Case Report

Diffuse dermal angiomatosis in a 53-year-old woman

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Abstract

A 53-year-old woman with a history of arterial hypertension, diabetes mellitus, and morbid obesity presented with abdominal lesions persisting for 3 months, resistant to previous treatment. Physical examination revealed characteristic erythematous-violaceous plaques with serpiginous distribution and central ulceration. Histological examination confirmed diffuse dermal angiomatosis with underlying fat tissue necrosis, a rare association. Diffuse dermal angiomatosis typically affects middle-aged women with cardiovascular risk factors, and treatment involves addressing underlying vascular alterations.

Introduction

Diffuse dermal angiomatosis is a rare, acquired vascular disorder characterized by benign cutaneous proliferation within the spectrum of reactive cutaneous angiomatosis. It primarily affects middle-aged women with significant cardiovascular risk factors and is typically associated with advanced atherosclerosis and chronic tissue ischemia. Clinically, it manifests as erythematous-violaceous plaques, often with ulceration and necrosis, predominantly on the limbs. Histopathologically, diffuse dermal angiomatosis is distinguished by endothelial and pericyte proliferation, requiring careful differentiation from other vascular lesions. We report a patient with diffuse dermal angiomatosis presenting with unique clinical features, emphasizing the importance of early diagnosis and targeted therapeutic strategies.

Case Synopsis

A 53-year-old woman with a history of arterial hypertension, diabetes mellitus, and obesity was admitted for evaluation of abdominal lesions present for 3 months. These lesions had been previously treated with oral antibiotics and topical corticosteroids, resulting in expansion.

Physical examination revealed an erythematous-violaceous indurated plaque with a reticular pattern in the left hypochondrium, extending in a serpiginous distribution across the groin to the left thigh, with a central ulcerated and abscessed area (Figure 1). Notably, the lesion extended within striae to the upper abdomen.

Histopathologic examination of a deep punch biopsy from the ulcer edge revealed vascular proliferation without atypia, extending from the superficial dermis to the deep margin (Figure 2) where only a few small lumina were found. Cluster of differentiation (CD) 31 and CD34 immunostaining demonstrated endothelial differentiation (Figure 3A). D2-40 and human herpesvirus 8 were negative, and smooth muscle actin highlighted the pericytic component, supporting a benign/reactive nature (Figure 3B). Additionally, necrosis of the underlying fat tissue was observed. Computed tomography scan confirmed fat tissue necrosis and atherothrombotic obstruction of both the common and left external iliac arteries, following the same trajectory of the patient's lesions (Figure 4).

Case Discussion

Diffuse dermal angiomatosis is an acquired benign cutaneous vascular proliferation within the spectrum of reactive cutaneous angiomatosis. Five entities in this group share intravascular and extravascular hyperplasia of endothelial cells and pericytes at the histologic level: diffuse dermal angiomatosis, acroangiodermatitis (pseudo-Kaposi syndrome), intravascular histiocytosis, glomeruloid

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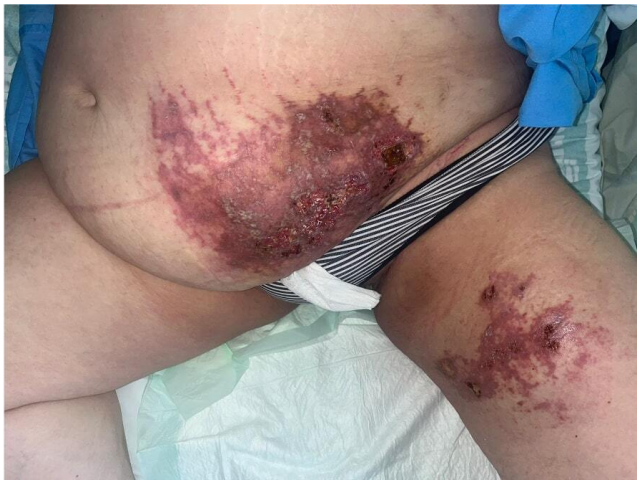

Figure 1. *Erythematous-violaceous indurated plaque with a reticular pattern in the left hypochondrium, extending in a serpiginous distribution across the groin to the left thigh, with a central ulcerated and abscessed area.*

angioendotheliomatosis, and cryoprotein angiomatosis.[1] Key characteristics of these entities are summarized in Table 1.

