

Facial Lymphangioma with Orbital Involvement

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Abstract

A 4 yr old male child came to our OPD with swelling over the left side of the face and forehead, gradually increasing in size, showing rapid growth over the past few days. There was h/o fever and difficulty in seeing. On examination, there was a 10 x 10 cm swelling involving the left face, left side of the cheek, left eye and forehead. The swelling was soft and compressible and no lymphnode was palpable. CT scan was s/o venous lymphangiomatous malformation. Histopathology confirmed it to be a lymphangiomatous tissue. Intraocular examination was normal.

Introduction

Lymphangiomas are uncommon, hamartomatous, congenital malformations of the lymphatic system that involve the skin and subcutaneous tissues. The classification of lymphangiomas lacks a standard clear definition and universal application, in part because of the nature of lymphangiomas, which represent a clinicopathologic continuum. The classification most frequently used divides these lesions into 2 major groups based on the depth and the size of these abnormal lymph vessels. The superficial vesicles are called lymphangioma circumscriptum. The more deep-seated group includes cavernous lymphangioma and cystic hygroma. Many categorize cystic hygroma as a variant of cavernous lymphangioma.

Lymphangiomas can occur anywhere in the skin and the mucous membranes. The most common sites are the head and the neck, followed by the proximal extremities, the buttocks and the trunk. However, they sometimes can be found in the intestines, the pancreas, and the mesentery. Deeper cystic

lesions usually occur in areas of loose and areolar tissue, typically the neck, the axilla, and the groin. Their skin involvement ranges from small, well-demarcated areas to large, diffuse regions with unclear borders.

Lymphangioma circumscriptum, the common form of cutaneous lymphangioma, is characterized by persistent, multiple clusters of translucent vesicles that usually contain clear lymph fluid (often compared to frog spawn). These vesicles represent superficial saccular dilations from underlying lymphatic vessels that occupy the papilla and push upward against the overlying epidermis. Each skin lesion may range from a minute vesicle to a small bulla-sized lesion. These vesicles can be clear or vary from pink to dark red because of serosanguineous fluid and haemorrhage. These vesicles often are associated with verrucous changes, which give them a warty appearance.

In the case of lymphangioma circumscriptum, the underlying lesions constitute abnormal dilated lymph vessels involving the upper part of the dermis. The sites of predilection are the proximal extremities, trunk, axilla, and oral cavity, especially the tongue. Involvement in other areas, such as the scrotum, is not uncommon.

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Lymphangioma *circumscriptum* has a high recurrence rate after excision because of its deep component.

Cavernous lymphangioma are also uncommon and usually arise during infancy. The most common sites are the head and neck areas and, less frequently, the extremities. These lesions are seated deep in the dermis, forming a painless swelling or thickening of the skin, mucous membranes, and subcutaneous tissue. Unlike lymphangioma *circumscriptum*, the overlying skin usually is uninvolved. Occasionally, patients report pain when the involved area is pressed. The affected area may be 1 cm, it may be as large as several centimeters in diameter, or it may involve an entire extremity. Upon examination and palpation, lipomas or cysts can be mistaken for these lesions. Lymphangioma *circumscriptum* can occur in conjunction with cavernous lymphangioma and cystic hygroma.

Some authors categorize cystic hygroma or cystic lymphangioma as an independent entity. Many authors agree that cystic hygroma is a form of cavernous lymphangioma in which the degree of involvement and character is determined by its location. These congenital lesions are deeply seated in areas of areolar or loose connective tissue. They appear early in life as large soft-tissue masses, usually on the axilla, neck, or groin. These lesions are soft, vary in size and shape, and tend to grow extensively if not surgically excised. Typical lesions are multilocular cysts filled with clear or yellow lymph fluid. Usually, cystic hygroma is diagnosed clinically with its large size, location, and translucence.

