ORIGINAL RESEARCH PAPER

Oral Medicine

LYMPHANGIOMA OF CHEEK – AN UNUSUAL PRESENTATION

KEY WORDS: hamartoma, Turners syndrome, Noonan syndrome

Aditi Tamrakar Post Graduate student, Department of Oral Medicine & Radiology, Manipal College of Dental Sciences, MAHE, Manipal

Komal Smriti* Assistant Professor, Department of Oral Medicine & Radiology, Manipal College of Dental Sciences, MAHE, Manipal*Corresponding Author

R Vineetha Professor and Head, Department of Oral Medicine & Radiology, Manipal College of Dental Sciences, MAHE, Manipal

ABSTRACT

Lymphangioma is a hamartoma of the lymphatic vessels and is categorized as a developmental lesion instead of true neoplastic tumor. This entity was described first by Redenbacher in 1828. According to the literature search, approximately 50% of lesions are present at birth and about 90% appear by early childhood. It has strong tendency of occurrence in the head and neck area. Anterior two-third region of the tongue is the most common site of occurrence of oral Lymphangioma resulting in macroglossia. They are reported to be coexistent with Turners syndrome, Noonan's syndrome, aneuploidies and fetal alcohol syndrome. They are relatively uncommon in buccal mucosa. Here we report one such rare case of Lymphangioma of right buccal mucosa in a 14-year-old boy.

INTRODUCTION

Lymphatic system is a diffuse network of endothelial channels, appears first during the 6th week of intrauterine development and further grows from five primitive sacs budding from the embryonic peripheral venous system [1]. Lymphatics channels are extensive unidirectional systems with the blunt endings, which retrieves excessive fluid from interststitium and carries it to local lymph node and drain into venous system by the means of thoracic duct. They also help in absorption of protein and lipid from liver and small intestine, respectively. Lymphangioma is considered a hamartoma of the lymphatic vessels and is categorized as a developmental lesion instead of true neoplastic tumor. This entity was described first by Redenbacher in 1828 [2].

According to the literature search, approximately 50% of lesions are present at birth and about 90% appear by early childhood. It has strong tendency of occurrence in the head and neck area. Anterior two-third region of the tongue is the most common site of occurrence of oral Lymphangioma resulting in macroglossia [3].

The lesions are composed from ectopic lymphatic system of lymphatics separated from the normal network of lymphatic vessels, yet communicating with the superficial lymphatics that become dilated by the continuous rise and fall of the pressure that is transmitted from the muscular walls of the deep cisterns. Thus, the blockage can determine the growth of the hydrostatic pressure with consecutive expansion of the lesions, until an equal pressure with the contiguous tissues will be realized. They are reported to be coexistent with Turners syndrome, Noonan's syndrome, aneuploidies and fetal alcohol syndrome. They are relatively uncommon in buccal mucosa. Here we report one such rare case of Lymphangioma of right buccal mucosa [4].

CASE REPORT

A 14-year-old male child reported to the outpatient department of oral medicine and radiology with the main complain of diffuse swelling on the right side of the face in the past 10 years. Patient's mother reported that mild swelling was present on the right side of his face since childhood approximately at the age of 3-4 years which gave more fullness to the right cheek. The swelling was asymptomatic and constant in size up to the age of 10-11 years but later it started gradually increasing in size. Three years back there was sudden increase in size of swelling with pain which subsided to some extent on taking antibiotics and analgesics. Two weeks back patient again noticed increase in size of swelling which has grown up to the current size with which patient presented to us. No history of trauma to the corresponding region. No pain or mobility associate with any tooth. No difficulty in chewing, swallowing, mouth opening and speech. No history of swelling elsewhere in the body. Patient did not report of nose block, epiphora, epistaxis or change in vision.

