Isolated Lymphangiomatous Polyp Nasopharynx in an Adult First Case Report in English Literature

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Abstract Lymphangiomas are rare benign, hamartomatous, congenital malformations of the lymphatic system involving the skin and subcutaneous tissues of head, neck and oral cavity. Occasional adult onset cases occur, this condition is thought to be a developmental malformation of lymph vessels which have poor communication with normal lymph system. Most of these malformations are present at birth or appear within two years of life. 75 % of cases occur in head and neck area, submandibular and parotid being the most affected parts. Lymphangioma arising in nasopharynx and in an adult has not been reported in english literature. This prompted us to report the very first case of Lymphangioma Nasopharynx.

Keywords Lymphangioma · IHC-immunohistochemistry · IHT-Immunohistochemistry · Hamartoma

Introduction

The lymphangioma is a benign hamartomatous, congenital hyperplastic malformation of lymphatic vessels. Though these malformation can occur at any age and in any part of the body but three-fourth of cases occur in the skin and subcutaneous tissues of head and neck region. Lymphangiomas are rare, accounting for 4 % of all vascular tumors in children. Although lymphangiomas can become evident at any age, 50 % are seen at birth and 90 % of lymphangiomas are evident by 2 years of age. Orvidas and

Kasperbauer in their review of 67 cases in pediatric age group found that submandibular area is most commonly affected (37 %) followed by parotid in 31 %. Most of lymphangiomas are benign, soft, slow growing doughy mass. Rarely they may impinge upon the vital structures of head and neck and may cause complications. Lymphangiomas arising in the nasopharynx is not reported in English literature. This prompted us to report this case, the first and the only one in an adult.

Case Report

Patient BS 18 years male attended our outpatient department with complaints of irritation of the posterior part of the throat for the last 1 year. He noticed an elongated structure in this throat hanging from above which was progressively increasing for the last 4 months. He had change in his voice. On examination, a pedunculated mass was seen in the right side of throat (Fig. 1). Nasal endoscopy done showed the soft mass attached to the posterior lip of the Eustachian tube opening. No bleeding on touch. CT scan neck showed a tubular well defined polypoidal mass 24×07 mm, attached to the opening of eustachian tube on right side with no invasive character (Fig. 2). The patient was operated under GA and the mass measuring 3.5×1 cm was removed endoscopically (Fig 3). The base/attachment of the mass was cauterized. No operative/postoperative complications. The histopathology examination revealed it to be consisting of stratified surface mucosa with small to large vascular spaces lined by flat endothelium, filled with lymph with collection of small mature lymphoid cells in fibrous stroma outside the spaces (Fig 4a, b). Immunohistochemistry for factor VIII related

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Fig. 1 Elongated mass seen on Right side Nasopharynx

antigen are weakly positive in the lining endothelial cells along vascular spaces (Fig. 5a, b).

Discussion

The lymphangiomas are the benign hamartomatous hyperplasia of lymphatic vessels. The tumor is thought to be a developmental malformation of vessels with poor communication with the normal lymph system. Occasional adult onset cases do occur [1]. Isolated lymphangiomas have been reported arising from different structures of head and neck. But lymphangioma arising from the nasopharynx has not been reported in english literature. A single case of lymphangioma in nasopharynx has been reported in Japanese literature [2].

These malformations can occur at any age and may involve any part of the body, but 90 % occur in children less than 2 years of age and involve the head and neck. These malformations are either congenital or acquired.

Fig. 2 CT Scan showing mass arising from the Eustachian Tube opening in nasopharynx

Fig. 3 Excised mass

Congenital lymphangiomas are often associated with chromosomal abnormalities such as Turner syndrome, although they can also exist in isolation. Lymphangiomas are commonly diagnosed before birth using fetal ultrasonography. Acquired lymphangiomas result from trauma, inflammation or lymphatic obstruction. Orvidas and Kasperbauer in their review of 67 cases in pediatric age group found that submandibular area is most commonly affected (37 %) followed by parotid in 31 %. 35 % of all lymphangiomas occur in cheek, tongue and floor of mouth, 25 % in the neck and 15 % in the axilla [3]. Most of lymphangiomas are benign, soft slow growing doughy masses. Rarely they may impinge upon the vital structures of head and neck and may cause complications.

Histologically, lymphangiomas consist of multiple, interwining lymph vessels in a loose fibrovascular stroma.

The lymphangiomas are classified as (1) Capillary lymphangiomas or microcystic or lymphangioma circumscriptum composed of small, thin walled, capillary-sized lymphatic vessels and are characteristically located in the epidermis. (2) Cavernous lymphangiomas or macrocystic most common and composed of dilated lymphatic channels, characteristically invade surrounding tissues. (3)

Fig. 4 Histopathology showing vascular spaces filled with lymph with lymphoid cells in fibrous stroma

Fig. 5 IHT showing positive factor VIII related antigen in endothelium of vascular spaces

Cystic lymphangiomas (Cystic hygroma) large, macrocystic lymphangiomas filled with straw-colored, protein-rich fluid (4) Hemangiolymphangiomas- lymphangiomas with a vascular component. There may not be any essential difference between these types but their apparent difference is due to their site [3]. Microcystic lymphangiomas are composed of cysts, each of which measures less than 2 cm^3 in volume, macrocystic lymphangiomas contain cysts measuring more 2 cm^3 in volume and mixed lymphangiomas contain both microcystic and macrocystic components. Lymphangiomas may be described in stages, which vary by location and extent of disease in relation to hyoid bone. Stage 1: unilateral infrahyoid. Stage 2: unilateral suprahyoid. Stage 3: unilateral suprahyoid and infrahyoid. Stage 4: bilateral suprahyoid. Stage 5: bilateral suprahyoid and infrahyoid.

Three theories have been proposed to explain the origin of this abnormality. The first suggests that a blockage or arrest of normal growth of the primitive lymph channels occurs during embryogenesis, the second that the primitive lymphatic sac does not reach the venous system, while the third advances hypothesis that, during embryogenesis, lymphatic tissues lays in the wrong area [4].

Treatment includes aspiration, surgical excision, laser, radio frequency ablation and sclerotherapy. Surgical resection still remains the best for lymphangiomas. Spontaneous regression has been found to be rare. As the

lymphangiomas are nonencapsulated, complete removal is often associated with excessive removal of surrounding tissue. Usually it is surgical debulking which may need to be addressed again. Other treatment options, such as sclerotherapy have been proposed as an alternative to reduce the impact and complication of surgery. Various products, such as sodium morrhuate, dextrose, tetracycline, doxycycline, bleomycin, ethibloc(alcohol) and OK-432 (lyophilized incubation mixture of group A *Streptococcus pyogenes* of human origin) [5], have been used as sclerotherapeutic agents. Apart from OK-432, other agents were reported to cause perilesional fibrosis and thus to complicate eventual surgical excision. OK432 stimulates and increase the permeability of the endothelium, thus accelerating drainage of the lymphatic fluid and reduction in the size of the lymphangioma. Okazaki et al. [6] recommended intralesional OK432 injection therapy for single cystic and macro cystic and surgical excision after pretreatment with OK432 in microcystic and cavernous types.

In this case, we were able to excise the lesions from its origin under direct vision and cauterization of the base and hence no chance of injury to the surrounding organs using endoscopic approach.

We report this rare isolated case with rare site of origin and unusual age of presentation. It is possibly the first case of lymphangioma being reported in english literature.

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