# Zosteriform lymphangioma circumscriptum

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SUMMARY

**Lymphangioma circumscriptum (LC) is a form of lymphangioma involving skin and subcutaneous tissue. It is evident as translucent vesicles of varying size, though commonly 2 to 4 mm, and of a pink, red, or black hue. It is localized to the dermis, frequently extending deeply and laterally. LC may resemble other entities, such as metastatic carcinoma of the skin, lymphangiectasis, or herpes zoster. We report an unusual verruciform, zosteriform form of LC.**

KEY WORDS

**Lymphangioma circumscriptum, zosteriform, congenital anomaly, vascular anomaly, lymphangiectasia**

## Introduction

Lymphangiomas are congenital malformations of the lymphatic system that may involve skin and subcutaneous tissue. These hamartomatous malformations comprise 25% of benign vascular tumors in children (1). They are not familial (2). Lymphangioma circumscriptum (LC), cavernous lymphangiomas, and cystic hygromas are the common forms of this condition. LC is a disorder involving superficial dilations of lymphatic vessels which communicate with deeper subcutaneous lymphatic cisterns via small channels. These abnormal malformations are not connected to normal lymphatics (3). Vesicles of LC exude a clear fluid and are prone to superinfection. They can appear over any area of the body and may have a verrucous appearance (3). LC may resemble a number of disorders including herpes zoster, viral warts, and molluscum contagiosum (4–7). Surgical excision is the mainstay of treatment, though recurrence is common, with a reported first-time cure rate of 75% (2, 8). It is recommended to surgically excise to the level of the deep fascia and obtain clear margins on frozen section to reduce recurrence (9).

## Case report

A 12-year-old boy was seen with a three-year history of a weeping and oozing plaque starting on the right upper back and extending around the flank to below the right axilla. It began as a small solitary nodule on the back that gradually enlarged. It was flesh-colored and painless. More oozing plaques developed, progressing from brown to black vesicles which eventually ruptured with a yellow fluid. These cycles of vesicles and rupture were associated with pain and tenderness. A primary care physician prescribed oral antibiotics which reduced the pain and led to further crusting. The diagnosis of molluscum contagiosum was suggested. After four

years of such cycles, the patient was admitted for a 10-day period, due to severe back pain attributed to the unilateral cutaneous plaque. Infectious disease specialists diagnosed herpes zoster, and he was started on intravenous acyclovir with no improvement.

On examination, this obese, otherwise apparently healthy boy with normal vital signs had a 10 × 30 cm plaque distributed along the fifth thoracic dermatome on the right mid-axillary skin (Figure 1). It appeared erythematous and verrucous and was studded with vesicles and dome shaped pink papulonodules. Areas of coalescence of the papules and yellowish crusting were apparent. The entire region was tender to light contact.

Complete blood count and routine serum chemistries were remarkable for a white blood cell count of 11,000/mL, 65% neutrophils, and 25% lymphocytes. The punch biopsy specimen

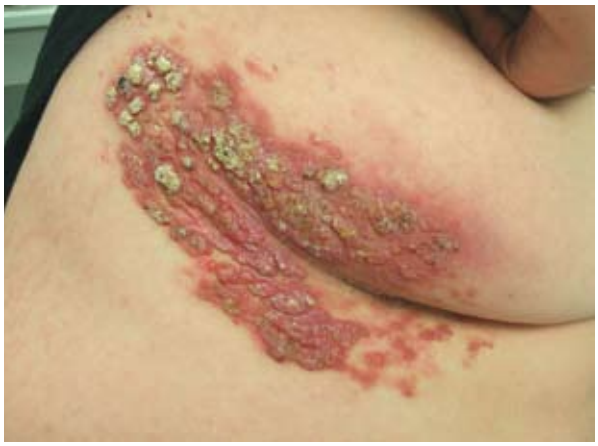

**Figure 1. 12-year-old boy with unilateral zosteriform lymphangioma circumscriptum on right T5 dermatome region.**

showed marked epidermal hyperplasia projecting downward into marked vascular dilation within the superficial and deep dermis. Grouped and solitary cystic lymphatic spaces lined by endothelium were present in the upper dermis contributing to the bulging appearance on the surface. Coagulated lymph or traces of red blood cells were present in the cystic spaces. CD31 staining of the vascular structures suggested a lymphatic origin (Figure 2). The specimen showed gram-positive bacteria in clusters on the surface and in the stratum corneum. Lymphangioma circumscriptum was confirmed. Bacterial culture showed heavy growth of *Staphylococcus aureus* and a few colonies of *Klebsiella pneumoniae*. Fungal cultures were negative. An accurate MRI reading was unattainable because of sub-optimal scan secondary to patient non-compliance. The available MRI interpretation was consistent with LC and epidermal nevus.

Treatment of infection included triple antibiotic ointment twice a day, an oral course of cephalexin 500 mg twice a day for 10 days, and antibacterial soap. After one week of treatment, the yellow crusting resolved to reveal vesicular verrucous plaques with multiple vesicles, some with a red tinge, clinically appearing as LC. Unfortunately, the patient's excessive weight made him an unacceptable surgical candidate.

