Atypical vascular lesion of the breast

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Atypical vascular lesions (AVLs) are vascular proliferations that develop after surgery and radiation for breast carcinoma and may represent precursors to angiosarcoma. AVLs are not well-known entities and currently lack official prognostic factors and guidelines for surgical treatment. We report the case of a patient who developed an AVL, vascular type, 4 years after lumpectomy and radiation therapy for ductal carcinoma in situ of the breast. The patient underwent wide local excision with 1-cm margins with subsequent pathologic examination confirming complete excision of the residual atypical vascular proliferation. This case highlights the importance of close cutaneous surveillance in patients with a history of surgery and radiation for breast carcinoma, and a low threshold for biopsy. More studies are needed to further delineate the risk of AVLs progressing to angiosarcoma and to identify histologic features or immunophenotypic markers, which may be predictive of this risk. Furthermore, formal treatment recommendations for these enigmatic entities would be helpful. (J Am Acad Dermatol 2010;63:337-40.)

Key words: angiosarcoma; atypical vascular lesion; dermatologic surgery; vascular neoplasms; pathology.

Atypical vascular lesions (AVLs) develop as one or more small erythematous to violaceous macules or papules after radiation therapy for breast carcinoma.[1] Although the specific type, technique, and dosage of prior radiation has not been specifically analyzed, all AVLs are observed within the radiation field. The latency period for development of these lesions ranges from 1 to 20 years after radiation, although presentation within 3 to 6 years is most common.[1,2] The effects of radiation therapy are typically divided into acute or early changes that occur within days to weeks and delayed or late changes that occur within months to years. Early radiation-induced changes occur secondary to necrosis of the rapidly dividing keratinocytes.[3] Dilation of capillaries and increased vascular permeability leads to erythema.[3] Decreased activity in hair follicles and sweat glands can lead to hair loss and xerosis, respectively.[3] By 3 to 4 weeks, warmth, tenderness, and edema can occur in addition to erythema.[3] Thrombosis of vessels, hemorrhage, desquamation, exudation, hyperpigmentation, and ulceration can also occur.[3] Typical late radiation-induced changes seen within the skin include hyalinization of dermal collagen fibers, swelling of endothelial cells, telangiectatic dilation of dermal vessels, and proliferation and hyalinization of deeper vessels.[4,5] Because such skin changes usually stop within 3 years of radiation therapy, observed changes after this period should alert the clinician.[5,6] We report the case of a patient who developed an AVL and suggest that more guidelines would be beneficial to help clinicians treat these patients.

Abbreviations used:

AVL: atypical vascular lesion
LT: lymphatic type
VT: vascular type

CASE REPORT

A 63-year-old woman with a medical history of hypertension, hypercholesterolemia, and estrogen receptor-positive, ductal carcinoma in situ of the right breast presented to the dermatology clinic for evaluation of an asymptomatic, unchanging macule of the right breast that had been present for 6 months. The patient had a history of right breast lumpectomy with axillary node dissection and adjuvant chemotherapy and radiation (32 treatments of external beam radiation therapy for a total of 5000 cGy) 4 years before presentation. She was currently taking the estrogen antagonist anastrozole. Physical examination revealed a 6-mm violaceous macule

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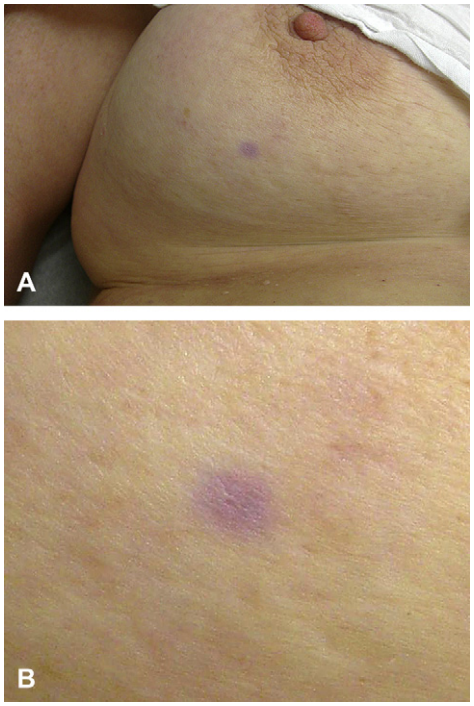

Fig 1. Low magnification (**A**) and high magnification (**B**) of a 6-mm violaceous macule of right breast.

