Case Report

Numerous Vascular Malformations in Orofacial Region – A Diagnostic and Therapeutic Challenge

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ABSTRACT

Vascular lesions always remain a challenge as far as its diagnosis and treatment are concerned because of its close proximity to vital structures and risk of severe hemorrhage. Different types of vascular lesions like hemangiomas, vascular malformations are often encountered in day-to-day dental practice, but presence of multiple vascular lesions in the orofacial region is quite rare. Here, a rare and interesting case of multiple vascular malformations in the head-neck region of a 39-year-old female patient involving the tongue, cheeks, lips, palate and eyes has been reported. After getting prior informed consent and routine preoperative haematological investigations, an incisional biopsy was done from the buccal mucosal lesion and tongue lesion. Histopathological examination of the biopsied samples revealed the presence of non-dysplastic, parakeratinized stratified squamous epithelium backed by connective tissue stroma. The stroma contains numerous vascular channels containing red blood corpuscles within. No active endothelial proliferation was noted. Sprinkling of chronic inflammatory cells was also noted. Based on the said histopathological features, a diagnosis of vascular malformation was confirmed. Proper diagnosis and making differentiation between haemangiomas and vascular malformations is utmost important because of their different treatment plan.

Key words: Intraoral Vascular lesion, Numerous Vascular malformation, Orofacial.

Vascular lesions always remain a challenge as far as its diagnosis and treatment are concerned because of its close proximity to vital structures and risk of severe hemorrhage. Forty percent of all the vascular lesions are usually seen in the orofacial region [1]. Different types of vascular lesions like hemangiomas, vascular malformations are often encountered in day-to-day dental practice, but presence of multiple vascular lesions in the orofacial region is quite rare. Hemangiomas usually manifest at an early age of infancy and undergo a rapid growth due to endothelial cell proliferation. Finally, it involutes with increasing age. Whereas vascular malformations are nothing but structural anomalies of blood vessels without endothelial cell proliferation. Unlike Hemangiomas, vascular malformations persist throughout the life [2]. Such vascular lesions show a wide range of clinical manifestations starting from a simple erythematous macule to a pulsatile lesion, often leading to a massive post-traumatic hemorrhage [3]. The color of the overlying skin of such vascular malformations also varies according to the depth of its presence within the tissues. Those vascular malformations lying in the superficial layer of the tissues appear purple and the ones lying in the deeper layers appear greenish blue [4]. Vascular malformations have an overall incidence of about 1 in 10000 [5]. Vascular malformations are usually locally aggressive and may undergo enlargement during puberty or adolescence resulting in an expansile lesion thereby causing both

cosmetic as well as functional disturbances. Such expansile mass often destroy adjacent normal structures and lead to certain complications viz. disfigurement of tissues, severe bleeding, painful ulceration, venous congestion, thrombosis, thrombolysis, localized intravascular coagulopathy (LIC) and disseminated intravascular coagulopathy (DIC) [6,7]. Here, the author described a rare and interesting case of multiple vascular malformations in the head-neck region of a 39-year-old female patient.

CASE REPORT

An otherwise healthy 39-year-old female patient (Figure 1) reported to the out-patient department of a tertiary health care centre situated in Kolkata of West Bengal. The patient complained of numerous abnormal nonspecific swellings on the tongue, cheeks, lips, palate and eyes. The swellings were slow growing, bluish green in colour and were not painful. Patient also reported about heavy bleeding following minimal trauma to those swellings.

