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Research Article

Multi-disciplinary approach towards lymphangiomas of the head and neck in adults

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ABSTRACT

Background: Lymphangiomas are congenital malformations of lymphatic vessels or certain acquired factors resulting in lymphatic obstruction, lymph fluid retention, lymphangiectasia and proliferation. Two-thirds of all reported cases are present in the head and neck region. Majority of them are often present at birth and diagnosed mostly (90%) before the age of two years. These lesions are rare in adults but may occur as late as the fifth decade of life.
Methods: A prospective study was done on 60 cases of Lymphangiomas of the head and neck in adults. All the cases were subjected to a clinico-pathological & clinico-radiological evaluation involving the otorhinolaryngologists, cytopathologists, interventional radiologist & the patients. The multidisciplinary approach was discussed to outline the management of Lymphangiomas of the head and neck in adults. The clinical data was analyzed by using Statistical Package for Social Sciences (SPSS) version 11.
Results: In our prospective study of 60 adult patients having lymphangiomas of the head and neck region, the age of the patients was ranging from 18 to 53 years with an average mean age of 29.5 yrs. Females & males were almost equally affected as the sex ratio Female : Male was 1.06 : 1.
Conclusions: The choice of treatment of lymphangiomas should be individualized and based on factors such as etiology, age of onset, growth rate, type (capillary/cavernous/cystic), depth, location, potential morbidity & potential surgical complications. The different available treatment modalities can be outlined either as a single modality of treatment or as a combination of two or more modalities in a sequential manner.

Keywords: Lymphangioma, Head & neck lymphangioma, Percutaneous sclerotherapy, Adult lymphangioma

INTRODUCTION

Lymphangiomas are the result of aberrations in the development of the lymphatic channels that belong to a large spectrum of vascular malformations. They are most commonly located in the head and neck region, but can occur anywhere there are lymphatic vessels.[1] Lymphangiomas occur either due to congenital malformations of lymphatic vessels or certain acquired factors such as trauma or infection resulting in lymphatic obstruction, lymph fluid retention, lymphangiectasia and proliferation. The majority of Lymphangiomas arise from parts of lymph sacs that fail to establish connections with the main lymphatic or venous channels by the end of 9th week of gestation.[2] A small proportion appears to arise from localized lymphatic malformations or obstruction. Although benign, lymphangiomas frequently present surgical difficulties and challenges due to their propensity to infiltrate and extend around neighboring structures. Although the treatment of choice is complete surgical

removal, their infiltrating nature & difficulty in distinguishing involved vital structures of head and neck from adjacent normal tissues makes complete surgical resection difficult. Other modalities available include laser excision, sclerotherapy, radiotherapy, electro-coagulation, cryotherapy & embolization.[3] We share our experience of individualized, comprehensive & multi-disciplinary approach towards lymphangiomas of the head and neck in adults.

METHODS

A prospective study was done on 60 cases of Lymphangiomas of the head and neck in adults. All the cases were subjected a thorough clinical examination followed by F.N.A.C., ultrasound, color doppler, C.T. and MRI to diagnose lymphangiomas & to assess size, type, extent and proximity to major neck vessels, cranial nerves & brachial plexus. A clinico-pathological & clinico-radiological evaluation was done in all cases involving the otorhinolaryngologists, cytopathologists, Interventional radiologist & the patient. All patients were explained about the advantages & disadvantages of different modalities of treatment available. The multidisciplinary approach was followed to guide the patient to choose the management of lymphangiomas of the head and neck in adults. All patients were subjected to a suitable intervention according to the size, type, consistency of lymphangiomas and their vicinity to major vessels & important nerves in head & neck region. The CO2 LASER of wavelength 10,600 nm (far-infrared light) with 0.2 mm spot size was used with continuous output power of 1–6W. The laser probe was kept at a distance of 0.5–1 cm from the surface & the lesions were gradually cauterized from surface to deep layer. For percutaneous sclerotherapy Pingyangmycin (PYM), a single-component A5 of various components of Bleomycin was used.[4] Patients with extensive lesions were treated with percutaneous sclerotherapy every two weeks in 3 to 4 sessions All patients were followed up for six months for any recurrence. All those six patients who had recurrence after surgery were subjected to revision surgery, sclerotherapy or LASER.