(Fig 1). A punch biopsy specimen revealed diffuse dermal proliferation of well-formed capillary structures with mild endothelial atypia (Fig 2). Staining was positive for the CD34 vascular marker and negative for the D2-40 lymphatic marker (Fig 3). A diagnosis of AVL, vascular type (VT), was made, consistent with the histologic appearance and the history of radiation. The patient underwent wide local excision with 1-cm margins, with pathologic examination confirming complete excision of the residual atypical vascular proliferation.

REVIEW

Patients who undergo conservative surgery and radiation therapy for breast cancer have been observed to have various complications including pneumonitis, arm edema, persistent breast or chest wall pain, breast fibrosis, fat necrosis, prolonged skin breakdown, rib fracture, cardiac complications, neuropathy, vascular thrombosis, and vascular neoplasms, including angiosarcoma and AVLs.[7] The term "atypical vascular lesion" was coined in 1994 by Fineberg and Rosen,[8] who described 4 women with cutaneous vascular proliferations after lumpectomy and radiation for breast carcinoma.[1] They believed these lesions to be benign and related to lymphatic obstruction after surgery and/or radiation-induced dilation of vascular channels.[1,5] Others have used the terms "atypical hemangiomas,"[9] "acquired progressive lymphangioma,"[10] "benign lymphangiomatous papules,"[11-13] "lymphangioma circumscriptum,"[14] and "benign lymphangioendothelioma"[8,14,15] to describe the presumptive benign nature of the same entity.

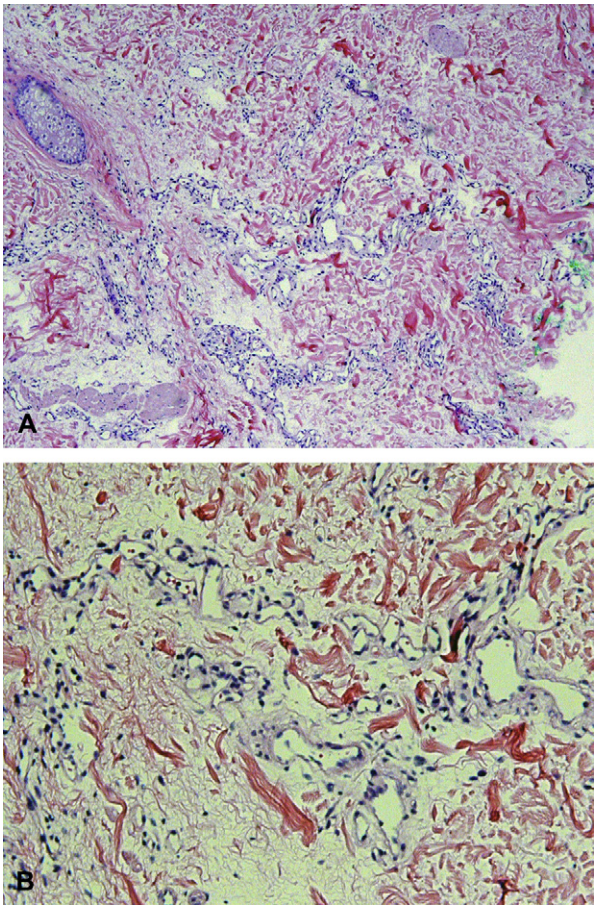

Fig 2. Low- (**A**) and high- (**B**) power image of hematoxylin-eosin stain of atypical vascular lesion, vascular type.

However, in their review of 42 cases, Brenn and Fletcher[14] concluded that AVLs were part of a continuum and were in fact precursors to angiosarcomas that warranted more aggressive treatment.[1] This opinion was based primarily on a patient with a classic AVL in whom serial biopsy specimens showed slow development of angiosarcoma during the next 5 years.[1,14] Other cases of malignant transformation to angiosarcomas are also reported in the literature.[14,16,17] However, despite such contradictory reports, the predominant consensus in the literature remained that AVLs represented a benign entity.[1]

The natural course and malignant potential of AVLs was once again called into question by a 2008 report by Patton et al[1] who reviewed 32 cases of AVLs after surgery and radiation of the breast. The authors divided these into two histologic types: the less common vascular type (VT) was observed to have a higher risk of development into angiosarcoma,