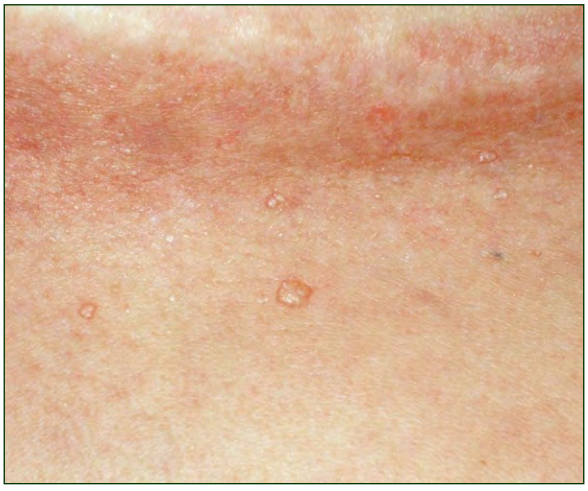
What Is Your Diagnosis?



A 56-year-old white woman with a history of stage III carcinoma of the right breast and subsequent right mastectomy, chemotherapy, radiotherapy, and a transverse rectus abdominis myocutaneous flap breast reconstruction 7 years prior presented with new skin lesions of several months' duration that had slowly developed on her right axilla and under her reconstructed right breast. The lesions were not painful and did not bleed, drain, or itch. One year prior to presentation she visited her primary care physician with a concern of back pain, and a bone scan revealed a lesion on the left ninth rib that was resected and determined to be benign on pathologic examination. She denied any further symptoms such as new breast lumps or masses. She was otherwise healthy with a history of hypertension, hyperlipidemia, bladder spasms, and gastroesophageal reflux. Her medications included esomeprazole, oxybutynin, and letrozole. Physical examination revealed multiple clusters of small, flesh-colored, fluid-filled vesicles ranging from 2 to 6 mm that were located just below the inframammary fold of her reconstructed right breast. There was no notable lymphedema, no palpable lymphadenopathy, and no breast masses or subcutaneous nodules. A 4-mm punch biopsy was obtained.

PLEASE TURN TO PAGE 291 FOR DISCUSSION

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The Diagnosis: Acquired Cutaneous Lymphangiectasia

Lymphangioma circumscriptum (LC) is a rare microcystic lymphatic malformation due to lymphatic hyperplasia that typically is seen in the pediatric population as a congenital disease.[1] It also can be acquired in patients with chronic lymphedema or damage to lymphatic drainage from surgery and/or radiotherapy. In these cases the term *acquired cutaneous lymphangiectasia* (ACL) may be preferred, though the acquired lesions are clinically and histologically indistinguishable from the congenital form.[2] Acquired lesions have been reported following the treatment of a variety of cancers such as vulvar cancer and cervical cancer,[3] but most reports are in breast cancer patients after mastectomy with adjuvant radiotherapy.[4-7] The classic clinical presentation of LC includes a frog spawn appearance due to the flesh-colored pink or red clusters of translucent vesicles filled with clear or serosanguineous lymph (Figure 1). Histologic examination reveals dilated irregular lymphatic channels with thin endothelium in the upper dermis and occasional epidermal acanthosis or hyperkeratosis, as seen in our patient's punch biopsy (Figure 2). Dermoscopy also may aid in diagnosis.[8]

Congenital LC and ACL have no malignant potential, as evidenced by a clinicopathologic analysis of 56 radiation-induced vascular proliferations in patients with breast cancer. The study determined that a large majority of the lesions had a benign clinical outcome.[6] Although vascular endothelial growth factor C is overexpressed in deeper cavernous lymphangiomas, which hints at the potential for proliferative activity, such expression is absent in superficial lymphangiomas such as LC and ACL.[9] Nevertheless, accurate diagnosis is important, particularly because ACL is most often seen in patients who have been treated for breast cancer. Lesions that clinically appear to be classic LC may actually be breast cancer metastases to the skin (eg, carcinoma telangiectodes).[10] Our patient had a history of breast cancer, back pain, and a bone lesion. Therefore, although the bone lesion was determined to be benign by pathology, a thorough history and physical examination assessing for new fevers, weight loss, lymphadenopathy, or breast masses was performed, as well as biopsy with histologic evaluation.

