Lymphatic malformations: A dilemma in diagnosis and management

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Abstract
Lymphatic malformations of the head and neck, also known as lymphangiomas or cystic hygromas, are a diverse group of lesions. Lymphangiomas represent benign hamartomatous tumors of lymphatic vessels with a marked predilection for the head, neck and oral cavity. These lesions, like most of the swellings occurring in the neck, frequently pose a dilemma in diagnosis and treatment. This is a case report of a lymphatic malformation which presented as a painless mass in the lower border of mandible.

Keywords: Hamartoma, neck swellings, neck tumors

Introduction
Lymphatic malformations are congenital malformations of the lymphatic system. They consist of channels and cystic spaces of varying size and result in the accumulation of fluid, often beneath the skin. Embryologically, they are thought to originate from the sequestration of lymphatic tissue during the development of lymphaticovenous sacs, which then fail to communicate with the remainder of the lymphatic or venous system. Later on the sequestered lymphatic tissues dilate, which results in the cystic morphology of the lesions.[¹]

Two-thirds of all the reported cases are found in the head and neck. Lymphatic malformations can be congenital or acquired. Acquired lesions generally arise from obstruction of the lymphatic system.[²] Trauma or infection is generally considered responsible for their pathogenesis.[³]

Indications for treatment include recurrent infection, cosmetic disfigurement and compression of local structures such as the airway, blood vessels, or upper gastrointestinal tract.[⁴] This is a case report of a lymphangioma which presented as a painless mass in the lower border of mandible.

Case Report
The present case report is about a 25-year-old female patient who reported to the Department of Oral and Maxillofacial surgery with swelling in the right lower third of face below the lower border of mandible since 6 months [Figure 1]. Recently, the swelling had suddenly increased in size with severe pain from the past 3 days. On examination, it was associated with infected molar, which was assumed to be the etiology of the swelling, for which extraction was carried out. After extraction, the pain subsided, however, the swelling was persistent and had not completely resolved even after a month. During the follow-up visit, the lesion was soft, non-tender and compressible and it measured 5 × 4 cm in size approximately. The skin over the swelling had normal texture and consistency. No bruits were heard on auscultation. The patient informed us that the swelling intermittently increased and decreased in size and has reached the present size. The growth although painless was cosmetically disfiguring.

On investigation, needle aspiration yielded a blood-tinged clear fluid which resembled lymphatic fluid, which was examined in light microscopy. After a routine blood investigations, the patient was advised an orthopantomograph which failed to reveal any bony changes. Later on, the patient was advised to undergo an ultrasonographic scan which revealed a multicystic lesion with internal septations. A computed tomography (CT) scan with contrast was carried out, which revealed a multicystic, homogeneous, non-invasive lesion with low attenuation.

The lesion was provisionally diagnosed as a lymphatic malformation and incisional biopsy was planned. A 5-cm
extraoral incision was placed in the first cervical crease. Blunt and sharp dissection was carried out to free the lesion from the overlying platysma muscle. Once the lesion was identified, aspiration of some content of the cyst was carried out to reduce the size of the lesion. Blunt dissection was carried out to protect vital structures of the neck. The medial part of the lesion extended up to the pharyngeal wall as per ultrasound and blunt dissection was carried out until the lesion was free from surrounding tissue [Figure 2]. The lesion was excised fully and the floor was examined for any perforations of the pharyngeal wall [Figure 3]. The surgical wound was cleaned and debrided and sutured in layers. After the surgery, the patient recovered well without any complications.

The lesion was sent for histopathologic examination which revealed large dilated lymphatic vessels filled with light eosinophilic proteinaceous fluid which confirmed the final diagnosis as lymphangioma. Post-operative follow-up after 1 month showed a good cosmetic healing of the surgical scar [Figure 4].

**Discussion**

The variety of anatomic structures within or bordering the parapharyngeal space accounts for many types of lesions that involve this area.\(^5,6\) Lymphangiomas represent hamartomas of malformed lymphatics. They are sometimes termed lymphangiectasias because they are actually cystic dilations of malformed lymphatic channels that fail to communicate with or drain into other lymphatic channels or veins and therefore will collect lymph. Lymphatic malformations range in size from small, asymptomatic masses to massive, disfiguring lesions.\(^7\) Nearly half of all lymphatic malformations are diagnosed in the head and neck regions and frequently pose treatment dilemmas for the surgeon.