On extraoral examination there was marked facial asymmetry due to dome shaped swelling present on the right side of the face. It extended from right canthomeatal line to lower border of mandible super inferiorly and from right nasolabial fold to right preauricular region anteroposterior approximately 4 cm x 5 cm in dimension. Overlying skin was tense with no alteration in color. Reddish vesicles 7-8 in number were evident around the ala of nose but no pus discharge or sinus opening was appreciable. On palpation the swelling was soft to firm in consistency and fluctuation was present. No pulsation or bruit was felt [Fig. 1].

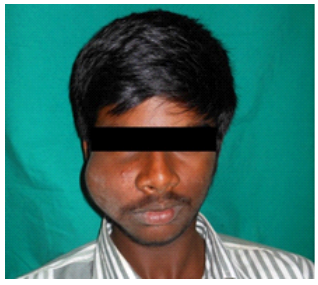

FIG. 1

Detailed **intraoral examination** revealed fullness of right buccal mucosa with obliteration of right maxillary buccal sulcus. Reddish vesicles, firm in consistency,4-5 in number on attached gingiva & depth of vestibule giving tapioca pudding appearance were evident [Fig. 2].

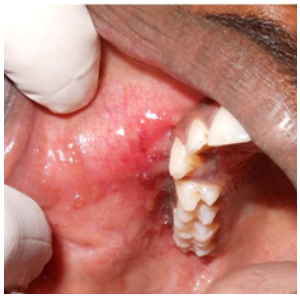

FIG. 2

No teeth in corresponding region were carious, mobile or discolored however cross bite present on right side indicated pressure effect of the lesion on the dentition. Keeping the above features in view a provisional diagnosis of Hemangioma was given. Lymphangioma and vascular malformation was given as

differentials. As chairside investigation vitality test of all the teeth of first quadrant was done which revealed all of them as vital. **An orthopantomogram** was done which showed no teeth associated pathology and gave clear indication of lesion completely in the substance of buccal FIG. 3mucosa [Fig. 3].

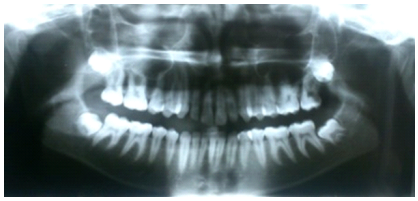

FIG. 3

Furthermore, **ultrasound imaging** of right middle third of face was performed which exhibited, large lobulated multifocal, anechoic lesion with low level internal echoes and echogenic internal septations measuring 5.5x 2.7x4.2 cm in the subcutaneous plane of right cheek superficial to the buccinators and masseter muscle [Fig. 4]. Color Doppler showed no evidence of color in flow suggestive of lymphatic malformation.

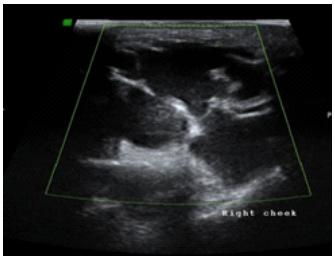

FIG. 4

To know the complete extent of lesion with precision Magnetic Resonance Imaging was performed which illustrated a well-defined lobulated multiseptated non-enhancing cystic lesion which was hyper intense on T2 measuring 4.3 X 3.9 X 5.5 cm [Fig. 5].

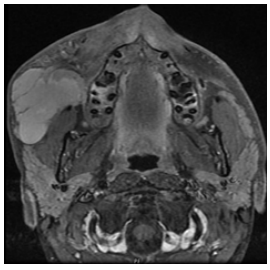

FIG. 5

It was situated in the subcutaneous and muscular plane, antero-lateral to the masseter muscle and anterior to the superficial lobe of parotid gland in the right cheek and maxillary region up to ala of the nose. There was no evidence of calcification in fluid levels within the lesion. These features were suggestive of Lymphangioma.