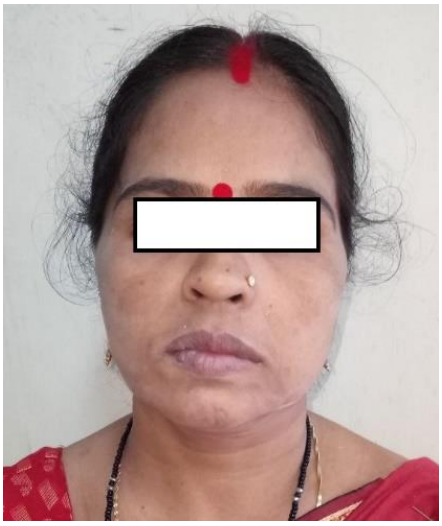

Figure 1: Frontal profile of 39-year-old woman

Patient's family history, general medical history was nil of note. Patient also denied about any oral deleterious habits. On extraoral examination, there was soft, non-tender, dome-shaped, compressible and reducible bluish swellings over right upper eyelid. Similar type of lesions was also noted within the eyes (Figure 2a, 2b, 2c).

Peri orally, there was soft bluish green coloured non-tender lesion on right side of lower lip near vermilion border (Figure 3a).

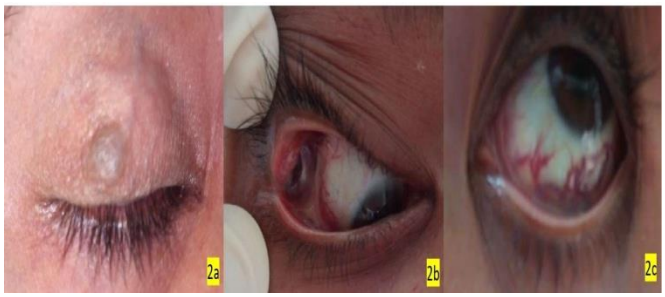

Figure 2: 39-year-old woman with multiple soft bluish swellings (a) on right upper eye lid; (b) inner aspect of right eye; (c) inner aspect of left eye.

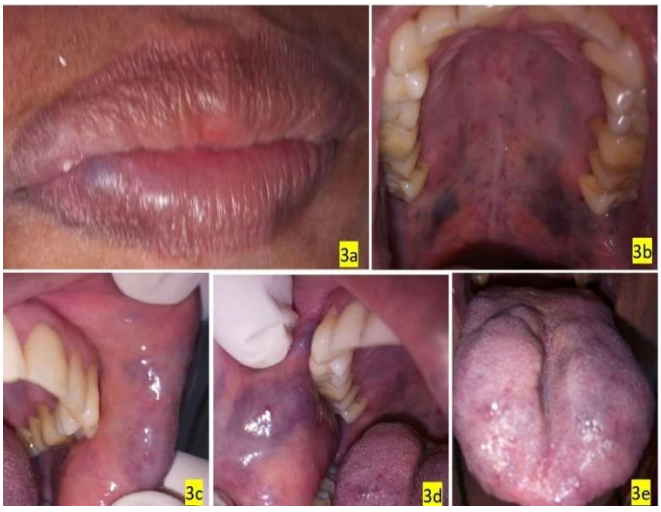

Figure 3: 39-year-old woman with multiple soft bluish swellings (a) on right side of lower lip; (b) posterior part of palate; (c) left labial mucosa; (d) right buccal mucosa near angle of the mouth; (e) dorsum of tongue.

Intra-oral examination revealed bluish, compressible non-tender swelling on either side of dorsum of tongue, right buccal mucosa near angle of the mouth, left upper labial mucosa and posterior region of palate (Figure 3b, 3c, 3d, 3e). Diascopy was done and blanching was noted. Aspiration revealed fresh blood (Figure 4).

Figure 4: Aspirated blood

Based on the above said clinical findings, a provisional diagnosis of vascular malformation was done. After getting prior informed consent and routine preoperative haematological investigations, an incisional biopsy was done from the buccal mucosal lesion and tongue lesion.

Histopathological examination of the biopsied samples revealed the presence of non-dysplastic, parakeratinized stratified squamous epithelium backed by connective tissue stroma. The stroma contains numerous vascular channels containing red blood corpuscles within (Figure 5). No active endothelial proliferation was noted (Figure 6). Sprinkling of chronic inflammatory cells was also noted. Based on the said histopathological features, a diagnosis of vascular malformation was confirmed.