Case History

A 4 yr old male child came to our OPD with swelling over the left side of the face and forehead, gradually increasing in size, showing rapid growth over the past

few days. There was h/o fever and difficulty in seeing. On examination, there was a 10 x 10 cm swelling involving the left face, left side of the cheek, left eye and forehead (Fig. 1). The swelling was soft and compressible and no lymphnode was palpable. CT scan was suggestive of venous lymphangiomatous malformation. Histopathology confirmed it to be a lymphangiomatous tissue. Intraocular examination was normal (Fig. 2).

Pathophysiology

In 1976, Whimster studied the pathogenesis of lymphangioma *circumscriptum*. According to Whimster, the basic pathologic process is the collection of lymphatic cisterns in the deep subcutaneous plane. These cisterns are separated from the normal network of lymph vessels, but they communicate with the superficial lymph vesicles through vertical, dilated lymph channels.

Whimster postulated that these cisterns might arise from a primitive lymph sac that fails to connect with the rest of the lymphatic system during its embryonic development. A thick coat of muscle fibres that cause



Fig. 1



Fig. 2

rhythmic contractions line these sequestered primitive sacs. Rhythmic contractions increase the intramural pressure, causing dilated channels to protrude from the walls of the cisterns toward the skin. He suggested that the vesicles seen in lymphangioma circumscriptum are outpouchings of these dilated projecting vessels.

Whimster's observations are supported by those of lymphangiographic and radiographic studies. These studies revealed that large multilobulated cisterns extend deep in the dermis and laterally beyond the obvious clinical lesions. These deep lymphangiomas show no evidence of communication with the adjacent normal lymphatics. The cause for the failure of these primitive lymph sacs to connect to the rest of the lymphatic system is not known.

Frequency

United States

Lymphangiomas are rare. They account for 4% of all vascular tumours and approximately 25% of all benign vascular tumours in children.

Mortality/Morbidity

Rarely do cutaneous lymphangiomas interfere with the well-being of patients. Patients are expected to live a full healthy life, and they usually seek medical intervention because of cosmetic reason.

- Lymphangiomas represent hamartomatous malformations with no risk of malignant transformation.
- In the case of cystic hygroma, total surgical excision is appropriate to prevent complications such as respiratory compromise, aspiration, and infections in critical areas, such as the neck.
- Lymphangiomas have a strong tendency for local recurrence unless they are completely excised. Recurrent episodes of cellulitis and minor bleeding are not uncommon.

Race

No racial predominance is reported.

Sex

Equal sex incidences are reported in most studies. Some groups have reported that lymphangioma circumscriptum is more common in females than in males, while others report a 3:1 male-to-female ratio.

Age

Lymphangioma can become evident at any age, but the greatest incidence occurs at birth or early in

life. About 50% of lymphangiomas are seen at birth, and most lymphangiomas are evident by the time the patient is aged 5 years.

History

- Lymphangioma circumscriptum
 - The typical history involves a small number of vesicles on the skin at birth or shortly after. In subsequent years, they tend to increase in number, and the area of skin involved continues to expand. Vesicles or other skin abnormalities may not be noticed until several years after birth.
 - Usually, lesions are asymptomatic, but, occasionally, patients may have spontaneous episodes of minor bleeding and copious drainage of clear fluid from ruptured vesicles.
- Cavernous lymphangioma
 - Typically, during infancy, a solitary rubbery nodule with no skin changes become evident in a single location, such as the face, trunk, or extremity. These lesions often have a fast growth phase similar to that of raised haemangiomas.
 - No family history of prior lymphangiomas is described.
- Cystic hygroma
 - Soon after birth, the infant is noted to have a deep subcutaneous cystic swelling, usually in the axilla, base of the neck, or groin. If these lesions are drained, they tend to fill up rapidly with lymph fluid.
 - The lesions tend to grow and increase to a large size if they are not completely excised at surgery.