Statistical analysis

The clinical data was analyzed by using Statistical Package for Social Sciences (SPSS) version 11 to assess the efficacy of multi-disciplinary approach towards Lymphangiomas of head & neck region in adults. Descriptive statistics was used to generate the results.

RESULTS

In our prospective study of 60 adult patients having Lymphangiomas of the head and neck region, the age of the patients was ranging from 18 to 53 years with an average mean age of 29.5 yrs. Females & males were almost equally affected as the sex ratio Female : Male was 1.06 : 1. Out of all potential locations in Head & neck region, the Lymphangiomas were more common (23%) in submandibular region than any other region (Figure 1). More than 2/3rd (46) of the lesions were found in above mylohyoid muscle & remaining (14) were below the mylohyoid muscle region. More than 85% of the patients were having the lesions since birth and the remaining patients were having a positive history suggestive of acquired etiology like trauma & chronic irritation (Figure 2). Based on morphology of the lesion , 30 patients were having cavernous Lymphangiomas, 24 patients with capillary lymphangioma & remaining 6 were having Cystic Lymphangiomas (Figure 3). More than 50% i.e. 36 patients were suitable for surgical intervention. Out of 36 patients recurrence was seen in six cases within six months & was treated with either revision surgery, LASER excision or sclerotherapy (Figure 4). Two patients, who had recurrence even after revision surgery responded nicely with sclerotherapy for the residual/recurrence of the lymphangioma. Small microcystic lesions on togue & cheek were excised with CO2 LASER.

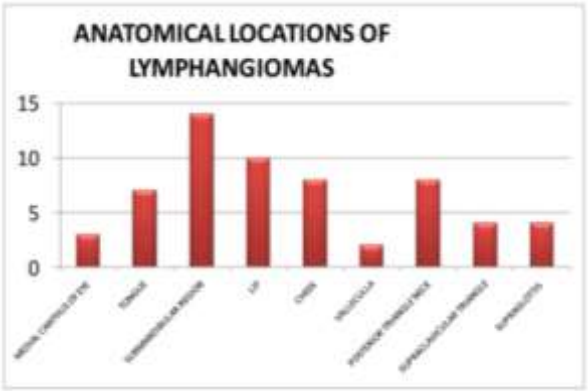

Figure 1: Distribution of lymphogiomas in head and neck region in adults.

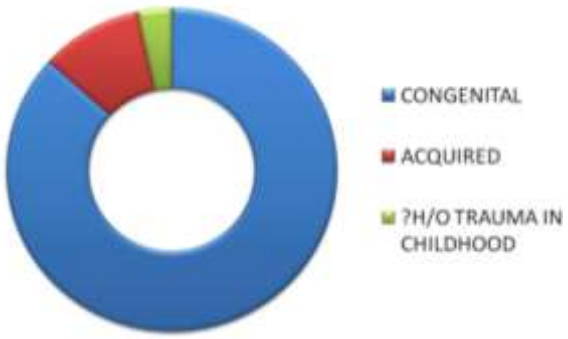

Figure 2: Etiology of lymphogiomas in head and neck region.

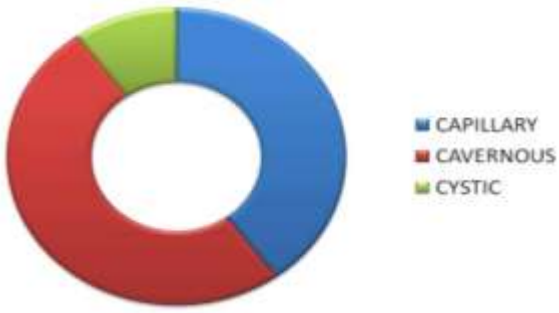

Figure 3: Types of lymphogiomas according to consistency.

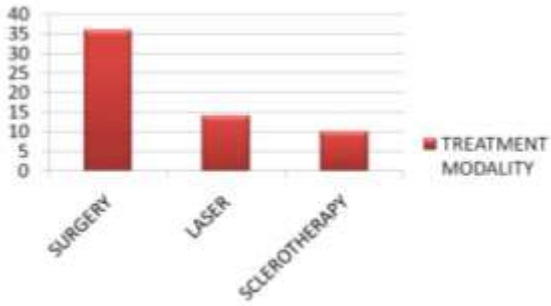

Figure 4: Primary treatment modality outlined for the patients.

