Diffuse dermal angiomatosis localized to abdominal striae

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ABSTRACT

Diffuse dermal angiomatosis is a rare, benign, reactive cutaneous vascular disorder considered to be a distinct variant of reactive angioendotheliomatosis. The disease typically presents in obese patients who smoke and have atherosclerotic risk factors, vasculopathies, or other comorbidities associated with hypoxemia. We present a case of a 40-year-old woman with ulcerated plaques on her abdomen consistent with diffuse dermal angiomatosis on histopathological evaluation. Unique to our patient was localization of the lesions to the abdominal striae.

KEYWORDS Diffuse dermal angiomatosis; striae

Diffuse dermal angiomatosis (DDA) is a rare, benign, reactive cutaneous vascular disorder.¹ It is associated with tissue ischemia secondary to atherosclerotic disease, vasculopathies, or other comorbidities that can cause tissue hypoxemia. This is theorized to cause up-regulation of endothelial cell production, resulting in the clinical manifestation of DDA.^{2,3} Here, we present a case of DDA with a unique presentation.

CASE DESCRIPTION

A 40-year-old white woman was evaluated for a 2-month history of nonhealing, painful, ulcerated plaques on her abdomen localized within striae. Her past medical history included tobacco use, prediabetes, obesity, chronic obstructive pulmonary disease (COPD), and hypercholesterolemia. She had previously been treated with several courses of systemic antibiotics without improvement. Her symptoms included numbness, tingling, and cramping in her lower extremities, consistent with claudication, and a progressive restriction in her ability to ambulate.

Examination revealed erythematous to violaceous atrophic plaques with central ulcerations occurring within the abdominal striae (*Figure 1*). Imaging studies demonstrated complete occlusion of the distal aorta and bilateral iliac arteries. A punch biopsy taken from one of the lesions revealed diffuse proliferation of monomorphic cells arranged between collagen bundles forming vascular channels with red

blood cell extravasation (*Figure 2*). No vasculitis or occlusion was noted. The cells were positive for CD31 and ERG immunostains and negative for human herpesvirus-8. The patient was diagnosed with DDA and underwent aortobifemoral bypass surgery. Her lesions were healing at her 2-week follow-up.

DISCUSSION

DDA, first described by Krell et al in 1994, is a distinct variant of reactive angioendotheliomatosis. The lesions typically present as livedoid, erythematous to violaceous plaques that may contain a central ulcer or erosion.¹ On histopathology, DDA is characterized by diffuse proliferation of endothelial cells arranged between collagen bundles, which form small vascular channels with scattered extravasated erythrocytes.⁴ The cells may have a spindle-shaped appearance with vacuolated cytoplasm. Clinically, the differential diagnosis may include vasculitis, acroangiodermatitis, and vascular tumors. Histologically, differentials include Kaposi sarcoma and well-differentiated angiosarcoma. DDA is thought to be caused by ischemia or inflammation that creates local hypoxia, leading to an increase in vascular endothelial growth factor. This, in turn, causes endothelial proliferation and neovascularization.^{2,3} The resolution of cutaneous lesions after revascularization of the blocked vessel supports this theory.

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Received March 25, 2020; Revised April 25, 2020; Accepted April 30, 2020.

Figure 1. Erythematous to violaceous atrophic plaques with central ulcerations/erosions occurring within abdominal striae.

Figure 2. Punch biopsy with hematoxylin and eosin revealing proliferation of monomorphic cells arranged interstitially between collagen bundles, forming vascular channels with red blood cell extravasation; no vasculitis or occlusion is noted.

The mean age of patients reported to have DDA is 54.3 years, and 74% are women. The female predominance is due to the overwhelming number of cases involving the breasts, usually associated with subclavian artery occlusion or large pendulous breasts. Macromastia is thought to result in tissue hypoxemia due to subclinical torsion, compression, and increased venous hydrostatic pressure.⁵ Pain is the most common complaint. Following the breast, the next most common location is the lower extremities. Our case is unique due to its involvement in the abdominal striae. Only one other case of DDA involving the abdominal striae has been reported. Although involvement of the buttock and lower

extremity was also reported in that case, these were explained by occlusion of the common iliac artery; involvement of the abdominal striae could not be explained.⁶

The most common associated comorbidity in DDA is smoking (62%), followed by hypertension (53%), peripheral vascular disease (41%), hyperlipidemia (18%), diabetes mellitus (15%), chronic kidney disease (15%), and obesity (12%). Other reported diseases associated with this cutaneous reactive pattern include Takayasu arteritis, calciphylaxis, hepatitis, monoclonal gammopathy, and cutis marmorata telangiectatica congenita.^{2,7-9} There is also a reported association with COPD, which was present in our patient as well. Pulmonary function impairment due to COPD, obstructive sleep apnea, and obesity hypoventilation syndrome may contribute to the tissue hypoxia seen in DDA.⁵

The treatment for DDA focuses on improving tissue hypoxemia with control of atherosclerotic risk factors and revascularization of the occluded artery. Many case reports describe the use of isotretinoin, topical and systemic corticosteroids, and topical and systemic antibiotics, but the results are variable, with many reporting little to no improvement.^{5,10-12} Isotretinoin proved to be effective for some, but recurrence occurred in several patients.¹³⁻¹⁵ Of patients with breast involvement, the treatment that was consistently successful was mastopexy with removal of the affected skin.^{5,16} Some reports have suggested that smoking cessation alone helps improve lesions.⁸ Other therapies attempted without success include hyaluronic acid, silver dressings, astringents, antiviral therapy, topical tacrolimus ointment, pimecrolimus cream, fluticasone propionate, Biafine emulsion, etanercept, and hydroxychloroquine sulfate.^{5,7} Most successful treatments involve revascularization, with resolution of the lesions within a few weeks to months.

The cutaneous manifestations of DDA should prompt imaging and diagnostic studies to find the underlying cause. Surgical intervention is usually curative and should be considered in patients with associated morbidities.

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Avocations

Now Covid 19

Why so many lives were allowed to wilt
 Before they were able to paint landscape.
 Because life became so ordinary
 That humanity lost concern for life.

Why so many blooms were being deplored,
 Before the petals could spread their fragrance.
 Because they were made available to
 The highest bidder on the trading floor.

Why the trees were becoming denuded
 Before the leaf drop of coming autumn.
 Because man's greed had taken heavy toll
 By choking the life, in a greenhouse tent.

The nature fairly implements its will.
 Treats a hovel or the palace alike.
 None of us will escape its brimming rage
 If we fail to obey the nature's rules.

—Amanullah Khan, MD, PhD

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