CORRESPONDENCE

Distinction of a postradiation atypical vascular lesion from cutaneous angiosarcoma in a patient with a history of breast cancer

Dear Editor,

A 60-year-old woman with a medical history of infiltrative ductal carcinoma of the right breast underwent breast-conserving surgery and axillary lymph node dissection, followed by adjuvant chemotherapy, hormone therapy, and radiotherapy (conventional radiotherapy, 25 fractions for a total dose of 5000 cGy). Six years after treatment, asymptomatic skin eruptions on the right side of the chest and axilla were observed for 4 weeks. The patient was referred to the dermatology department for suspected cutaneous metastases. Physical examination revealed multiple discrete papules, clear fluid-containing vesicles, and lesions with a mild erythematous base on the right side of the chest. A grayish, rubbery but compressible cystic nodule, 1.5 cm in diameter, was observed near the right axillary fossa (Figure 1). No regional lymphedema or significant sclerotic change of the skin was noted. Excisional biopsy of the 1.5-cm cystic nodule was performed.

Histopathological analysis revealed diffuse anastomotic vessels and dilation of the vascular space, mostly in the deep dermis (Figure 2A), with concomitant dissection of collagen bundles in some areas (Figure 2B). In the subcutis, more dilated vascular channels with stromal projection were observed (Figure 2C). The endothelial cells were plump, discontinuous in a single layer, without cytologic atypia (Figure 2D). Immunohistochemistry revealed positive staining for CD31, positive staining for CD34 (Figure 2E), and negative staining for smooth muscle actin (Figure 2F). A postradiation lymphatic type atypical vascular lesion (AVL) was diagnosed.

The patient decided to forego complete residual lesion removal. On follow-up, breast cancer recurrence was not noted.

The term AVL was coined by Fineberg and Rosen in 1994.¹ They examined seven patients with breast cancer who had undergone lumpectomy and adjuvant radiotherapy. Three patients were subsequently diagnosed as having cutaneous angiosarcoma (AS) and four were diagnosed as having AVLs. Pathologically, AVL is defined as the proliferation of anastomosed vascular structures with relative circumscription in the dermis, lined with hyperchromatic endothelial cells, with stromal projection into the preliminary lumen, and slight to no dissection of dermal collagen.¹ Several reports have confirmed the original description of AVLs.²⁻⁴ Other conditions such as “acquired progressive lymphangioma”, “benign lymphangiomatous papules”, “lymphangioma circumscriptum”, and “benign lymphangioendothelioma” have previously been described as AVLs; these are now collectively termed as AVLs.

Clinical distinction of AVLs from well-differentiated cutaneous AS is challenging because their clinical settings and cutaneous morphologies overlap. Postradiation cutaneous AS usually presents as bruise-like plaques, nodules, indurated skin, and skin ulceration.⁵,⁶ In addition, chronic edema may indicate AS, known as a condition of Stewart–Treves syndrome. AVLs often manifest as one or more skin-colored papules, erythematous to violaceous macules, and the appearance of clear fluid-containing vesicles or nodules.⁴,⁵,⁶ AS usually arises from thickened mammary skin, often with multiple large lesions (median, 7.5 cm), whereas AVLs are smaller (median, 0.5 cm).⁵,⁶ The time period from radiation to the development of lesions is slightly longer for AS (4–6 years) than for AVL (3–4 years).²,⁴,⁶ Dermal anastomosing vessels and hyperchromatic endothelial cells are common features of AVLs and cutaneous AS. However, AVLs lack the characteristic features of AS such as infiltration into the subcutis, diffuse dissection of collagen bundles, hemorrhage with blood lakes, multilayering of endothelial cells, papillary endothelial hyperplasia, and most importantly, cytologic atypia including prominent nuclei and mitotic figures.¹,⁴,⁶

Figure 1 A grayish, elastic nodule, measuring 1.5 cm, is seen near the right axillary fossa. There are multiple vesicles filled with clear fluid on the right side of the chest. Some lesions display a mild erythematous base.

Conflicts of interest: The authors declare that they have no financial or non-financial conflicts of interest related to the subject matter or materials discussed in this article.
The relationship between AVL and AS remains unclear. The initial report by Fineberg and Rosen\(^1\) suggested that they were distinct entities. However, other studies have suggested the progression of AVL to AS,\(^2,4\) and AS to AVL.\(^3\) Some histopathological features of mastectomy specimens from patients with AS were indistinguishable from those of AVL.\(^3,4\) These rare reports imply the possibility of a morphological continuum between AVL and AS. From the viewpoint of molecular pathogenesis, Santi et al.\(^7\) reported that mutations in the tumor suppressor gene TP53 in contrast to atypical vascular lesions after radiotherapy are exclusively noted in postradiation AVLs.\(^8\) This finding might be a potential diagnostic tool to differentiate the ambiguous lesions of AVL from AS.

Currently, there are no guidelines for the management of postradiation AVLs. The most frequent complication of AVLs is postsurgical recurrence, rather than transformation to AS, and the development of new lesions in the same radiation field, the so-called “field effect”.\(^1\) The incidence of residual or recurrent lesions after variable extent excision is approximately 20–30%.\(^2,9\) Patients with a history of radiotherapy should be carefully monitored for vascular eruptions on the irradiated skin. Once AVL is diagnosed, complete excision with a negative margin is indicated. Careful surveillance and biopsy of any clinically suspicious lesion are necessary.

References