Diffuse dermal angiomatosis primarily affects middle-aged women with cardiovascular risk factors, especially advanced atherosclerotic disease and smoking, though it may also be associated with systemic diseases such as infections, chronic kidney disease, calciphylaxis, monoclonal gammopathy, or anticardiolipin antibodies.[2,3] Its etiology is thought to result from atheromatous plaques, causing prolonged tissue ischemia and subsequent release of histiocytes and proinflammatory cytokines that promote neoangiogenesis.[1,2] Clinically, it presents as erythematous-violaceous or purpuric plaques, occasionally ulcerated or necrotic, which may appear on any region, including the upper and lower limbs.[2]

Fewer than 100 cases have been reported, most involving the breast region, particularly in patients with gigantomastia or breast dependency, where lesions are often associated with severe pain.[2] Surgical management, including reduction mammoplasty or mastectomy, has proven effective, though isotretinoin or corticosteroids have also been used successfully.[2,4] Fewer than 20 cases have been linked to atherosclerosis, and just 1 prior case described involvement of striae similar to our patient.[5] A comparison with previously reported cases is presented in Table 2.

Histopathologically, diffuse dermal angiomatosis is characterized by diffuse proliferation of endothelial cells and intravascular and extravascular pericytes, primarily in the reticular dermis.[1,5] Differential diagnosis includes acroangiodermatitis, Kaposi sarcoma, and angiosarcoma, for which conventional histology and immunohistochemistry (CD31, CD34, and HHV-8) are essential.[2] Histologic and immunohistochemistry comparisons are summarized in Table 3.

Therapeutic options focus on correcting underlying vascular alteration, with documented resolution following revascularization.[3] Isotretinoin and corticosteroids may also be considered in selected patients to reduce proangiogenic factors.[2]

Conclusion

Diffuse dermal angiomatosis is a rare reactive cutaneous vascular proliferation, predominantly associated with advanced atherosclerosis and ischemia. Diagnosis is confirmed by histopathology, including endothelial and pericyte hyperplasia. Management focuses on correcting underlying vascular abnormalities, with adjunctive therapies such as isotretinoin or corticosteroids to reduce angiogenesis.

Potential conflicts of interest

The authors declare no conflicts of interest.

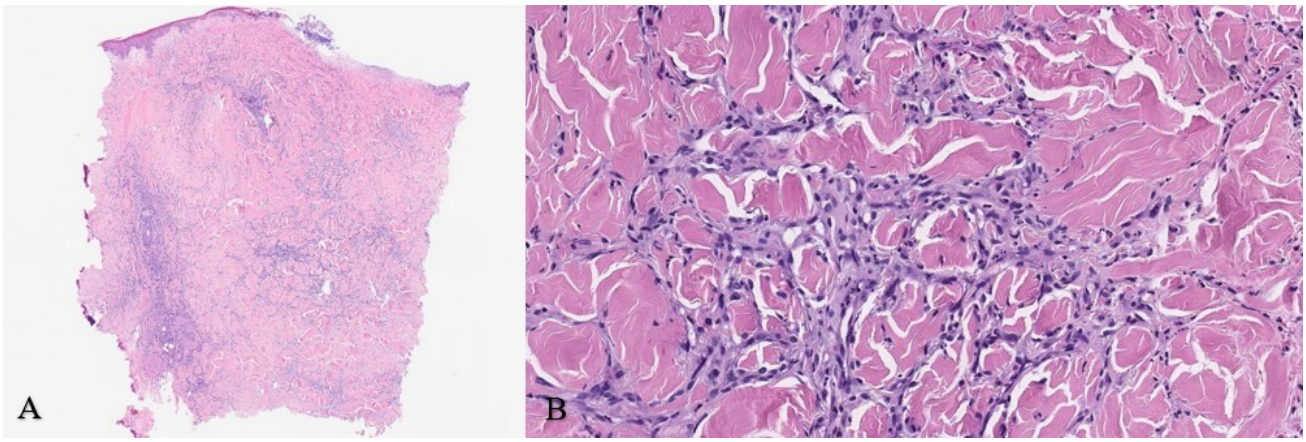

Figure 2. ***(A,B)*** *Punch biopsy showing interstitial cellular proliferation dissecting collagen bundles of the dermis. Spindle cells display significant atypia, monomorphic nuclei and basophilic cytoplasms with indistinct borders. Few erythrocytes and small lumina are barely identifiable (hematoxylin-eosin, original magnification ×200).*

