Extensive and invasive lymphangioma circumscriptum in a young female: A rare case report and review of the literature

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ABSTRACT

Lymphatic malformations, or lymphangiomas, are benign proliferations of lymphatics, classified as superficial lymphangioma circumscriptum (LC) and deep (cavernous lymphangioma). LC, as the name suggests, is a well-circumscribed lesion but, rarely, extensive and invasive forms can also be seen. Surgical excision is the main modality of treatment. We report a case of extensive and invasive type of LC where surgical excision is difficult and recurrence is the rule.

Key words: Extensive, lymphangioma circumscriptum, malformed lymphatics

INTRODUCTION

Lymphangiomas are not true benign tumors because they are usually associated with lymphatic malformations. They account for 4% of all vascular malformation and 26% of all benign vascular tumors.[1] LC is classified as: (1) lymphangioma circumscriptum (LC), (2) cavernous lymphangioma, (3) cystic hygroma and (4) benign lymphangioendothelioma. LC is a benign hamartomatous lymphatic malformation of the skin and subcutaneous tissue characterized by thin-walled vesicles mimicking frog spawn. The content of the vesicles may be clear or blood-tinged fluid. The term LC was coined by Morris.[2] Treatment is by surgical excision and recurrences are very common. Other treatment modalities with success are intra lesional sclerotherapy using doxycycline or picibanil (OK-432) and vaporization by carbon dioxide laser. We report a case of extensive and invasive type of LC in a young female where surgical excision is difficult and complete cure is highly impossible.

CASE REPORT

A 21-year-old female presented to the dermatology out-patient department with multiple hemorrhagic vesicles over the right side of the abdomen, buttock and thighs since birth. Immediately after birth, bluish-colored red plaque was noticed by her mother in the anterior aspect of the right thigh. Gradually, over the next 6 months, multiple water-filled lesions appeared over the whole of the right buttock and right side of the lower abdomen. It was associated with recurrent episodes of pain and fever. At 1 year of age, she was taken to a plastic surgeon and the whole lesion was excised under general anesthesia. After 1 year, the lesions recurred with greater involvement of the thighs, buttocks and right lower abdomen. At 5 years of age, magnetic resonance imaging (MRI) scan showed extensive and deep LC with involvement of deep fascia, muscles of abdomen and buttocks and large vessels, including right iliac and right femoral arteries. The new fluid-filled vesicles appeared over the thighs and legs as well. There was history of episodes of bleeding from the lesion associated with pain. After the MRI scan, she was advised for rest and non-surgical management of the LC as it was deep and extensive.

Examination revealed a 20 cm X 15 cm-sized large area of hemorrhagic fluid-filled vesicles of varying size from 3 mm to 1 cm over the right lower abdomen and buttocks and extending down toward the right thigh [Figures 1 and 2]. Few scattered clusters of vesicles were also seen over the posterior aspect of the right thigh [Figure 3]. The diameter of the right thigh was 6 cm more than that of the left one. Areas of...
Figure 1: Lymphangioma circumscriptum: Clusters of vesicles of varying size with clear to hemorrhagic fluid are seen over the right lower abdomen and buttock

Figure 2: Lymphangioma circumscriptum: Close up view of the tumor

Figure 3: Lymphangioma circumscriptum: Isolated group of clustered vesicles with hemorrhagic fluid are seen over the back of the right thigh

Figure 4: Lymphangioma circumscriptum: [H & E, 10X]. Showing irregular acanthosis and multiple dilated lymphatics lined by endothelial cells in the dermis and containing numerous erythrocytes and few lymphocytes

Histopathology of a vesicle showed irregular acanthosis of the epidermis, dilated lymphatics lined by endothelial cells in the dermis and containing numerous erythrocytes, macrophages and lymphocytes within the lumen [Figures 4 and 5].

The patient was referred to the department of plastic surgery for cosmetic debulking of the tumor. She was also reassured regarding the benign nature of the tumor.

DISCUSSION

LC, or microcystic lymphatic malformation, is a benign tumor of the lymphatic system with inherent hamartomatous lymphatic malformation. The synonyms are “capillary lymphangioma,” “lymphangiectasia” and “dermal lymphangioma.” The basic
pathology in LC is presence of dilated muscle-coated lymphatic cistern in the subcutaneous plane with communication to the large dermal lymphatics and finally forming multiple clustered vesicles with clear, yellow, pink or hemorrhagic fluid over the skin surface as blow-out phenomenon. These abnormal lymphatics are not connected to the normal lymphatics. The basic developmental defect is sequestration of cisterns in the plane of subcutaneous tissue. Lymphedema will be absent in the diseased area because of the presence of normal lymphatics.

In 1970, Peachey *et al.*[^3] divided LC into classic and localized forms. The classic variety of LC is seen at or soon after birth, and is often larger than 1 cm², seen over the proximal limbs. The localized form can be seen at any age and is smaller than 1 cm². For clinical practice and from the treatment point of view, LC can be divided into smaller lesion (less than 7 cm) and larger lesion (more than 7 cm). LC can occur as a congenital or acquired form. The congenital form will be larger in presentation and the acquired form will be small and circumscribed. In a large case series by Ghaemmaghami *et al.*, they described 13 cases of congenital and 24 cases of acquired lymphangioma circumscriptum over the vulva.[^4]

The LC is asymptomatic in its localized form, but the most common symptom is oozing of clear fluid mixed with blood, which occurs spontaneously or after minor trauma. The common sites of involvement are axillary folds, shoulder, upper arm, scrotum, penis, rectum and vulva.[^5] Rarely, it can present in zosteriform distribution.[^6] LC may mimic a number of disorders like herpes zoster, molluscum contagiosum and viral warts.

The complications are rare in LC. The most common complications are bleeding, pain and infection by *Staphylococcus aureus* causing cellulitis. The chance of malignancy is rare. Few cases of lymphangiosarcoma have been reported in patients previously treated by radiation therapy. The prognosis of LC is excellent as it is non-malignant. Over the years, few reports of squamous cell carcinoma have been reported in old lesions of LC. The common associations of LC are lymphedema of lower limb and cystic hygroma, and there few reports of Proteus, Cobb and Maffucci syndromes.

Apart from histology and lymphangiography, MRI is very useful in the diagnosis of LC and the modality of choice in the management of LC. The dermatoscopic examination of LC reveals a lacunar and saccular pattern. Presence of clear fluid appears as brown lacunas with pale septa; if there is blood in the LC, it may show scattered or uniform areas of redness in the lacuna depending on the extent of bleeding.

The surgical excision is the main modality of treatment, although the recurrence is common, with a cure rate of 75%.[^7] Other treatment options are carbon dioxide laser, pulsed dye lasers, intense pulse light,[^8] sclerotherapy, cryotherapy, superficial radiotherapy and electrocautery. Mosche Lapidoth *et al.* have successfully used the combination of radiofrequency current and 900 nm diode laser in treating six patients of LC.[^9] The primary goal of treatment is to remove or destroy the diseased lymphatics and subcutaneous components that serve as a nidus for recurrence.

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**Figure 5:** [H & E, 40X]. Dermis showing dilated lymphatics lined by plump endothelial cells.

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