As the extent of lesion was large fine needle aspiration cytology was performed which exhibited clear fluid with scanty blood cells predominantly lymphocytes thus indicative of lymphatic malformation. Biopsy from the lesion was obtained which was routinely processed. Hematoxylin and eosin stained section showed stratified squamous epithelium with juxta epithelial stroma showing large thin endothelial lined vessels, few vessels showed eosinophilic material resembling lymph and few lymphocytes. Based on this a final diagnosis of simple lymphangioma based on Watson and Mc McCarthy classification was made.

DISCUSSION

The lymphangioma is a hamartomatous proliferation of lymphatic vessels, accounting more than three-fourths of cases which are reported in the head and neck region. This tumor is thought to be a developmental in origin with abnormal malformation of vessels with poor communication to the normal lymphatic vessels. Most of the cases are superficial in origin but may extend deep into the connective tissues [5,6]. Most of the lesions seen in lymphangioma are single and rarely multiple lesions are reported in childhood and may pose a life-threatening disease when involvement of viscera occurs.

A classification of the lymphangioma has been suggested by Watson and McCarthy based upon their study of 41 cases. In this classification the following divisions are proposed: (1) simple lymphangioma, (2) cavernous lymphangioma, (3) cellular or hypertrophic lymphangioma, (4) diffuse systemic lymphangioma, and (5) cystic lymphangioma or hygroma [7,8].

Clinical Features: the majority of cases of lymphangioma are present at birth, according to Watson and McCarthy, in whose series 95 per cent of the tumors had arisen before the age of 10 years. In the series of 132 patients reported by Hill and Briggs, 88 per cent of the lesions had developed by the end of the second year of life [9]. In contrast to that of the hemangioma, the gender distribution of the lymphangioma is nearly evenly divided. Watson and McCarthy reported head and neck as the primary site of tumor in 52 percent of cases. The most common location in head and neck region is the lateral neck, and is commonly called cystic lymphangioma or cystic hygroma [10].

ORAL MANIFESTATIONS

Tongue is the most common location followed by palate, buccal mucosa, gingiva and lips. The lesions on the surface are manifested as papillary lesions which may be of the same color as the surrounding mucosa or of a slightly redder hue. The lesions which are deeper in origin does not show any change in surface texture or color. In some cases, relatively large areas of tissue may be involved. If the tongue is affected, considerable enlargement may occur, and to this clinical feature the term 'macroglossia' may be applied. The irregular nodularity of the surface of the tongue with gray and pink projections is the commonest sign of the disease, and when associated with macroglossia, is pathognomonic of lymphangioma [11].

Lip involvement and its attendant deformity are referred to as macrocheilia. The cystic hygroma is a common and distinct entity that is not manifested in the oral cavity but occurs in the neck as a large, deep, diffuse swelling. This has been discussed in particular by Bill and Summar and by Paletta [12].

An unusual form of lymphangioma termed lymphangioma of the alveolar ridge in neonates has been reported by Levin and his associates. They found domed shaped bluish fluid lesions on the alveolar ridges in 55 (3.7 per cent) of 1,470 normal black newborns; none were found in whites. Histologically, those biopsies were lymphangiomas [13]. The natural history of this lesion is unknown, although spontaneous regression was noted in several cases. Occasional cases of central lymphangioma of bone are also known to occur such as that in the tibia reported by Bullough and Goodfellow, as well as in the jaw. There categories are somewhat artificial and many lesions are combinations of categories [14]. Treatment and Prognosis

The recommended treatment is surgical excision as tumor is radio resistant and not sensitive to local sclerosing agents, such as sodium morrhuate [8]. Spontaneous regression, according to most studies is rare. Lymphangioma is non-encapsulated and infiltrating, thereby, removal of complete tumor is often impossible and is difficult to salvage normal structures. Removal of the bulk of the tumor by surgical means is the treatment of choice especially in a growing child [8,15]. In our case also choice of surgical excision was given but patient's parents refused to seek so due to expense of the treatment. Patient was recalled to review after 1month but he was lost to follow-up.

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