Lymphangioma of cheek region—an unusual presentation

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ABSTRACT

Background and Objectives: Lymphangiomas are benign hamartomatous tumors of the lymphatic vessels. A lymphatic malformation is a congenital defect that occurs during early embryonic development when the lymphatic vessels do not properly form. The vessels may become blocked and enlarged as lymphatic fluid collects in the vessels, forming a mass or a cyst. About 50% of all lesions are noted at birth and around 90% develop by 2 years of age.

Materials and Methods: We report the case of lymphangioma, with an unusual presentation in the cheek region resulting into vestibular obliteration and expanding to the left maxilla diagnosed on the basis of histopathology. The review of literature concerning the clinical and histological features as well as the proper management concerning this hamartomatous entity is included.

Results and Conclusion: The various treatment modalities for lymphangioma are surgical excision, radiation therapy, cryotherapy, electrocautery, sclerotherapy, steroid administration, embolization, ligation, laser surgery with Nd-YAG and CO₂ and radiofrequency tissue ablation technique. An early diagnosis and intervention helps in reducing the functional, psychological disturbances and cosmetic disfigurement.

Keywords: Hamartoma, lymphangioma, maxilla.

INTRODUCTION

Lymphangiomas are rare benign hamartomatous lesions of the lymphatic system. They rarely represent as neoplasm; most likely present as developmental malformations arising from the sequestration of the lymphatic tissue that do not communicate with the rest of the lymphatic channels.¹ They have been classified as (1) lymphangioma simplex (capillary lymphangioma) consisting of small, capillary-sized vessels; (2) cavernous lymphangioma, comprising large, dilated lymphatic vessels, and (3) cystic lymphangioma exhibiting large macroscopic cystic spaces.²

Lymphangiomas have a marked predilection for head and neck which accounts for about 75% of all cases. About 50% of all lesions are noted at birth and around 90% develop by 2 years of age. Oral lymphangiomas are most frequent on the anterior two-thirds of the tongue where they often result in macroglossia.³ Small lymphangiomas <1 cm in dimension occur on the alveolar ridge in about 4% of black

neonates. These lesions occur bilaterally on an alveolar ridge.⁴ Lymphangioma simplex involves the cluster of vesicles measuring about 2–4 mm. These clear vesicles range from pink to red to black due to secondary hemorrhage. Cavernous lymphangiomas appear as subcutaneous nodules, with a rubbery consistency. They have a clear dimension than lymphangioma simplex. The overlying skin has no lesions or changes. Cystic lymphangiomas are usually larger than cavernous lymphangiomas and more commonly occur in the head and neck in the parotid region.⁵

CASE REPORT

A 15-year-old boy, accompanied by his father, reported to the outpatient department, with complaint of poor esthetics and difficulty in speech due to unerupted permanent left upper front two teeth since 9 years. Moreover, the patient

Figure 1 Intra-oral photograph revealing dento-alveolar enlargement and vestibular obliteration.

gave a history of congenital swelling in the left upper face region since birth and he had forceps delivery at the time of his birth.

Extra-oral examination revealed facial asymmetry with respect to the left middle third of the face region, extending from the left lateral surface of ala of the nose, superiorly to the left inferior border of the orbital rim, and extending to the left ala tragus line. The diffuse swelling measured around 4×4 cm in diameter, and obliterated the left nasolabial groove. On palpation, swelling was soft, nonfluctuant, nonreducible and compressible to some extent, associated with left solitary submandibular lymphadenopathy.

Intra-oral examination revealed retained deciduous teeth 61 62, palatoversion 22 23, class I molar relation and missing 21. Soft tissue examination revealed diffuse dento-alveolar enlargement with respect to 22 23 and vestibular obliteration (Figure 1).

The differential diagnoses put forth were chronic inflammatory cyst and hamartomatous lesion. The patient was subjected to varied hematological investigations that were found to be within normal limits. Posteroanterior skull projection revealed an expansile lesion with respect to the left anterior maxillary region. Under local anesthesia, infiltration was given and vestibular incision was made. Labial vertical bone in 22 23 region was exposed; buccal window of 5×5 mm was made; bone piece and small amount of the soft tissue were removed that were sent for histopathological examination.

The soft tissue specimen revealed fibrous connective tissue exhibiting diffuse, dense, predominantly chronic inflammatory cells, extravasated red blood cells and capillaries. A small fragment of the connective tissue was associated with a nonkeratinized stratified squamous epithelium. A few

Figure 2 Focal area of dense dilated spaces with fibrous connective tissue.

empty spaces, filled with amorphous eosinophilic material, were seen in the connective tissue. The decalcified tissue section showed spicules of vital bone associated with the fibrous connective tissue marrow exhibiting chronic inflammatory cells, infiltrate and extravasated blood cells, and the focal area of dilated spaces suggestive of lymphangioma (Figure 2). On subsequent follow-up, when the wound was healed, orthodontic correction and prosthodontic rehabilitation were carried out that improved the esthetics of the patient.

DISCUSSION

Lymphangiomas are rare benign hamartomatous lesions of the lymphatic system. These lesions, more correctly referred to as the lymphatic malformations, have a marked predilection for the head and neck region. No clear sex predilection is demonstrated, and whites appear to be affected more often than people of other races. Embryologically, it is derived from five primitive buds developing from the venous system which include paired jugular sacs, paired posterior sacs and a single retroperitoneal sac.²

Since the lesions are translucent, transillumination is positive unless there is secondary hemorrhage into the lesion. Magnetic resonance imaging can define the degree of involvement and entire anatomy of the lesion. Immunohistochemical studies for laminin show typical multilayered basal lamina in lymphangiomas.⁶

The treatment of lymphangiomas usually consists of surgical excision. Total removal may not be possible in all cases because of the large size or the involvement of the

vital structures. Recurrence is common especially for cavernous lymphangioma, which are more radioresistant and insensitive to sclerosing agents such as sodium morrhuate than hemangiomas.

The various treatment modalities for lymphangioma are surgical excision,⁷ radiation therapy, cryotherapy, electrocautery, sclerotherapy, steroid administration, embolization and ligation, laser surgery with Nd-YAG and CO₂ and radiofrequency tissue ablation technique.^{8–10} An early diagnosis and intervention helps in reducing the functional, psychological disturbances and cosmetic disfigurement.

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