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## Case 11

### Painful lesion with central necrosis on the thigh

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**Patient 11.1:** A 49-year-old Thai male from Bangkok

**Chief complaint:** Painful lesion on the right thigh for 1 month

**Present illness:**

The patient developed a small painful lesion on right thigh from a minor trauma for 1 month. He had a history of minor trauma prior to the development of the lesion. The lesion gradually increased in size and became ulcerated at the center of lesion, which was extremely painful. He lacks of other systemic symptom.

**Past history:**

- Coronary artery disease S/P percutaneous coronary intervention
- Hypertension, diabetes mellitus type 2, dyslipidemia
- Smoking history of 15 pack years

**Current medications:** Aspirin 81 mg/d, carvedilol 50 mg/d, simvastatin 40 mg/d, glipizide 5 mg/d, hydralazine 50 mg/d, isosorbide dinitrate 20 mg/d

**Skin examination:**

- Solitary, well-defined, irregular, erythematous violaceous, non-blanchable patch diameter 10x12cm, with superficial central ulceration covered by thick hemorrhagic crust

**Physical examination:**

VS: BT 37.4<sup>0</sup> c, BP 140/90 mmHg, HR 80/min, PR 80/min, RR 12/min

HEENT: No pale conjunctivae, no icteric sclerae, no oral ulcer

Heart: Normal S1 S2, no murmur

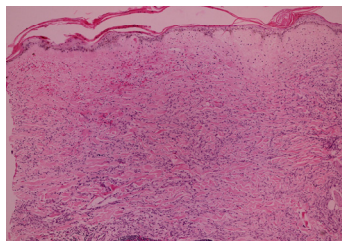
Lung: Normal breath sound

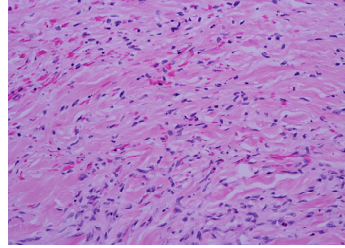
Abdomen: Soft, no hepatosplenomegaly

Neurological system: Intact

Vascular: Absent of femoral, popliteal, posterior tibial, and dorsalis pedis pulses on both sides

Case 11.1





**Histopathology:** (S13-17819, right thigh)

- There is interstitial proliferation of spindle cells in association with extravasated red blood cells and sparse perivascular inflammatory cell infiltration in the dermis
- Immunostains: Positive for CD31 and CD34, but negative for HHV 8

**Investigation:**

Laboratory tests

CBC: Hct 43%, WBC 9,440/mm<sup>3</sup> (N 48% L46% M 5% E 1%),  
Platelet 371,000/mm<sup>3</sup>

Anti-HIV: Negative

ANA: Negative

Imaging study

Computed tomography angiography:

- Total occlusion of right infrarenal aorta, extended to aortic bifurcation, both common iliac arteries, internal, and external iliac arteries

**Diagnosis: Diffuse dermal angiomatosis**

**Treatment:** Axillo-femoral and femoro-femoral bypass surgery

**Patient 11.2:** A 62-year-old Thai male from Bangkok

**Chief complaint:** Painful patch on the left thigh for 3 month.

**Present illness:**

Six months earlier, the patient developed intermittent claudication of both thighs and legs during walking and were quickly relieved at rest, in which the pain was worse on his left thigh. Three months earlier, he developed an extremely painful erythematous patch on his left thigh, which became quickly enlarged and ulcerated. He lacks of other systemic symptom.

**Past history:**

- Coronary artery disease S/P percutaneous coronary intervention
- Hypertension, dyslipidemia
- Smoking history of 20 pack years

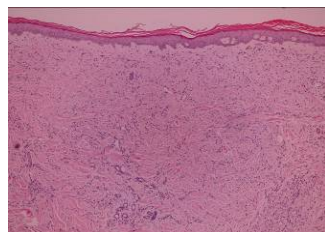
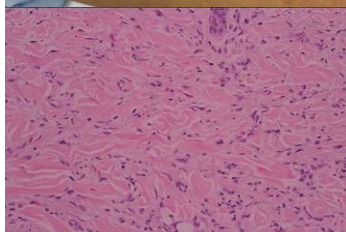
**Current medications:** Aspirin 81 mg/d, beraprost 120 mg/d, cilostazol 100 mg/d, hydralazine 75 mg/d, telmisartan 80 mg/d, manidipine 20 mg/d, atorvastatin 40 mg/d, isosorbide mononitrate 40 mg/d

