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CASE REPORT

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Giant Haemangio- Lymphangioma of The Upper Lip- A Case Report

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Background: Vascular anomalies are congenital defects in vasculogenesis. They are classified into vascular tumors (hemangiomas) and vascular malformations (venous malformations, arteriovenous malformations, lymphatic malformations). Sometimes, lymphatic channels may be evident in hemangiomas. Depending on predominant component, it will be categorized either as hemangiolymphangiomas or lymphangiohemangiomas, vice versa is true for lymphangiomas. Hemangiomas of the oral cavity are not commonly observed though head and neck are common sites.

Case presentation: The patient was a 13-year-old female who presented herself to the Department of Oral and Maxillofacial surgery at Dental college, with the complaint of swelling in upper lip since 12 years. The swelling was soft in consistency and bluish in colour. The lesion was diagnosed as hemangioma after clinical examination and excised under general anaesthesia. Histopathology was consistent with Hemangio-lymphangiomas.

Conclusion: Hemangio-lymphangiomas are not common in oral cavity but all suspicious cases should be properly investigated. Early detection and biopsy is necessary to determine the clinical behavior of the tumor and potential dentoalveolar complications.

KEYWORDS

Hemangio-lymphangioma; lip; hemangiomas; oral cavity

1 | INTRODUCTION

Vascular tumors are endothelial neoplasms having the hallmark feature of increased cellular proliferation, the most common to which is hemangioma exclusively found in infants which arises because of abnormal development of vascular elements during embryogenesis and fetal life. Endothelial turnovers are generally not demonstrated by these malformations which are classified according to the predominant channel type as capillary malformations, lymphatic malformations, venous malformations, arteriovenous malformations, and complex forms like capillary- venous malformation.[1] 75 % of all cases

[*]HK, AKJ: Conceptualization, Methodology, HK,SV: Data curation, Writing- Original draft preparation. YTL: Visualization, Investigation.

of lymphangioma occur in the head and neck region, of which 50 % of all lesions appear at birth. Around the age of 2 years 90% of them are developed.[2] Tongue, palate, gingiva, buccal mucosa, lips, and the alveolar ridge of the mandible are the most commonly affected sites in the oral cavity, tongue being the most preferred site. Lymphangioma is rarely seen in buccal mucosa.

The incidence of lymphangiomas has been reported to range from 1.2 to 2.8 per 1000 newborns.[3] The most prominent sign or symptom of all lymphangiomas is the presence of a mass. Surgical excision is the treatment of choice. The prognosis is good for most patients, although large tumours of neck/tongue may result in airway obstruction and death. We here report a case of Haemangio-lymphangioma of the upper lip, and reviewing the literature.

2 | CASE REPORT

A 13-year-old female patient reported to the Department Of Oral And Maxillofacial Surgery, Sri Rajiv Gandhi College of Dental Sciences and Hospital, Bangalore, Karnataka. Her chief complaint was swelling in upper lip since 12 years. Upper lip shows diffuse swelling measuring about 5x 2cm in size. The extension of swelling was from left side corner of the mouth to right side corner of the mouth and surface is tensed. Swelling was non-tender with incompetent lips. The consistency was soft to firm and was non reducible in nature [Figure 1(a)]. Provisional diagnosis of haemangioma was made.

Excision of the lesion was carried out under general anaesthesia. Intense bleeding was noted which was controlled using electro-cautery. Complete wide excision and primary closure of the surgical site with double layer suture technique was done [Figure 1(b)]. Specimen obtained was soft in consistency and bluish in colour. [Figure 2]

3 | HISTOPATHOLOGY

The H & E stained soft tissue section shows keratinized epithelium with intracellular oedema and acanthosis. Connective tissue shows numerous dilated irregular endothelial lined lymphatic channels containing eosinophillic homogenous material suggestive of lymph fluid. Loosely arranged fibro cellular stroma also shows endothelial lined capillaries and chronic inflammatory cell infiltration [Figure 3 (a) & (b)]

4 | DISCUSSION

Lesions larger than 5 cm are term as giant hemangioma. In our case also the measurements was found to be larger than 5 cm hence we could consider it as a giant hemangio-lymphangioma. Histologically, hemangiomas in the proliferating phase show endothelial hyperplasia and large number of mast cells. In contrast, vascular malformations show normal number of mast cells, and consist of mature, often combined, capillary, arterial, venous, and lymphatic elements. In the histopathological picture of the case presented we did not find any mast cells. However, in some instances the channels may be filled with blood, a mixed hemangio-lymphangioma which can invade the underlying tissue and recur locally. This feature differentiates it from simple lymphangioma and hemangioma. In the presented case multiple large sized lymphatic channels were filled with lymph and RBC's, lined by endothelium were noted. So the final diagnosis as hemangio-lymphangioma was made.

Lymphangioma is a rare, benign, congenital disease of unknown aetiology. Origin of the lesion is considered abnormality of the lymphatic system rather than a true neoplasm. The first description of lymphangioma in the literature is credited to Redenbacher who, in 1828, referred to a lesion as a ranula congenital.[4] Virchow (1854) gave the first accurate description of lymphangioma.[5] In 1872 Krester hypothesized that hygromas were derived from lymphatic tissue.[6] Hemangiomas are endothelial tumors with a unique biologic behaviour. The three stages in the life cycle of a hemangioma are (1) the proliferating phase (0–1 year of age), (2) the involuting phase (1–5 years of age), and (3) the involuted phase (>5 years of age). These stages are typically clinically apparent and can be distinguished microscopically and immunohistochemically. In the proliferating phase, the hemangioma is composed of plump, rapidly dividing endothelial cells that form tightly packed sinusoidal channels. Even at this early stage, the endothelial cells express phenotypic markers of mature endothelium , in addition to markers of activated endothelium. Urinary markers of angiogenesis, such as basic fibroblast growth factor and high molecular weight (MW)

matrix metalloproteinase (MMPs) are usually high in infants with proliferating haemangiomas and diminish to normal levels during regression. In the involuting phase, there is decreasing endothelial proliferation, increasing apoptosis, and the beginning of fibro-fatty replacement of the hemangioma.. During the involuted phase, after regression is complete, all that remains are a few tiny capillary-like feeding vessels and draining veins (some of which can be abnormally large) surrounded by islands of fibro-fatty tissue admixed with dense collagen and reticular fibres. The endothelium lining these vessels is flat and mature. Multilaminated basement membranes persist around the residual tiny capillary-sized vessels.[7]

