Case Report

Lymphangioma CircumscriptumA Rare Form Of Lymphangiomas

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Abstract:
Superficial lymphangiomas are either cavernous type, diffuse or circumscribed (circumscriptum). Lymphangima circumscriptum (LC) is a rare benign condition characterized by the appearance of persistent clusters in the skin and subcutaneous tissue which may contain fluctuant cysts[1]. The first reported description ofLC was done by Fox and Fox[2] in1878. They named it as lymphangiectodes[2]. The term LC is first used by Morris[3] in1889. Complete surgical excision is the main treatment. Here we are reporting a case of lymphangioma circumscritum of the thigh in a six-year old girl.

Lymphangiomas are congenital malformations of the lymphatic system that may involve skin and subcutaneous tissue. They are hamartomatous malformations which account for 25% of benign lympho-vascular tumours in children[4]. They are not familial[5].Lymphangioma circumscriptum, cavernous lymphangiomas, and cystic hygromas are the common forms of this condition.LC is a lesion that leads to superficial dilations of lymphatic vessels, which communicates with deeper subcutaneous lymphatic cisterns via small channels. These abnormal malformations are not connected to normal lymphatic[6].Vesicles of LC discharge clear fluid and are prone to super-infection. They can appear over any part of the body and may have a verrucous appearance[6].LC may look like a number of disorders including herpes zoster, viral warts and molluscum contagiosum[7,8].
Surgical excision is the cornerstone in treatment but recurrence is common, with a reported[5,9] first-time cure rate of 75%.It is recommended to surgically excise to the level of the deep fascia and obtain clear margins on frozen section to avoid recurrence[10].

Case report:

A six-year old girl presented with two swellings in the medial aspect of the right thigh for one year. One swelling was above the other. The upper one was excised but soon recurred. Physical examination revealed a residual soft tissue mass with unhealed part of the wound and multiple clusters of vesicles which discharge colorless smelly fluid.

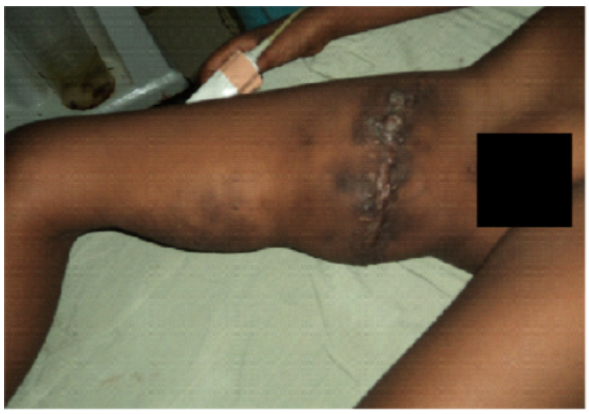

Figure1: Skin appearance before excision.

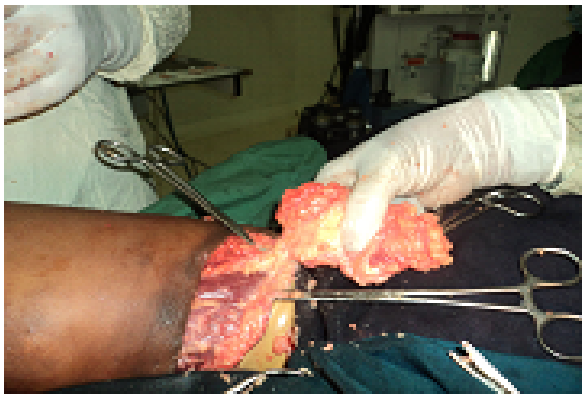

Figure 2: Specimen during excision.

There was no other swelling in any other part of the body. The lower swelling which was not operated before was 12x9cm.The upper one which was operated was about 18x10cm. Complete excision of the recurrent mass was done and sent for histopathology. Figures1,2

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and 3 show intra operative views. Histopathology revealed Lymphangioma circumscreptum as in figures 4 and 5. Follow up at three months revealed no recurrence of the lesion.

Discussion:

Originally LC was diagnosed as a soft- tissue mass. LC the skin and subcutaneous tissue is uncommon. The dilated vessels cause expansion of the papillary dermis. There is some acanthosis and hyperkeratosis of the overlying epidermis. The lymph fluid within the vesicles may contain erythrocytes, lymphocytes, macrophages, and/or neutrophils. It is a benign disorder affecting lymphatic channels. Its diffuse and cavernous subtypes are inter-related, and may occur in association with each other[14] since the basic pathogenesis is almost the same in all types.