**Skin examination:**

- Solitary, well-defined, stellate, erythematous violaceous, non-blanchable patch diameter 9x8 cm., with central irregular-border shallow ulcer and hemorrhagic crust



Case 11.2



**Physical examination:**

VS: BT 37.2<sup>0</sup> c, BP 130/80 mmHg, HR 72/min, PR 80/min, RR 10/min

HEENT: not pale conjunctivae, no icteric sclera

Heart: Normal S1 S2, no murmur

Lung: Normal breath sound

Abdomen: Soft, no hepatosplenomegaly

Neurological system: Intact

Vascular: Absent of femoral artery, popliteal artery, posterior tibial artery, dorsalis pedis artery pulses both sides

**Histopathology:** (S14-7778, left thigh)

- Proliferation of vascular lumens with occasional plump endothelial cells
- Immunostains: Positive for CD31 and CD34, but negative for HHV 8

**Investigation:**Laboratory tests

CBC: Hct 40%, WBC 8,940/mm<sup>3</sup> (N 49% L45% M 6%),  
Platelet 231,000/mm<sup>3</sup>

Anti-HIV: Negative

Imaging studyComputed tomography angiography:

- Total occlusion of left distal infrarenal aorta, extended to aortic bifurcation, both common iliac arteries, internal, and external iliac arteries

**Diagnosis: Diffuse dermal angiomatosis**

**Treatment:** Axillo-bifemoral bypass surgery

**Discussion:**

Diffuse dermal angiomatosis (DDA) is a rare benign vascular disorder of the skin and was classified in the group of cutaneous reactive angiomatosis.<sup>1</sup> DDA has been described in association with severe atherosclerotic peripheral vascular disease, arteriovenous fistula, and large pendulous breasts.<sup>2-5</sup>

The pathogenesis of DDA is unclear. It is hypothesized that ischemic process or an inflammatory vascular reaction that generates a localized hypoxic stimulus causing a local increased vascular endothelial growth factor, which induces endothelial proliferation and neovascularization, and the occlusions of the vessel caused by atherosclerotic plaques may be source of emboli to distal, small cutaneous vessels, where they could induce neoangiogenesis.<sup>2</sup>

The clinical presentations of DDA are erythematous, violaceous, livedoid plaques, often with central ulceration. The frequently painful lesions may be solitary or multiple and typically found on the lower extremities in patients with severe vascular atherosclerotic disease. It has also been reported on the forearm secondary to iatrogenic arteriovenous fistulas in chronic hemodialysis patients and in women presenting with non-healing, ulcerating lesions on large breasts. Differential diagnosis of DDA includes vasculopathy, medium to large vessel vasculitis, benign and malignant vascular tumor.

Histologically, DDA is characterized by a diffuse proliferation of endothelial cells interstitially arranged between the collagen bundles within the papillary and reticular dermis. Focal areas of proliferating cells may show a spindled morphology, vacuolated cytoplasm, and formation of small vascular channels, suggesting neoangiogenesis. Scattered extravasated erythrocytes and hemosiderin deposition may also be appreciated.<sup>6</sup> Atypical endothelial mitoses and cytologic atypia are absent. Immunohistochemistry showed positivity of endothelial cells for Ulex europaeus agglutinin-1, factor VIII-associated antigen, CD31, and CD34. Histological differential diagnosis of DDA includes, Kaposi's sarcoma and well-differentiated angiosarcoma, but the frank atypia of cells, the diffuse slit-like lumen formation with the promontory sign and an inflammatory component are lacking.<sup>9</sup>

The treatment of DDA includes cessation of smoking led to a substantial improvement, accompanied by strict control of cardiovascular risk factors, such as hypertension or hyperlipidemia. Systemic steroids and isotretinoin have also been successfully used based on their inhibitory effect on neoangiogenesis.<sup>5</sup> Surgical procedure includes vascular surgery for correcting and bypass the vascular occlusion could reach good results.<sup>7,8</sup>

Both of our patients had undergone the vascular bypass operations, along with tightly control of hypertension, hyperlipidemia and diabetes, which yield the successful results. The erythematous violaceous patches and painful symptoms rapidly disappeared within a week.

In conclusion, DDA should be considered for a red purpuric patch or ulcer that presents subacutely in a patient with either established vascular compromised or risk factor for peripheral vascular disease. The diagnosis can be made by skin biopsy along with the demonstration of ischemic disease in the affected limb.

## References:

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