The pathogenesis of ACL is thought to be secondary to damaged lymphatic tissue, resulting in ineffective drainage and increased hydrostatic pressure, which in turn causes dilatation of the superficial lymph vessels. However, clinically ostensible lymphedema is not always present. In addition, although

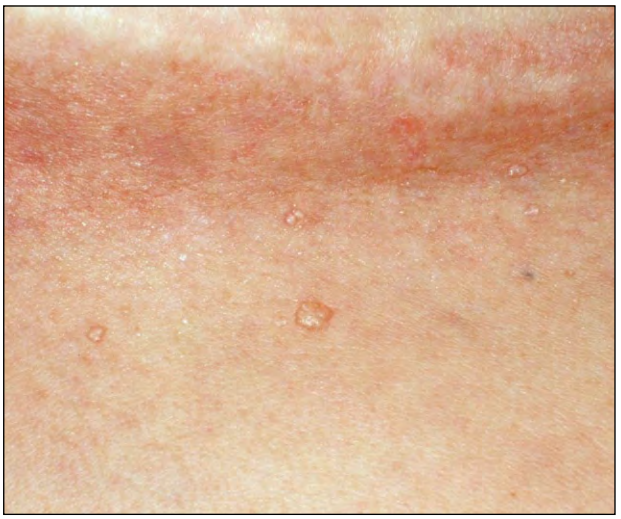

Figure 1. Multiple clusters of small fluid-filled vesicles below the right inframammary fold.

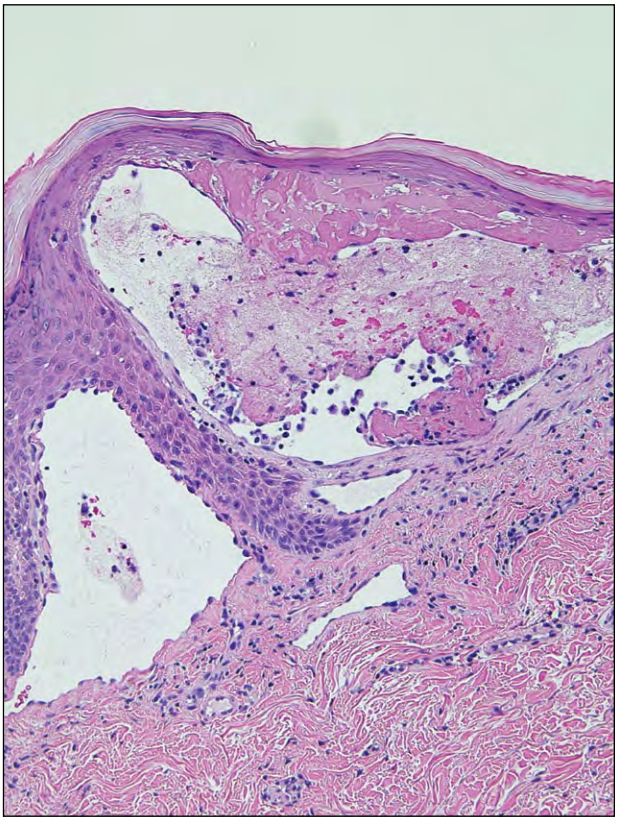

Figure 2. Punch biopsy specimen from below the right inframammary fold (H&E, original magnification ×20).

most cases of ACL arise in patients who have had both surgery and radiation therapy, either treatment alone presumably can induce sufficient lymphatic damage, as ACL has been reported after a radical mastectomy with no radiation[11] and after radiation therapy with no surgery.[12] Although LC and ACL are benign lesions, they can have deep components comprised of large lymphatic cisterns, thus explaining the high recurrence rate. A report of an LC lesion spreading to the muscle and subcutaneous fat adjacent to the left kidney of a 7-year-old boy suggests that even deeper spread is possible.[1] Ultrasonography or magnetic resonance imaging can be useful in these cases to adequately assess the depth of the lymphatic cisterns before surgery. Most LC and ACL lesions are asymptomatic, except for cosmetic concerns, but some may be associated with chronic drainage, pain, or infection.

Treatment of LC and ACL is difficult due to its deep components and different opinions exist regarding the most adequate therapy. Surgical therapy is the most common treatment and has a low recurrence rate, but other modalities such as cryosurgery, electrocautery, laser phototherapy, and even radiotherapy have been utilized with success. Sclerotherapy and intense pulsed light also are treatment options.[13-18]

Acquired cutaneous lymphangiectasia is a benign finding seen in patients with lymphatic damage. Most cases of ACL have been reported following therapy for breast cancer, but accurate diagnosis is important. Due to the potential for cancer recurrence and the possible clinical similarity to the frog spawn appearance of LC, sufficient workup must be established.

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