DISCUSSION

Lymphangiomas are one of the most common benign lesions, consisting of localized areas of abnormal development of the lymphatic system. Most of the patients present due to cosmetic reasons, however dysphagia, pain in the swelling, sudden rapid enlargement in size, airway compromise can also be the mode of presentation.

Lesions in oral mucosa commonly present as microcystic, isolated or multiple sporadic, pink to dark red soft round nodules or punctuate lesions. The deep seated lesions may present with macroglossia, macrocheilia and may be associated with mandibular hypertrophy. Ultrasound, C.T. and MRI are important adjunctive diagnostic approaches to identify the location, size and extent of the lesions. Lymphatic malformations (LMs) are low-flow vascular anomalies, often appearing as multiple cystic spaces. The signal intensity is intermediate on T1-weighted image; hyperintense on T2-weighted image with no enhancement after contrast injection.[5] LMs can be classified according to their size, content, consistency and location. The Lymphangiomas can be macrocystic, microcystic & mixed (macro- and microcystic) depending upon their size, such as macrocystic lesions in the neck with microcystic lesions in the tongue or cheek. The LMs can be divided into serous and chylous type based on the contents in the cysts. Based on morphologic and histological characteristics, Lymphangiomas can also be classified in to the capillary, the cavernous, and the cystic lymphangioma.[6] Lymphangiomas in Head and Neck region are also classified into type I lymphangioma, which is located below the level of the mylohyoid muscle, and type II lymphangioma, found above the level of the mylohyoid muscle involving the cheek, oral cavity, lip, and tongue.[7] De Serres et al, proposed a staging system based on the location and extent of the lesions: stage I is unilateral infrahyoid, stage II is unilateral suprahyoid, stage III is unilateral infrahyoid and suprahyoid, stage IV is bilateral infrahyoid, stage V is bilateral infrahyoid and suprahyoid.[8] This type of staging system does offer some importance in prognosis of the Lymphangiomas of Head & Neck region.

The localized microcystic lesions can be resected completely either by laser or electro-coagulation but impossible for extensive and diffuse ones. Surgical excision has been the cornerstone of treatment for Lymphangiomas of the head and neck. However, complete surgical resection is a viable option only when the lesion is well defined & excision of lesion will not lead to obvious disfigurement or damage to vital structures such as orbit (Figure 5), cranial nerves, sympathetic chain or major neck vessels. Involvement of certain areas like angle of mandible, angle of mouth and tongue may lead to severe cosmetic deformity and morbidity when excised and hence alternative options should be considered. Watson and McCarthy advocated sclerotherapy as early as in 1964.[9] Because the endothelial lining of lymphangiomas seems to be vulnerable to infections and chemical irritants, and spontaneous infection of lymphangiomas can lead to total regression of the lesion, sclerotherapy was initiated in an attempt to exploit this phenomenon.[10] In recent years, many centers have switched to using sclerotherapy as a primary treatment, followed by surgery if needed, rather than using sclerotherapy as rescue treatment. In our study, two patients who had extensive lesions were managed by percutaneous sclerotherapy as a primary intervention followed by surgical excision (Figure 6). Macrocystic, well defined lesions in posterior triangle of neck (Figure 7) can be successfully excised without any obvious morbidity.

Figure 5: Surgical excision of cystic lymphogiomas for right upper eye lid.

Figure 6: Percutaneous sclerotherapy results in 3 sessions.

Figure 7: Macrocystic Lymphangioma in left posterior triangle of neck.

CONCLUSION

The various classifications & staging system of lymphangiomas offer little help in their management. Percutaneous sclerotherapy is a safe & effective alternative to surgical intervention particularly on imaging studies, when the lymphangioma lesion is in the vicinity of important major vessels & nerves in the neck. Sclerotherapy can be used as the primary treatment of choice or postoperative adjunctive treatment in case of extensive lesions. The choice of treatment of lymphangiomas should be individualized and based on factors such as etiology, age of onset, growth rate, type (capillary/cavernous/cystic), depth, location, potential morbidity, potential surgical complications, available technology & expertise. The different available treatment modalities can be outlined either as a single modality of treatment or as a combination of two or more modalities in a sequential manner. Further studies with a larger sample size are required to evaluate advantages, limitations & long-term results of different modalities for management of lymphangiomas of the head and neck in adults.

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Ethical approval: The study was approved by the institutional ethics committee

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