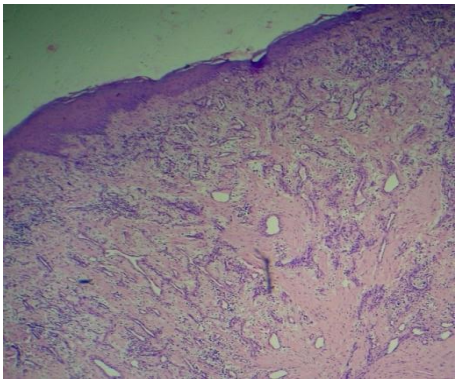

Figure 5: 10X view of H & E-stained section showing parakeratinized stratified squamous epithelium backed by connective tissue stroma which contains numerous vascular channels.

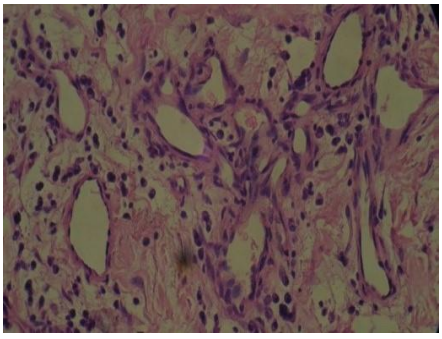

Figure 6 : 40X view of H & E-stained section showing numerous blood vessels without endothelial proliferation.

The patient was referred to the Department of Oral surgery for further management under a multidisciplinary team consisting of an oral and maxillofacial surgeon, haematologist and vascular malformation specialist. Considering the risk of heavy bleeding per-operatively, initial treatment was done with sclerosing agents for management of intraoral lesions. Here, Sodium tetradecyl sulphate, a sclerosing agent, was injected within the lesion at a regular weekly interval. It works by increasing the formation of blood clots and scar tissue inside the vessels. Thus, the size of the vascular lesions decreased and bleeding also subsided due to fibrosis of the vascular channels. Then the patient was called for regular follow up at monthly interval. Patient reported with neither any adverse effects nor any exacerbation of the lesion within next one year.

DISCUSSION

Vascular malformations may be defined as a heterogeneous group of vascular aberrations having a spectrum of various subtypes differentiated by its vessel type, rate of blood flow, and size [8]. They can be classified into three types viz. capillary, venous and arterial based on the type of vessel involved. According to the haemodynamic features, vascular malformations can be low flow or high flow.

The pathogenesis of such vascular malformations is not clear till date. Various researchers have tried to explain the reason behind such defective morphogenesis of blood vessels. Few genes and its certain forms of mutation causing such anomalies have been identified. Such as germline mutations in the TEK gene (chromosome 9p), encoding the endothelial cell (EC) tyrosine kinase receptor TIE2, resulting in cutaneo-mucosal venous malformation (VMCM) due to increased phosphorylation of TIE2. 40% of sporadic vascular malformations are due to somatic mutations in TIE2 [9].

Histopathological features of vascular malformation show numerous engorged blood vessels within the stroma, which are packed with red blood cells. Endothelial cell proliferation is absent. To distinguish between vascular tumours/ neoplasms and vascular malformations, Wilms' tumor-1 (WT-1) immunoexpression can be done. Vascular malformations usually show negative WT-1 immunoexpression [10].

The main risk factor associated with the management of such malformations is heavy bleeding and chances of developing LIC, which further predisposes during surgical management. Oral mucosal vascular malformations are also a challenge because of increased chances of trauma either during having food or undergoing any dental procedure. So, this type of clinical case with multiple lesions in the orofacial region should be carefully dealt with.

CONCLUSION

Proper diagnosis and making differentiation between haemangiomas and vascular malformations is utmost important because of their different treatment plan. Haemangiomas usually regress with advancing age, but malformations persist through out the life and may increase in size resulting in cosmetic disfigurement and functional disturbances, thereby hampering patient's daily activities. The treatment of vascular malformations is also complex and requires multidisciplinary involvement.

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