Lymphatic malformations are either congenital or acquired. Most of the lesions are congenital and develop in infants and children below 2 years of age. Commonly accepted theories for the origin of congenital lymphangiomas include failure of lymphatics to connect with the venous system, anomalous budding of the lymphatic structures and abnormal
sequestration of lymphatic tissue. Recently, the vascular endothelial growth factor C gene has been found to be a critical factor for normal lymphatic development. Trauma or respiratory infections are generally considered responsible for their pathogenesis in acquired type of lymphatic malformations due to obstruction of lymphatic system.

Three types of lymphangiomas have been described: The superficial multicystic type; the deep cavernous type and the cystic hygroma. However, these actually represent a single type of defect in lymphatic development manifesting different degrees of severity. In general speaking, lymphangiomas are less common than hemangiomas. Most of those occurring in the head and neck area (50-65%) are present at birth, while 90% are clinically apparent by age 3 years; the majority of these are the deep cavernous type.

The most common presentation is that of a painless soft mass that gradually enlarges and then remains static over a long period. Although occasional enlargements and shrinkages occur, a residual mass remains. Table 1 discusses the anatomic classification of lymphatic malformations.

Ultrasound usually shows a multicystic lesion with internal septations and no blood flow is detected on Color-Flow Doppler. The latter is particularly helpful in detecting the different vascular characteristics of mixed vascular malformations and it also differentiates hemangiomas from vascular malformations.

CT and magnetic resonance imaging (MRI) delineate extension and show relation with other anatomical structures better than ultrasound. CT shows multicyrstic, homogeneous, non-invasive lesions with low attenuation. MRI is particularly useful for malformations that involve muscle and whilst T1-weighted images are helpful in locating lesions, a T2-weighted scan can readily distinguish them from normal muscle. Cross-sectional imaging is also helpful in measuring how much of the lesion is close to the great neck vessels, particularly when surgery is being planned.

There are a number of contemporary treatments for lymphatic malformations, which range from simple, relatively non-invasive methods to advanced surgery with adjuvant techniques. The management of head and neck lymphatic malformations is challenging due to the close association with the adjacent vital structures and poor demarcation.

Current treatment methods of lymphatic malformations include surgery, sclerotherapy and laser therapy, or a combination thereof. Non-surgical conservative treatments include radiotherapy, electrocoagulation, cryotherapy, ligation and embolization.

Localized lymphatic malformations can be treated with surgery, laser therapy or sclerotherapy, usually with excellent results. Excision can be considered either due to the potential complications related to the lesion or for esthetic reasons. Lesions can become infected and in some cases may become life-threatening if the airway is compromised or if there are difficulties feeding young children. For diffuse lesions or lesions with cosmetic and functional problems, an individualized treatment plan should be made based on the status of the patient and technology and expertise available.

Although lymphatic malformations are benign lesions, spontaneous regression is rarely seen and only 12.5-15.0% respectively. Therefore, observational monitoring may be appropriate in some cases. However frequently, a regression is usually followed by recurrence in most cases. Surgery remains the treatment of choice in well-localized lesions with least recurrence rate when complete excision is possible.

It has to be remembered that complete excision provides the least recurrence rate, but surgery is not feasible in all cases. In this particular case, the persistent inflammation of the lymphangiomata, the wall of the lesion had thickened, which facilitated easier dissection and complete excision was possible.

**Conclusion**

Management of these diverse groups of lesions is complex and multidisciplinary and depends on expertise and experience. Whilst some lesions may resolve spontaneously (for this reason treatment should ideally be delayed in young children), the mainstay of management is to eradicate the lesion whilst causing minimal damage to adjacent structures. Various treatment modalities such as surgical excision, laser therapy, sclerotherapy or combination therapy have been proposed in management of such lesions.

Complete excision, although not feasible in all lesions, has shown to result in the least recurrence rate among all treatment modalities and remains the mainstay in treating localized lymphatic malformations.

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**Table 1: Classification of lymphatic malformations as suggested by De Serres et al.**

| Stage | Anatomic location |
|-------|-------------------|
| I     | Unilateral infrahyoid lesion |
| II    | Unilateral suprathyoid lesion |
| III   | Unilateral lesion extending both above and below the hyoid |
| IV    | Bilateral infrahyoid lesion |
| V     | Bilateral lesion extending both above and below the hyoid |
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