A 60-year-old Caucasian female presented with a 3-month history of exquisitely tender, ulcerating, and bleeding breasts, with a tremendous amount of exudate material bilaterally. This eruption started approximately 6 weeks after cardiac surgery. During the procedure, the patient received heparin, but was not placed on coumadin. She denied exacerbating or alleviating factors. Past medical history is significant for cardiovascular disease, transient ischemic attack, hypertension, and hypercholesterolemia. The patient was a smoker when she was evaluated for this eruption. Her medications upon evaluation included atorvastatin, clopidogrel, lisinopril, metoprolol, and topical lidocaine. Family history was noncontributory. All labs were found to be within normal limits.

Physical exam revealed livedo reticularis on the breasts, bilaterally. The left breast was much more affected than the right, with associated healed punctuate ulcerations and changes of healed infarcts. The rest of her cutaneous exam was negative (Figures 2, 3). Histologic sections of a punch biopsy from the left breast revealed a diffuse capillary network along with mild mixed cell infiltrate and dermal edema, consistent with a vascular process. The biopsy includes CD31 and CD34, vascular markers characteristic of a vascular proliferation. Subsequent pathology revealed focal positivity with CD34, focal cell proliferation between collagen bundles and uniform positivity with CD31.

Discussion

First described in 1994 by Krell et al., diffuse dermal angiomatosis (DDA) is a rare skin condition primarily affecting females and characterized by erythematous, violaceous, indurated plaques which are often ulcerated and tender and commonly localized to the lower extremities.\(^1\)^\(^2\)\(^3\) Although the pathogenesis is unknown, it is often noted in patients with severe peripheral vascular disease among other co-morbidities.\(^2\)^\(^3\) A few authors have reported a correlation between DDA and trauma, namely from surgery. While DDA is rare, with 14 total cases reported, involvement of the breast is even less frequently diagnosed.\(^2\)\(^3\) To date, only 5 cases of DDA of the breast (DDAB) have been described.\(^1\) Although often affecting large pendulous breasts bilaterally, these patients presented in an otherwise atypical fashion without relevant medical history or vaso-occlusive disorder.\(^2\)

Histologically, however, they demonstrated diffuse dermal vascular and endothelial cell proliferation between collagen bundles and uniform positivity with immunoperoxidase stains CD31 and CD34, vascular markers characteristic of DDA.\(^4\)^\(^5\) HBV-8 is also often used to aid in diagnoses and is uniformly negative in DDA.\(^1\)

The exact process underlying the development of DDA has yet to be determined but is thought to be a result of tissue ischemia.\(^1\) The current hypotheses regarding the pathogenesis of the disease are as follows: (1) atherosclerotic plaques may embolize to distal small vessels and create endothelial hyperplasia; (2) vascular steal syndromes can give rise to ischemic necrosis with subsequent ulceration; or (3) ischemia leads to increased vascular endothelial growth factor and subsequent endothelial proliferation.\(^6\)^\(^7\) Given this understanding, it is believed that reversing ischemia and achieving revascularization can be beneficial in improving the clinical signs of disease.\(^8\) Despite a clear mechanism of disease development, several associations have been made between DDA and other co-morbid conditions. Many authors have reported associations between DDA and peripheral vascular atherosclerosis, arteriovenous fistulas, anti-cardiolipin antibodies, hypercoagulable states, and breast ulceration.\(^8\)^\(^9\)\(^10\) The most common and widely accepted association, however, has been vascular exclusive disease.\(^1\) Smoking and DDA have also been found to be strongly associated, with patient’s often having a significant clinical history of long-term tobacco-use. Hypertension has also been reported to be associated with DDA.\(^1\)^\(^4\)

References