pulsed dye laser and Nd: YAG laser acts on intravascular oxy haemoglobin resulting in vascular injury.[2]

Selective Embolisation is indicated when there is no response to systemic treatment for small lesions and for residual deformity after conservative or laser therapy.[2]

CONCLUSION

Hemangioma is a benign vascular abnormality of endothelial cells. Hemangiomas most commonly occur in head and neck region. Intraorally the most common location is buccal mucosa. Most of hemangiomas involute by age of 9-12 years without treatment. But some isolated hemangiomas do not involute and require treatment.

The dental practitioners should keep in mind that certain lesions can mimic hemangioma especially when the lesions are bluish red in colour. Also, considering the chances of heavy bleeding which can

occur during management of haemangioma, due care should be followed while surgical excision of such lesions.

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