

What Is Your Diagnosis?



Patients reported leakage of straw-colored fluid when the lesions were traumatized.

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The Diagnosis: Lymphangioma Circumscriptum (Benign Lymphangiectasia)



Lesions of lymphangioma circumscriptum look like clusters of clear fluid-filled vesicles resembling frog eggs. The lesions may occur in a zosteriform distribution.¹ Verrucous hyperplasia of the epidermis and associated tissue edema may be noted. Some vesicles contain blood in addition to lymph and Kim et al² reported solitary nodular angiokeratoma in a lesion of lymphangioma circumscriptum. Lymphangioma circumscriptum has been reported to masquerade as contact dermatitis.³

Rarely, lymphangioma circumscriptum may involve the eyelids and conjunctiva.⁴ Oral lesions have been described.⁵ Pauwels and Koedam⁶ reported on lymphangioma circumscriptum involving the tongue. Vulvar, penile, perianal, or scrotal lymphangioma circumscriptum may have a markedly verrucous or cerebriform appearance

and may be misdiagnosed as condylomata acuminatum.^{7,8} Vulvar lesions may occur following surgery or radiation therapy, or they may arise spontaneously.⁹ The most common patient complaint relates to the clear fluid oozing from the lesions. Pain and cellulitis also may occur as complications.¹⁰ Squamous cell carcinoma has been reported in long-standing lymphangioma circumscriptum.^{11,12} Cobb syndrome, the association of spinal angioma with a cutaneous angioma or nevus flammeus, has been reported in association with lymphangioma circumscriptum.¹³

Rather than representing a neoplastic proliferation of lymphatic vessels, lymphangioma circumscriptum appears to represent localized lymphangiectasia in response to chronic lymphostasis. Lymphangioma circumscriptum may be associated with deep lymphatic involvement

and enlargement of the extremity.¹⁴ Superficial lesions of lymphangioma circumscriptum may be associated with deep cystic hygroma, mediastinal lymphangioma, or retroperitoneal lymphangioma.^{15,16} Lesions presenting after surgery or radiation therapy suggest damage to lymphatic vessels resulting in lymphostasis.¹⁷ Lymphangioma circumscriptum may occur in areas of chronic lymphedema several years after mastectomy and radiation therapy.¹⁸ Vulvar lymphangiectasia may occur in association with or without chronic lymphedema of a lower extremity following therapy for squamous cell carcinoma of the cervix.^{19,20} Scarring of lymphatics following infections such as erysipelas or cellulitis may result in acquired lymphangiectasia resembling "spontaneously occurring" lymphangioma circumscriptum.²¹ Chylous reflux into the skin in a patient with intestinal lymphangiectasia was reported to resemble lymphangioma circumscriptum.²² Ultrastructure of superficial and deep vessels in lymphangioma circumscriptum suggests that the lesions are the result of chronic localized lymphostasis.²³ Histopathologically, there are no features that differentiate "spontaneously occurring" lymphangioma circumscriptum from secondary lymphangiectasia.²⁴

Lymphangiosarcoma rarely arises in the setting of lymphangioma circumscriptum. Prior irradiation of the lymphangioma has been implicated as a possible etiologic factor of malignant degeneration.²⁵ Rarely, angiosarcoma occurring in a chronically lymphedematous extremity following mastectomy may resemble benign lymphangioma circumscriptum.²⁶

Recurrence is common after surgical excision. Steps to improve surgical outcome include complete excision of the lesions down to fascia, which can be aided by frozen section, or partial excision of only the deepest or most troublesome areas of extensive lesions.^{27,28} Selective excision of the deep underlying cisterns has been suggested and vesicles have been reported to regress after deeper components of larger lesions have been excised.²⁹ Magnetic resonance imaging of the deeper component may be helpful in presurgical evaluation of the patient.³⁰ Obliteration of deep underlying cisterns using suction-assisted lipectomy has been reported.³¹ Although obliteration of deep lymphatics has been suggested in patients with penile lesions, less aggressive surgical management can achieve good results in some cases.³² We have seen significant scarring and recurring lesions within large scars as complications of surgical treatment of penile lymphangioma circumscriptum.

Treatment with the argon laser has resulted in some scarring and a high rate of recurrence.³³

Anecdotal success has been reported with both the ultrapulsed and conventional CO₂ laser, as well as the flashlamp pulsed dye laser and sclerotherapy.³⁴⁻³⁸ Long-term follow-up of these patients will determine the frequency of recurrence. Superficial radiation has been reported to produce almost complete regression of lymphangioma circumscriptum.³⁹ This mode of treatment is intriguing because deeper radiation has been implicated as a cause of lymphangioma circumscriptum. Radiation has been suggested as an appropriate therapy for patients with unresectable lesions or for patients who are not willing to undergo surgery.⁴⁰

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