Physical

- Lymphangioma circumscriptum
 - Lymphangioma circumscriptum involves small clusters of vesicles measuring about 2-4 mm. These clear vesicles can vary from pink to red to black secondary to haemorrhage.
 - The lesions can have a warty appearance on their surface; as a result, these lesions are often confused with warts.
- Cavernous lymphangioma
 - Typically, cavernous lymphangiomas appear as subcutaneous nodules with a rubbery consistency. They may have large dimensions.
 - The overlying skin has no lesions or changes.
 - The area of involvement varies, ranging from lesions smaller than 1 cm in diameter to larger lesions that involve an entire limb.

- Cystic hygroma
 - Cystic hygromas are usually larger than cavernous lymphangiomas, and they more commonly occur in the neck and parotid area.
 - Often, deep cavernous lymphangiomas are not evident on superficial examination, but cystic hygromas are detected with ease because of their size and location. These large cystic lesions are soft and translucent.

Other Problems to be Considered

Other cysts and soft subcutaneous masses should be considered.

Occasionally, acquired lymphangioma circumscriptum of the vulva may mimic genital warts.

Imaging Studies

- MRI can define the degree of involvement and the entire anatomy of the lesion
- MRI can help prevent unnecessary extensive, incomplete surgical resection, because of the association with a high recurrence rate. (Figs. 3 and 4).

Other Tests

- Immunohistochemical study is useful in differentiating lymphangiomas from haemangiomas in difficult cases.
- Test results with factor VIII-related antigen are positive for haemangiomas but negative or weakly positive in the endothelium of

lymphangiomas.

- Immunohistochemical studies for laminin show the typical multilayered basal lamina of normal blood vessels and the discontinuous basal lamina in lymphangiomas.

Procedures

Biopsy: The diagnosis of lymphangiomas is based mainly on the clinical history and findings from physical examination and conventional light microscopy.

Histologic Findings

Microscopically, the vesicles in lymphangioma circumscriptum are greatly dilated lymph channels that cause the papillary dermis to expand. They may be associated with acanthosis and hyperkeratosis. These channels are numerous in the upper dermis and often extend to the subcutis. These deeper vessels seem to have a large caliber, and they often have a thick wall that contains smooth muscle. The lumen is filled with lymphatic fluid, but it often contains red blood cells, lymphocytes, macrophages, and neutrophils. These channels are lined by flat endothelial cells, which stain positive for Ulex europaeus agglutinin-I. The interstitium often has numerous lymphoid cells and shows evidence of fibroplasia.

Nodules in cavernous lymphangioma are characterized by large, irregular channels in the reticular dermis and subcutaneous tissue that are lined by a single layer of endothelial cells. An

