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Visual Diagnosis in Emergency Medicine

RECURRENT CHEST WALL AND AXILLARY LESIONS IN A TEENAGE BOY

Emily M. Ayub, DO,* Davis W. Bateman,† and Shabana Yusuf, MD, MED‡

*Department of Pediatrics, Section of Emergency Medicine, Baylor College of Medicine, Texas Children's Hospital, Houston, Texas, †Baylor College of Medicine, Baylor University, Houston, Texas, and ‡Division of Pediatric Emergency Medicine, Department of Pediatrics, Baylor College of Medicine, Texas Children's Hospital, Houston, Texas

Reprint Address: Shabana Yusuf, MD, MED, Department of Pediatrics, Section of Emergency Medicine, Baylor College of Medicine, Texas Children's Hospital, 6621 Fannin St., A 2210, Houston, TX 77030

CASE REPORT

A 13-year-old boy presented to the emergency department (ED) with a history of recurrent skin lesions on his chest and axilla. They acutely increased in size with intermittent bleeding, prompting him to present to the ED. The lesions first appeared in infancy. His family had recently relocated from another state where the patient reportedly had received multiple laser treatments. The patient had not established care with a primary care physician. No medical records relating to diagnosis or treatment received were available at the time of the ED visit. The lesions were not painful, were mildly pruritic, and bled with irritation to the area (Figure 1). They continued to recur despite a history of multiple laser treatments and surgical resections. Dermatology was consulted in the ED and the patient was diagnosed with lymphangioma circumscriptum (LC). He was referred to the vascular anomalies clinic for further care.

DISCUSSION

LC is a benign hamartomatous malformation of the lymphatic channels of the skin originally described by Fox and Fox and termed "lymphangiectoides" (1). It is a cutaneous subtype of lymphangioma and was later named LC by Morris (2). Lymphangiomas are a group of disorders that are lymphatic malformations and were originally classified as simple, cavernous, and cystic by Wegner (3). They were later reclassified into superficial and deep types. The former includes cutaneous LC and the latter includes cavernous lymphangioma and cystic hygromas. Lymphangiomas occur at the rate of 1.2–2.8 per 1000 live births and account for 4% of all vascular tumors. They may present at birth or appear in the first few years of life.

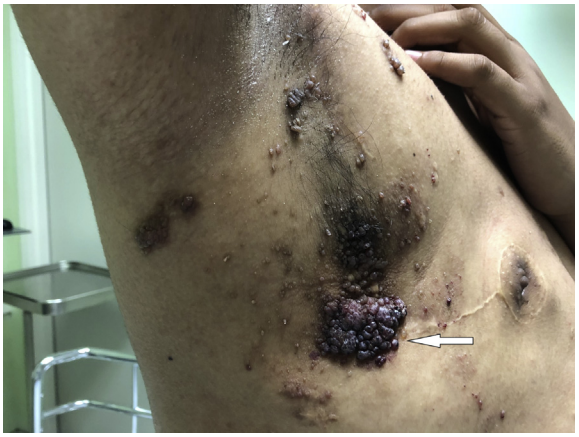

Figure 1. A mass of clustered vesicles lateral to the right nipple with numerous satellite vesicles across the chest and axilla with multiple scars from previous surgeries.

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The lymphatic system develops abnormally in these patients, with lymphatic channels called cisterns communicating with the lymphatic system in the subcutaneous tissue without systemic lymphatic communication (4). Abnormal dilatations of these cisterns lead to protrusions of lymph vessels into the skin.

LC first appears as translucent vesicles that cluster together and may coalesce to large bullae. The color varies from pink and purple to black because of hemorrhage and resembles a frog spawn (5). The vesicles may intermittently leak blood-tinged lymphatic fluid and can undergo hyperkeratotic transformation with a verrucous appearance resembling warts. It most frequently involves the oral cavity including the tongue, proximal extremities, axilla, and trunk. It can uncommonly involve the groin, vulva, penis, and scrotum.

The differential diagnosis includes carcinoma telangiectoides, hemangioma, and lymphangiectasis. Lymphangiectasis patients often have a history of surgical resection or radiation in the area affected. Carcinoma telangiectoides is a cutaneous metastatic condition with underlying, undetected malignancy. Timely diagnosis of carcinoma telengiectoides and location of the underlying primary malignancy is imperative. Other conditions, such as warts, molluscum contagiosum, angiokeratoma, and lymphangioendothelioma can mimic LC. Diagnosis can be established clinically. When in doubt, lesions can be biopsied or visualized by dermoscopy (6). The most common complications include bleeding and secondary infection, typically by *Staphylococcus aureus*. Lymphedema and cystic hygromas are commonly associated with LC. A small percentage of patients are at risk of developing lymphangiomasarcoma and squamous cell carcinoma, especially in chronic cases.

LC is difficult to treat given its deep cisternal element. The goals of therapy are to improve cosmetic outcome, decrease pain, reduce infection, and prevent recurrence. If infection is suspected, antibiotic therapy with staphylococcal coverage and wound cultures are warranted. Palliative treatments include cryotherapy, sclerotherapy, and laser therapy. Radiation therapy is not used because it predisposes to malignancy.

Definitive treatment is surgical excision, although complete resection is often difficult with extensive disease (7). The recurrence rate with surgical resection is about 17% (4). Resections should include clear margins so that all the cisternal elements are excised; otherwise, recurrence is high. Imaging with ultrasound and magnetic resonance imaging can guide surgical excision. If resection is extensive, primary closure might not be possible and skin grafting will be required.

In summary, LC is a benign lymphatic malformation resembling frog spawn that often presents in the first few years of life. It can be difficult to treat and may present with hemorrhage, discomfort, or secondary infection. Surgical resection is definitive treatment, with laser or sclerotherapy as adjuncts. It is important to recognize this rare condition because timely referral to a specialist likely improves outcomes.

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