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CASES FOR DIAGNOSIS

Violaceous Plaque in the Left Leg

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Clinical History

The patient was a 19-year-old male with no past medical history of interest. He consulted for the appearance of a painful, slow-growing lesion on the left pretibial region that had appeared after an injury 2 years earlier.

Physical Examination

On examination, a violaceous plaque of 8 × 6 cm, with well-defined borders, was observed on the distal third of the left pretibial region (Figure 1). The local temperature was not raised and there were no varicose veins.

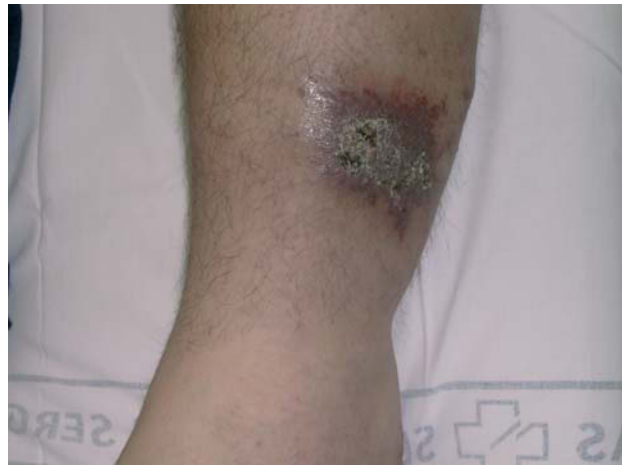

Figure 1.

Complementary Tests

The blood tests including complete blood count, biochemistry, coagulation studies, and serology for human immunodeficiency virus were normal or negative. Echo-Doppler study showed an underlying arteriovenous fistula.

Histopathology

The histopathological study revealed an increase in the number of newly formed capillaries in the superficial and mid dermis. There was a mild chronic inflammatory infiltrate around this vascular proliferation, and hemosiderin deposits in the deep dermis. The overlying epidermis was normal (Figure 2).

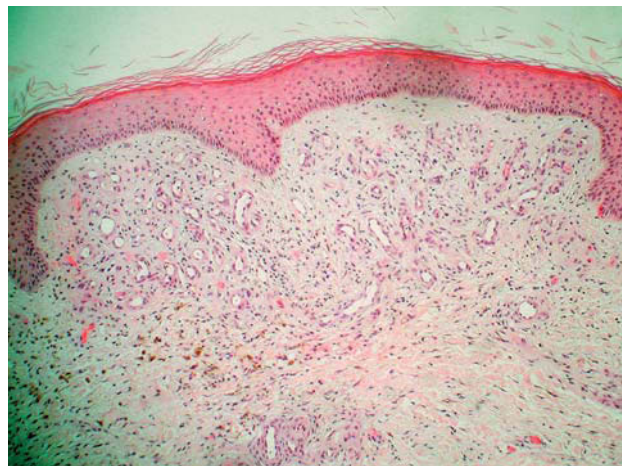

Figure 2. Hematoxylin-eosin, ×100.

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What Was the Diagnosis?

Diagnosis

Acroangiodermatitis (Stewart-Bluefarb syndrome).

Clinical Course and Treatment

The patient was referred to the Vascular Surgery Department for treatment of the arteriovenous fistula.

Discussion

Acroangiodermatitis, also known as pseudo-Kaposi sarcoma, is an unusual condition characterized by slow-growing macules, papules, or violaceous plaques most commonly found on the lower limbs.[1] It is separated into 2 variants: Mali acroangiodermatitis,[2,3] which is associated with chronic venous insufficiency in elderly individuals and which usually affects the distal parts of both legs; and Stewart-Bluefarb syndrome,[4,5] which is secondary to the presence of an arteriovenous fistula that develops in young individuals and usually only affects one limb.

Similar lesions have been reported in areas of skin distal to an arteriovenous fistula in patients with chronic renal failure on hemodialysis, in paralyzed limbs, in the skin of an amputation stump,[6] in patients with Klippel-Trenaunay syndrome,[7] in many other conditions,[8] and even in healthy individuals.

The disorder has a characteristic histopathology, with an increase in the number of capillaries, which have thick walls, extravasation of red blood cells, and hemosiderin deposits in the papillary dermis in the case of Mali acroangiodermatitis and throughout the dermis in the Stewart-Bluefarb syndrome.

The etiology is unknown. It appears that the anoxia induced by changes in blood flow leads to a proliferation of endothelial cells and fibroblasts. However, alterations of blood flow are very common and acroangiodermatitis is a relatively rare condition. It has therefore been postulated that other mechanisms, such as the action of prostaglandin E or exogenous microtrauma to the limb, could play a role in the pathogenesis of the disease.[2]

Knowledge of this condition is essential to avoid incorrect treatments. This is based on the underlying pathology: measures that increase venous return are sufficient in venous insufficiency; embolization or surgical excision may be required in the case of arteriovenous fistulas, though these treatments are not risk-free. Laser therapy may be used in certain cases.[1,2]

The main differential diagnosis is with Kaposi sarcoma. From a histological point of view, Kaposi sarcoma presents an irregular proliferation of slit-like blood vessels; these vessels are lined by flattened, fusiform endothelial cells and tend to respect the papillary dermis. Immunohistochemistry with CD34 stains the endothelial cells and perivascular fusiform cells diffusely throughout the dermis in contrast to pseudo-Kaposi sarcoma in which the pattern of uptake is limited to the vessels.

Other differential diagnoses include multinucleate cell angiohistiocytoma, lymphangioendothelioma, progressive pigmentary dermatosis, stasis dermatitis, and vasculitis.[2]

Conflicts of Interest

The authors declare no conflicts of interest

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