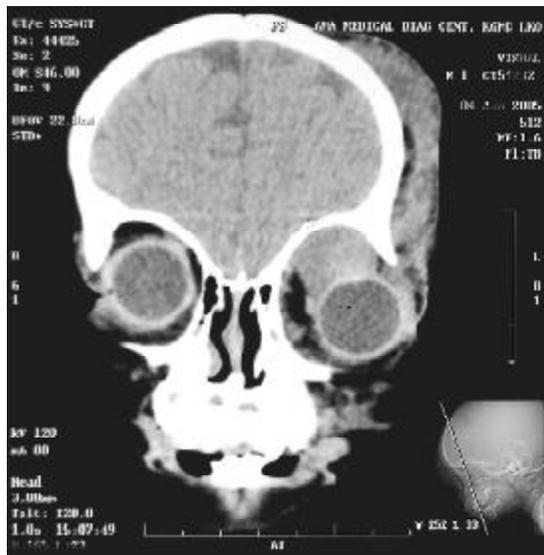


Fig. 3

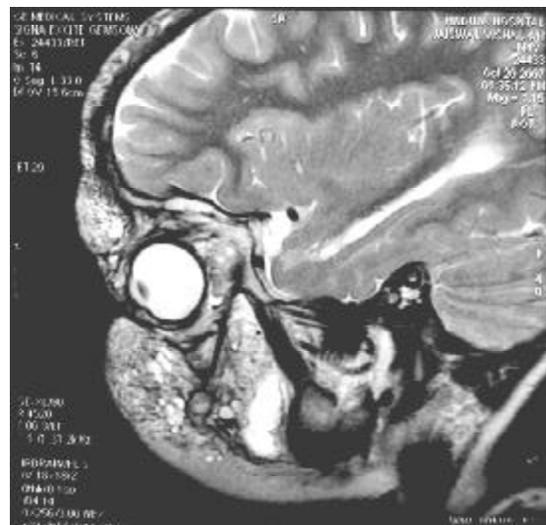


Fig. 4

incomplete layer of smooth muscle often lines the walls of these malformed channels. The surrounding stroma consists of loose or fibrotic connective tissue with a number of inflammatory cells. These tumours often penetrate muscle.

Cystic hygroma is indistinguishable from cavernous lymphangiomas on histology.

Medical Care

No proven medical care for lymphangiomas exists. This condition is not responsive to radiation therapy or steroids. Antibiotics are given for secondary cellulitis.

Surgical Care

The preferred treatment for lymphangiomas is complete surgical excision.

- Local recurrences are common in lymphangiomas.
- On the basis of the Whimster hypothesis, the large subcutaneous cisterns should be removed to prevent the lesion from resurfacing.
- Adequate excision of lymphangiomas can be difficult and, at times, unfeasible. This problem is the main reason for the high recurrence rate.
- Tumours that are confined to the superficial dermis are more amenable to surgical excision, which is associated with a high rate of success.
- The use of other treatment modalities have been advocated; these include cryotherapy, sclerotherapy, and cautery.
- Vaporization with a carbon dioxide laser has been tried with good results.
- A new therapeutic option for lymphangioma circumscriptum is 23.4% hypertonic saline sclerotherapy.
- The use of intralesional OK432 (Picibanil) is a new and effective treatment for macrocystic lesions, but the response of microcystic or cavernous lesions to OK-432 has been disappointing and surgery remains the most effective treatment for these microcystic and cavernous lesions.

Further Inpatient Care

In the case of lymphangioma circumscriptum, severe recurrent cellulitis may warrant inpatient care at times, especially in patients who are immunocompromised.

Further Outpatient Care

Patients should be monitored and treated for

cellulitis, especially those with ruptured vesicles, which provide a portal of entry for infection.

Regular skin examination should be included in the follow-up treatment to evaluate recurrence and the response to treatment.

Complications

- Lymphangioma circumscriptum may occur.
 - This condition is associated with minor bleeding, recurrent cellulitis, and lymph fluid leakage.
 - Two cases of lymphangiosarcoma arising from lymphangioma circumscriptum have been reported. However, in both of the patients, the preexisting lesion was exposed to extensive X-ray therapy. Therefore, radiation therapy should be avoided in lymphangiomas.
- In cystic hygroma, large cysts can cause dysphagia, respiratory problems, and serious infection, if they involve the neck.

Prognosis

Lymphangiomas are benign hamartomatous malformations instead of true neoplasms.

The prognosis is excellent.

Patient Education

- Patients should receive reassurance.
- Lymphangiomas represent benign lymphatic malformations and not premalignant lesions.
- Patients should be aware of the risk of recurrence.

Medical/Legal Pitfalls

Failure to evaluate for congenital disorders, such as Turner syndrome and Down syndrome, in patients with cystic hygroma

Special Concerns

- Cystic hygroma can be associated with the following congenital disorders:
 - Turner syndrome
 - Chromosomal aneuploidy
 - Hydrops foetalis
 - Down syndrome and other trisomy disorders
 - Foetal alcohol syndrome
 - Noonan syndrome
 - Several other congenital syndromes
- Patients with cystic hygroma should undergo cytogenetic analysis for chromosomal aneuploidy.
- Parents should receive genetic counselling because aneuploidic conditions can recur in subsequent pregnancies.