## Discussion

LC is a benign disorder affecting the lymphatic channels. It was initially described as lymphangiectodes in 1879 by Fox and Fox (10). It was renamed to lymphangioma circumscriptum by Morris in 1889 (11). Its dermatopathology was well described by Whimster et al. (3) as a cluster of small, thin-walled vesicles resembling frog spawn. These vesicles communicate with deeper subcutaneous lymphatic cisterns and contain a clear, yellow- or pink-colored fluid, depending on the extent of hematologic extravasation. The deeper abnormal cisterns have a thick, smooth muscle coat which serves to pump lymph into the more superficial vesicles over extended periods of time (3). LC histology shows dilated lymphatics which expand into the papillary dermis. They are lined by flat endothelium, and contain red and white blood cells (3) (Figure 2).

In 1970, Peachey et al. (12) divided LC into two main groups: classic and localized. The classic form of LC is typically seen at or soon after birth, is often larger than 1 $cm^2$, and usually covers the proximal limbs. It is unusual for the classic form to progress from its original vesicular appearance to warty plaques. Conversely, the localized form is seen at any age, is often less than 1 $cm^2$, and may appear anywhere on the body (12). In neither form has communication between the abnormal lymphatics and the normal lymph system been shown (12). The patient discussed here is a unique overlap of Peachey's classifications, as he is 12 years old and possesses a large 10 × 30 cm lesion. At least one other case has been reported of a giant LC in an adolescent, but not with a zosteriform appearance (7).

This patient was also unusual for having tender, painful LC with recurrent vesicular rupturing and secondary infection. Most often, LC is asymptomatic. The verrucous appearance and extended course led the early diagnosis to molluscum contagiosum. Intertriginous sites similar to this

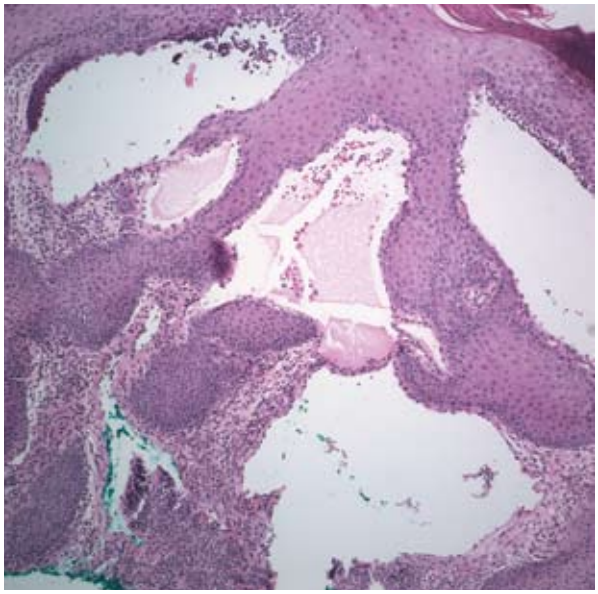

**Figure 2. Histological features showing cystic lymphatic spaces lined with a flattened endothelium within the dermis of a 12-year-old patient (Hematoxylin – Eosin × 100).**

patient's LC are common (5, 6). Herpes zoster is another differential diagnosis. The appearance in this patient was zosteriform, within the T-5 dermatome region. At least one other case of such a presentation has been reported in a middle-aged patient with localized LC, devoid of superinfection (13). Herpes zoster is rare in children; fewer than 10% of all herpes zoster patients are under 20 years of age, and fewer than 5% under 15 years of age (4). Resemblance is possible, because herpes zoster may present with a vesicular rash which can become superinfected. In fact, many skin diseases may have distributions in a zosteriform pattern (14). Other differential diagnoses include herpes simplex, dermatitis herpetiformis, hemangiomas, acquired lymphangiectasis, and metastatic cancer of the skin (15–19). In addition to histology, an MRI may assist LC diagnosis and treatment by describing the extent of the malformation.

This patient did not qualify for standard treatment with surgical excision. His obesity restricted medical clearance. However, he received therapy for the overlying infections and wound care for the ruptured vesicles. The usual treatment for LC is surgical excision after antibiotics have been employed. Additional options include carbon dioxide lasers, pulsed dye lasers, sclerotherapy, cryotherapy, and cautery (8, 20, 21). Radiation therapy is not recommended as malignant conversion has been described (22). The primary goal is complete removal or destruction of both the diseased lymphatics and the subcutaneous components which serve as a nidus for recurrence.

Malignant transformation of LC is rare. Ruocco's immunocompromised district might be recalled (23,24), as should other potential triggering factors (25). A small number of lymphangiosarcomas have been noted on sites of LC where radiation therapy was previously performed (22). There is an additional report of a Dabska tumor, or papillary intralymphatic angioendothelioma, forming within an adolescent patient's LC. In this case, the lymphangioma recurred after excision, but the Dabska tumor did not (26,27). Squamous cell carcinoma has also been previously described arising within LC (28).

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