The clinical appearance of lymphangioma depends on the extension of the lesion. Superficial lesions consist of elevated nodules with pink or yellowish colour. Usually, there is a plaque constituted from small vesicles with thin walls, translucide like frog eggs. They may feel like a ball of worms on palpation, but are usually rather nonspecific and ill-defined. Often with blue-black or deep red hemnorrhagic blebs.[8] Deeper lesions are described as soft, diffuse masses with normal colour. Oral lymphangioma may occasionally assume a red or blue aspect because of the rupture of blood capillary into the lymphatic inner spaces.[9] These lesions are three to five times more common in females, and we are also reporting a female patient with hemangio- lymphangioma of upper lip. There is an increased frequency of hemangiomas in premature infants with a reported incidence of 23% in neonates who weigh less than 1200 g.[1] In the presented case also the swelling was initially small in size which gradually increased to the present size. The swelling was not associated with pain. In adult patients, neoplasm can switch to squamous cell carcinoma. The possible etiological factors include VEGF-C, vascular endothelial growth factor receptor 3 (VEGFR-3), and transcription factor Prox-1.VEGF-C and VEGFR-3 have been shown to be upregulated in lymphatic malformed tissue, and both are involved in lymphatic tissue proliferation.

The treatment of lymphangioma depends upon their type, size, involvement of anatomical structures and infiltration to the surrounding tissues as well as the risk of complications.[8] There are various other treatment modalities like injection of fibrosing agents intralesionally, radiation, cryosurgery, embolization, electrocoagulation, interferon a-2b, laser therapy and plasma knife surgery.[10] Microcystic lesions do not respect tissue planes, are diffuse and difficult to eradicate, whereas macrocystic lesions are localized and easily excised.

The most effective treatment is surgery when vital structures are not involved and the aims are to restore adequate breathing and swallowing, leave a tongue capable of normal speech, taste, sensation and oro-facial development and to achieve a good cosmetic result. However surgical complications, which occur in 19 % to 33 % of the cases, include formation of hematoma, lymphocele, scar, abscess, infection, wound dehiscence and nerve palsy. Nerve involvements also leads to complications such as dysphagia, dysphonia, and difficulty with breading. Some clinicians do not recommend surgery for non-enlarging lymphangiomas of the tongue because of difficulties in removal and the high recurrence rate (39%). The most common sites for recurrence are the tongue and hypoharynx and/or larynx.

5 | CONCLUSION

Hemangio-lymphangioma is a rare entity encountered in the oral cavity. The early diagnosis can help in appropriate treatment and prevent possible complications. Early detection and biopsy is necessary to determine the clinical behavior of the hemangio-lymphangioma and potential dento-alveolar complications.

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Nil

Conflict of interest

The authors have no conflicts of interest to declare

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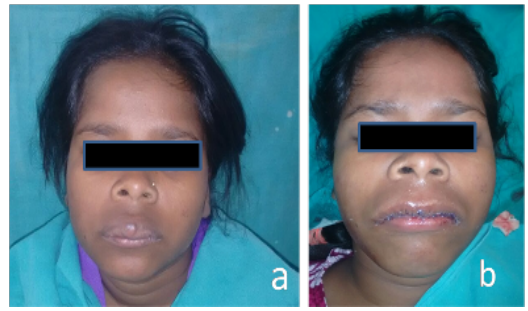

FIGURE 1 (a) Pre-operative picture showing huge swelling on upper lip. (b) Post- Surgical View

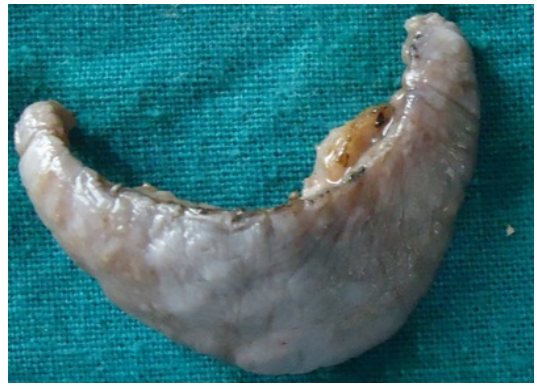

FIGURE 2 Grossing specimen was greyish white in colour, firm in consistency measuring about 5x2 cm and 3x1 cm in size.

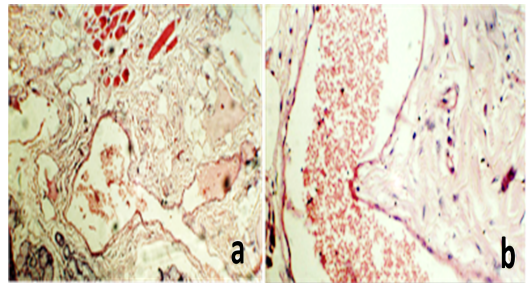

FIGURE 3 (a) H & E stained section showing lymph vessels containing red blood cells (Magnification 10x) (b) H & E stained section showing multiple large lymph vessels and blood vessels (Magnification 10x)