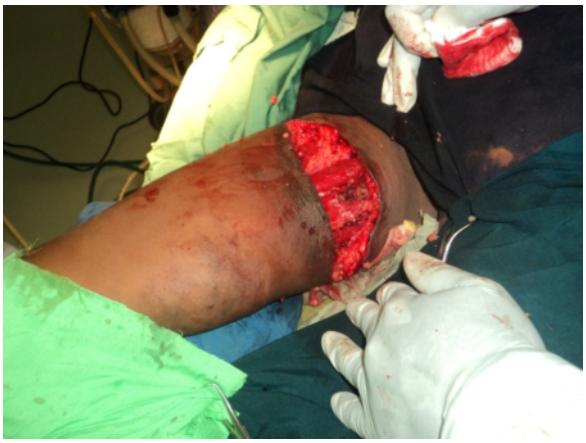

Figure 3: Look at the clean uncut tissue and muscle after excision

In LC, there is dilated muscle coated lymphatic cistern in the subcutaneous plane, communicating with large dermal lymphatics upwards, and finally erupting as superficial vesicles on the surface of the skin as blow-out phenomenon. There may be one or more abnormal lymphatic cistern and vessels.
Lymphatics form closed system, neither communicate with each other nor with the main normal lymphatic channels[11]. The sequestration of cisterns is considered as developmental defects. It is reported after radiation therapy for malignant tumours[12].
Lymphoedema is absent in the affected area, the reason being that the presence of normal lymphatic drainage. Also LC can be divided into smaller lesions (less than seven

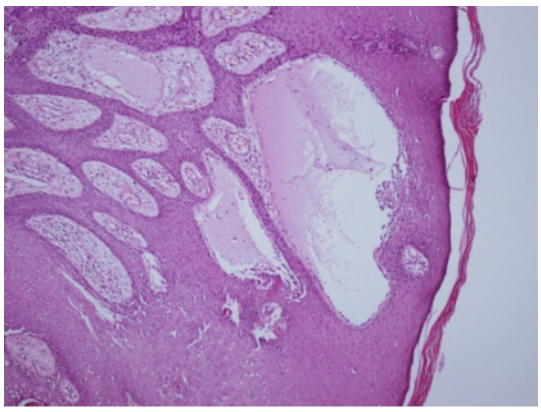

Figure 4: histology slide that shows dilated lymph vessels in the upper dermis that extend into the subcutis.

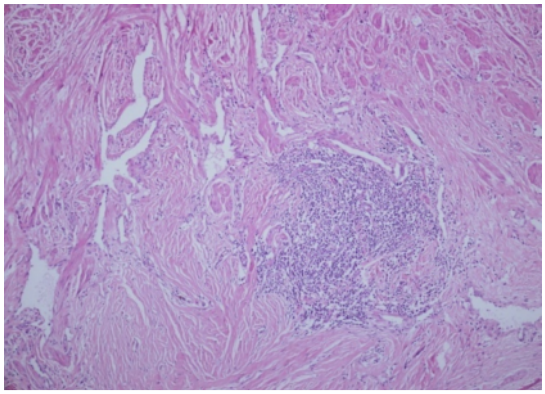

Figure 5: Histology slide, high power.

centimetres) and extensive lesions (more than that)[10]. The incidence of LC is higher in the first and second decade of life has preponderance females[10,13]. The proximal part of limbs, axillary folds, adjacent chest wall and flanks are the common sites[13].
The classical subtype is characterized by extensive swelling with thickened, hypertrophied, hyperpigmented, lichenified skin. Clusters of vesicles may be scattered over the involved skin. These vesicles may vary in appearance from pale white to reddish brown. Some of these vesicles may become oozing[13,17]. Lymphangiography was the procedure of choice to map the involved Lymphatics[11,13]. Magnetic resonance imaging (MRI) is the investigation of choice to see the extent of disease[13].
The diagnosis is made on histopathology. Surgical excision is still the only option of

treatment for extensive/classical subtype despite chances of recurrence[11]. Other modalities are intra-lesional sclerotherapy using doxycycline, Picibanil (OK-432)[14], or vaporization by carbon dioxide laser[15]. The principle of surgery is based on the excision of all sequestrated lymphatic cisterns in the subcutaneous plane, since they are the main culprits. The diseased subcutaneous tissue is recognized by the presence of cysts, fibrous strands, and wetness in the deep fascia[11,10]. The prognosis is excellent as it is a non-neoplastic condition. However, squamous cell carcinoma has been reported in an existing LC[16].

Conclusion:

Our patient was a six-year old girl, suffering from LC in the inner aspect of the proximal part of right thigh. Cure was obtained in the second surgical excision, with an acceptable cosmetic result. To minimize the chances of recurrence, wide local excision of diseased subcutaneous tissue is mandatory rather than wide excision of skin.

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