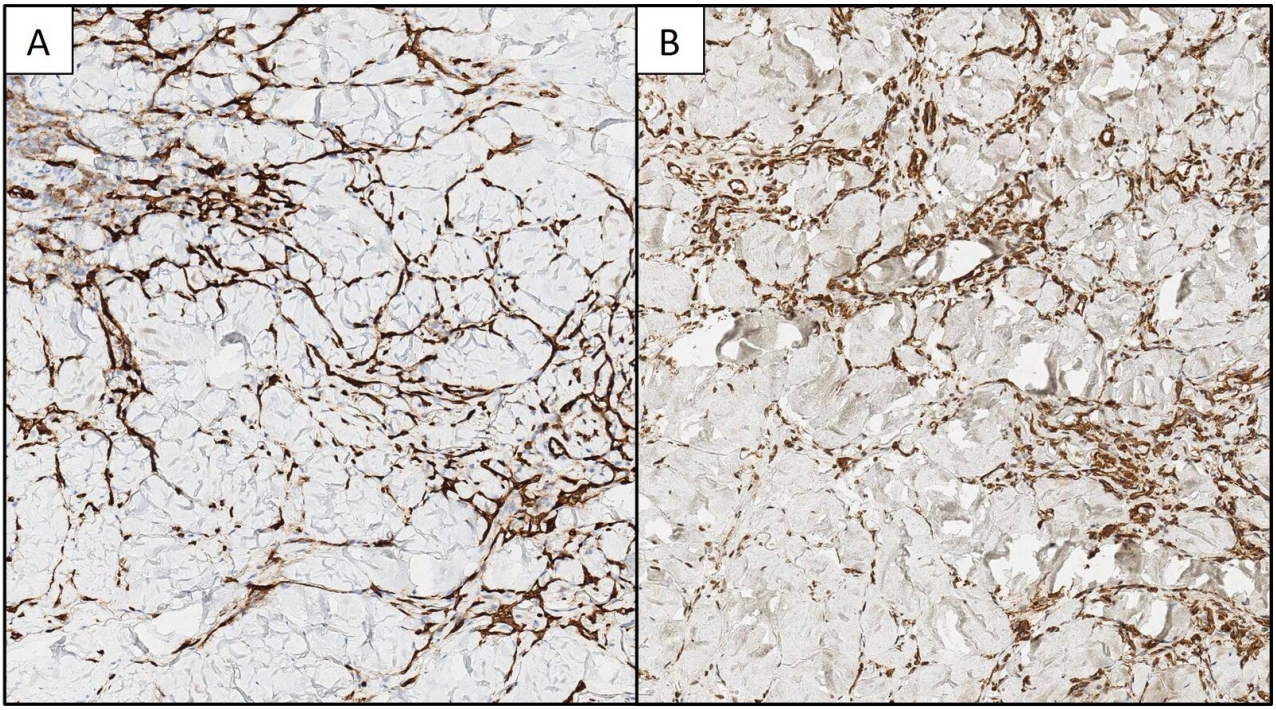

Figure 3. ***(A)*** *Endothelial differentiation of the cell population (cluster of differentiation [CD] 31 immunostaining, original magnification ×100).* ***(B)*** *Smooth muscle actin highlighting the pericytic component surrounding each vascular structure, supporting a benign/reactive nature (CD31 immunostaining, original magnification ×100).*

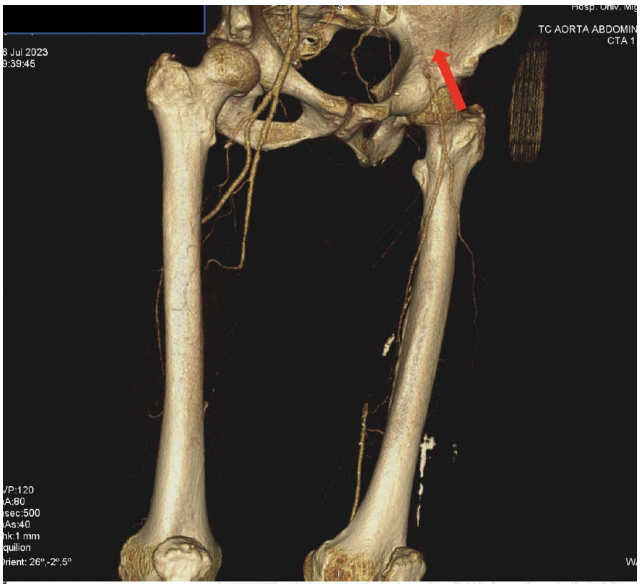

Figure 4. *Three-dimensional reconstruction of computed tomography showing atherothrombotic obstruction in both the common iliac and left external iliac arteries (red arrow).*

