A 58-year-old woman presented for evaluation of asymptomatic vesicles near the right breast. She first noticed them 1-2 years ago and denied receiving prior treatment for the lesions. Medical history was significant for lobular invasive carcinoma of the right breast—status after chemotherapy, right mastectomy, and completion of radiation therapy 5 years ago. She also underwent a left mastectomy and bilateral breast reconstruction 4 years ago. On physical examination, multiple clustered, translucent papulovesicles were noted lateral and inferior to the right breast (Fig 1). The remainder of the examination was unremarkable. A 4-mm punch biopsy was performed (Figs 2 and 3).
Question 1: Based on the clinical presentation and histopathologic findings, what is the most likely diagnosis?

A. Lymphangiosarcoma  
B. Lymphangioma circumscriptum (LC)  
C. Carcinoma telangiectodes  
D. Acquired cutaneous lymphangiectasia (ACL)  
E. Herpes zoster

Answers:

A. Lymphangiosarcoma — Incorrect. Stewart-Treves syndrome is lymphangiosarcoma that develops in the setting of chronic lymphedema after mastectomy. It is classically characterized by red-blue macules or patches that progress to nodules on the ipsilateral extremity.\(^1\) It is less likely in this patient without long-standing lymphedema. Proliferating vascular channels and hyperchromatic, pleomorphic endothelial cells are seen on histology.

B. LC — Incorrect. LC is a result of developmental malformation of lymphatic vessels. Though this presents clinically and histologically similar to ACL, LC typically presents at birth or early childhood.\(^2,3\)

C. Carcinoma telangiectodes — Incorrect. Carcinoma telangiectodes is cutaneous metastasis, most commonly associated with breast cancer, that results from local invasion of the vascular or lymphatic system. It may present as an erythematous telangiectatic patch or as papules, nodules, or pseudovesicles. Tumor emboli within dilated lymphatic vessels are seen on histology.

D. ACL — Correct. ACL typically presents as clusters of 2-10-mm papules and vesicles that vary in color from translucent to red or purple, due to the varying content of red blood cells.\(^3,4\) In some cases, they have a verrucous surface or appear as hyperpigmented maculae.\(^3\) ACL often occur in an area with existing lymphedema; however, it is not always present,\(^5\) as in this case. Dilated lymphatic vessels in the papillary and reticular dermis are seen on histology. Lumina may contain scant or absent erythrocytes.\(^3\)

E. Herpes zoster — Incorrect. A patient this age may develop shingles and present with a vesicular eruption. However, lesions are usually painful, follow a dermatomal distribution, and resolve within weeks.

Question 2: Which of the following is most commonly associated with the development of these lesions?

A. Surgery and radiation therapy  
B. Breast cancer  
C. Infection  
D. Older age  
E. Immunocompromised state

Answers:

A. Surgery and radiation therapy — Correct. Surgery and radiation therapy for malignancy are the most common causes of ACL.\(^3\) Pathogenesis involves the destruction of previously normal lymphatics. The congestion and resulting superficial lymphatic dilation manifest as the lesions of ACL, which have been reported to develop as late as 25 years after the causative factor.\(^4\)

B. Breast cancer — Incorrect. Breast cancer is the most common cause of carcinoma telangiectodes and is the most common cause of cutaneous metastases in women.\(^3\)

C. Infection — Incorrect. Infection-related complications (eg, tuberculosis-associated scarring)\(^3\) have been associated with ACL, but they are not a common cause. However, rupture of ACL lesions can create a portal of entry for bacteria, and this can give rise to complications such as cellulitis.\(^4\)

D. Older age — Incorrect. Older age is a risk factor for developing shingles, which is most common in individuals over the age of 50.

E. Immunocompromised state — Incorrect. Immunocompromised individuals are at an increased risk for the development of shingles and viral warts, which are lesions that ACL may mimic.

Question 3: Which is an appropriate option for the management of this patient’s lesions?

A. Wide local excision  
B. Oral antivirals  
C. Chemotherapy  
D. Oral antibiotics
E. Observation

Answers:

A. Wide local excision — Incorrect. This is an option for Stewart-Treves syndrome, though the prognosis remains poor given the disease’s aggressive course.

B. Oral antivirals — Incorrect. This is the treatment for uncomplicated herpes zoster.

C. Chemotherapy — Incorrect. This would be an option for treating an underlying primary malignancy such as breast cancer.

D. Oral antibiotics — Incorrect. This patient does not have an infection, though topical mupirocin may be used to prevent superinfection of any ruptured ACL lesions.¹

E. Observation — Correct. ACL lesions are generally stable, chronic, asymptomatic, and do not require treatment. Treatment options that have been reported with varying efficacies include cryotherapy, electrodessication, laser therapy, surgical excision, and sclerotherapy.¹,²

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Abbreviations used:

ACL: acquired cutaneous lymphangiectasia
LC: lymphangioma circumscriptum

Conflicts of interest

None disclosed.

REFERENCES