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Breast nodularity and ulceration: diffuse dermal angiomatosis a corticosteroid responsive disease

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Abstract

Diffuse dermal angiomatosis of the breast (DDAB) is an uncommon ulcerative angiomatosis, which occurs in middle aged women with large pendulous breasts, a history of cigarette smoking, and risk factors for atherosclerosis. Based on its rarity, no well-defined therapeutic regimen has been elucidated. We report a case of DDAB in a woman with no history of smoking or radiographic evidence of occluded vasculature who presented with ulceration and pain-associated breast nodularity. She had a complete reproducible response to oral corticosteroids.

Keywords: dermal angiomatosis, vascular proliferation, breast nodularity, breast ulceration, prednisone

Introduction

Diffuse dermal angiomatosis of the breast (DDAB) is an uncommon ulcerative angiomatosis with fewer than 50 reported cases. DDAB occurs in middle aged females with large pendulous breasts, a history of cigarette smoking, and risk factors for atherosclerosis. Based on its rarity, no well defined therapeutic regimen has been elucidated. We report a case of DDAB in a woman with no history of smoking or radiographic evidence of occluded vasculature. Unique features included pronounced underlying breast nodularity and a complete and reproducible response to oral prednisone.
A 50-year-old woman was referred to our clinic with a 2 month history of an erythematous plaque on her right breast that had become tender and ulcerated over a four week period. She denied trauma to the breast or a prior history of tobacco use. Her past medical history was significant for end-stage renal disease, hypertension, and obesity. Examination revealed pendulous breasts with pronounced underlying nodularity. The right breast contained an extremely tender peri-areolar ulcer with overlying hemorrhagic crust (Figure 1). The left breast demonstrated a subtle, slightly tender, erythematous and reticulate patch.

Figure 1. Diffuse dermal angiomatosis: large periareolar ulcer with hemorrhagic crusting. Note adjacent subtle reticulated red-brown patch (arrow).

Figure 2. Diffuse dermal angiomatosis: right breast, H&E 100x. The specimen exhibits a percolating dermal proliferation of plump CD31 positive endothelial cells forming numerous small ectatic blood vessels arranged between collagen bundles and dense dermal fibrosis.
Histopathologic examination of a wedge biopsy demonstrated a bland CD31 positive, extravascular endothelial proliferation dissecting collagen bundles, with focal vascular lumen formation (Figures 2, 3). Calcification of subcutaneous vessel walls suggestive of calciphylaxis was not identified and Von kossa staining for calcium was negative. Findings were diagnostic of DDAB.

Vascular studies revealed no upper limb arterial occlusion. Despite adequate wound care, the ulcer rapidly enlarged after biopsy. Prednisone was started at 40 mg daily, tapering by 10 mg weekly until 10 mg, followed by maintenance on 5 mg until resolution. Pain was dramatically reduced within two days of initiating prednisone, accompanied by a rapid decrease in ulcer size and nodularity. Complete resolution was seen over a period of approximately 12 weeks. After 2 months, the patient had recurrence of nodularity, pain, and ulceration. Prednisone 40 mg was again prescribed and again she experienced rapid decrease in pain and complete healing within 3 weeks.

Discussion

Diffuse dermal angiomatosis (DDA) was initially described as a type of reactive angioendotheliomatosis [1], but has been reclassified as a distinct reactive vascular entity owing to the extravascular location of the proliferating vessels. Diffuse dermal angiomatosis of the breast (DDAB) begins with tender reticulate patches, which evolve into painful ulcers in fat-rich dependent areas [2]. Interestingly, both our case and one other, describe underlying tender breast nodularity [2]. This feature likely represents extensive vascular proliferation and when present in association with breast ulceration of unknown origin, may serve as a clinical clue to the diagnosis of DDAB. Nodularity may precede ulceration (as noted in the left breast of our patient), and our case suggests it may be an early clinical indicator disease. Recognition, therefore, may be of particular importance in patients with darker skin types where the early reticulated erythema is subtle and easily missed.

DDAB is rare and occurs in females with pendulous breasts who smoke [2, 3, 4, 5, 6]. Some patients have radiologic confirmation of partial arterial occlusion [2]. The reported associations with positive anticardiolipin antibodies and calciphylaxis, have raised the possibility that DDA represents a reactive proliferation secondary to microvascular occlusion [4, 7]. Cigarette smoking induced endothelial dysfunction and hypercoagulability seem contributory in the majority of reported cases, though not in ours’. Large pendulous breasts appear to be the universal risk factor for DDAB. Repetitive low-grade trauma to relatively poorly perfused, dependent adipose tissue, in the setting of macro- or micro-vascular occlusion, induces a hypoxic tissue state. In this hypoxic state, excessive angioproliferative factors, including VEGF, are produced resulting in excessive endothelial proliferation.
References