Table 1. *Comparison of Conditions Included as Reactive Cutaneous Angiomatoses.*

Condition	Clinical Presentation	Histopathologic Features	Proliferating Cells	Associated Conditions
Reactive angioendotheliomatosis	Erythematous-violaceous plaques; sometimes ulcerated or necrotic	Endoluminal endothelial cell proliferation	Endothelial cells with or without pericytes	Infections (endocarditis), cholesterol emboli, arteriovenous shunt, antiphospholipid syndrome, renal disease, rheumatoid arthritis, monoclonal gammopathy, hepatitis
Diffuse dermal angiomatosis	Ulcerated violaceous plaques; often painful	Interstitial endothelial proliferation throughout the dermis	Endothelial cells	Atherosclerosis, arteriovenous shunt
Acroangiodermatitis (pseudo-Kaposi's sarcoma)	Coalescent red-brown macules or violaceous papules and plaques; primarily on the legs	Lobular proliferation of thick-walled vessels in the papillary dermis	Endothelial cells and pericytes	Venous insufficiency, arteriovenous shunt, limb paralysis, amputation stump, thrombophilia, Klippel-Trénaunay syndrome
Intravascular histiocytosis	Erythematous-violaceous patches and plaques	Intraluminal proliferation with vessel occlusion	Histiocytes	Monoclonal gammopathy, vascular insufficiency, rheumatoid arthritis
Glomeruloid reactive angioendotheliomatosis	Erythematous-purpuric patches and ulcerated necrotic plaques	Intraluminal glomeruloid capillary tufts	Endothelial cells	Cold agglutinins, lymphoma
Angiopericytomatosis (angiomatosis with cryoproteins)	Ulcerated necrotic plaques and erythematous papules	Periluminal proliferation with or without thrombi	Pericytes (with or without histiocytes)	Multiple myeloma, cryoproteinemia

Table 2. *Comparison of Patients With Diffuse Dermal Angiomatosis Affecting Striae.*

Feature	Current Case	Previously Reported Case, Frizzell et al[5]
Age	53 y	50 y
Sex	Female	Female
Medical history	Hypertension, diabetes mellitus, morbid obesity	Tobacco use, prediabetes, obesity, COPD, hypercholesterolemia
Clinical presentation	Erythematous-violaceous indurated plaque with a reticular pattern, extending from the left hypochondrium to the left thigh, with central ulceration and abscess	Painful ulcerated plaques within abdominal striae, non-healing over 2 months
Histopathologic findings	Vascular proliferation without atypia, interstitial pattern with pericytes (SMA+), necrosis of fat tissue	Diffuse proliferation of monomorphic cells forming vascular channels, red blood cell extravasation, CD31+ and ERG+
Imaging findings	CT showed fat tissue necrosis and atherothrombotic obstruction in iliac arteries	Complete occlusion of distal aorta and bilateral iliac arteries
Treatment and outcome	Prior treatment with oral antibiotics and topical corticosteroids, torpid evolution	Aortobifemoral bypass surgery, lesion healing at 2-week follow-up

Abbreviations: CD31, cluster of differentiation 31; COPD, chronic obstructive pulmonary disease; CT, computed tomography; ERG, ets-related gene; SMA, smooth muscle actin.

Table 3. *Comparison of H&E and IHC Staining of Diffuse Dermal Angiomatosis, Acroangiodermatitis, and Angiosarcoma.*

Feature	H&E Staining	IHC Staining
Diffuse dermal angiomatosis	Small capillaries with erythrocytes; mild inflammatory infiltrate (lymphocytes)	CD31+, CD34+, factor VIII+; negative for pancytokeratin
Acroangiodermatitis	Vascular proliferation with neutrophils and extravasated red blood cells	CD31+, CD34+, factor VIII+, SMA+
Angiosarcoma	Irregular large blood vessels, pleomorphic endothelial cells, high mitotic activity	CD31+, CD34+, ERG+, VEGF+, pancytokeratin–

Abbreviations: H&E, hematoxylin-eosin; IHC, immunohistochemistry; CD31, cluster of differentiation 31; CD34, cluster of differentiation 34; factor VIII, antihemophilic factor; SMA, smooth muscle actin; ERG, ets-related gene; VEGF, vascular endothelial growth factor.

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