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Non-healing ulcer on the foot: early onset unilateral Mali-type acroangiodermatitis

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Abstract

Acroangiodermatitis (pseudo-Kaposi's sarcoma, AAD) is a benign vascular dermatosis that resembles Kaposi's sarcoma clinically and histopathologically (1). Four types have been defined: the Stewart–Bluefarb type accompanying chronic arteriovenous malformations, the Mali type accompanying stasis dermatitis, a type accompanying the first gestation, and a type accompanying arteriovenous shunts in patients with chronic kidney failure (3). Although AAD development is associated with chronic venous failure, less frequently AAD can develop as a complication of extremity paralysis, hemodialysis, post-traumatic arteriovenous fistula, amputated extremities, and vascular malformations (e.g., Klippel–Trénaunay syndrome). Pseudo-Kaposi's sarcoma can be histopathologically and clinically confused with malignant diseases such as Kaposi's sarcoma (1, 4). A 22-year-old male was referred to our outpatient clinic with a complaint of a non-healing wound on the distal phalanx of the left first toe. The patient was referred to various centers for 2 years and stated that he had received infection treatments but that his complaints did not disappear. An AAD diagnosis was established for the patient based on clinical and histopathologic evidence. Because he had early-onset disease and it was unilateral, the diagnosis was delayed. In addition, due to the rare occurrence of the disease, we histopathologically diagnosed this patient as having acroangiodermatitis.

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Introduction

Acroangiodermatitis (pseudo-Kaposi's sarcoma, AAD) is a benign vascular dermatosis that clinically and histopathologically resembles Kaposi's sarcoma (1). Clinically, it is characterized by violaceous or reddish-brownish macules, papules, plaques, and/or nodules (1, 2). Four types have been defined: the Stewart–Bluefarb type accompanying chronic arteriovenous malformations, the Mali type accompanying stasis dermatitis, a type accompanying the first gestation, and a type accompanying arteriovenous shunts in patients with chronic kidney failure (3). Although AAD development is associated with chronic venous failure, less frequently AAD can develop as a complication of extremity paralysis, hemodialysis, post-traumatic arteriovenous fistula, amputated extremities, and vascular malformations (e.g., Klippel–Trénaunay syndrome). Pseudo-Kaposi's sarcoma can be histopathologically and clinically confused with malignant diseases such as Kaposi's sarcoma (1, 4).

Case

A 22-year-old male was referred to our outpatient clinic with a complaint of a non-healing wound on the distal phalanx of the left first toe. The patient was referred to various centers for 2 years and stated that he had received infection treatments and that his complaints did not disappear. A yellowish-brown crust on the distal phalanx of the first toe was observed upon dermatological examination (Fig. 1a). When the crust was lifted for biopsy, a hemorrhagic nodule with a 2 cm diameter formed by the fusion of the livid-red papules was observed (Fig. 1b). No fading was observed when diascopy was applied to the nodule. Pulses from the distal part of the foot and popliteal pulses were taken. In the histopathologic evaluation, multiple foci of vascular proliferation were seen in the upper dermis. The vessels were small with plump endothelial cells. There were hemosiderin-laden macrophages and extravasated erythrocytes around the vessels. Fibroblastic activity was prominent (Fig. 2a, b). A diagnosis of AAD was established according to the morphologic and clinical features. In the medical history of the patient, there was a history of thrombophlebitis on the left leg during the period that he received treatment due to pulmonary tuberculosis in 2008. At the same time, the patient stated that his symptoms were ameliorated by enoxaparin treatment. Grade 2 to 3 bilateral failure was observed in a venous Doppler ultrasound examination of the bilateral lower extremity. An HIV test was negative. His routine laboratory parameters were unremarkable. Leg elevation and an elastic bandage were recommended to the patient as treatment. Two cycles of cryotherapy were administered to the vascular lesions with a 2-week break. Significant recession in the lesions was observed 1 month later (Fig. 1c).

Discussion

AAD is a rare dermatosis that occurs due to the reactive proliferation of the small vessels. Chronic venous failure can occur with syndromes that involve arteriovenous fistulas, such as congenital or acquired (traumatic or iatrogenic) arteriovenous fistulas, Prader–Labhart–Willi syndrome, and Klippel–Trénaunay syndrome (5). Stewart–Bluefarb syndrome is characterized by congenital arteriovenous malformation of the lower extremity accompanied by multiple arteriovenous shunts and an early onset age. Stewart–Bluefarb syndrome usually occurs unilaterally, it is observed at the lower extremities, and it begins with painful, livid papules and plaques. Mali-type acroangiodermatitis is usually accompanied

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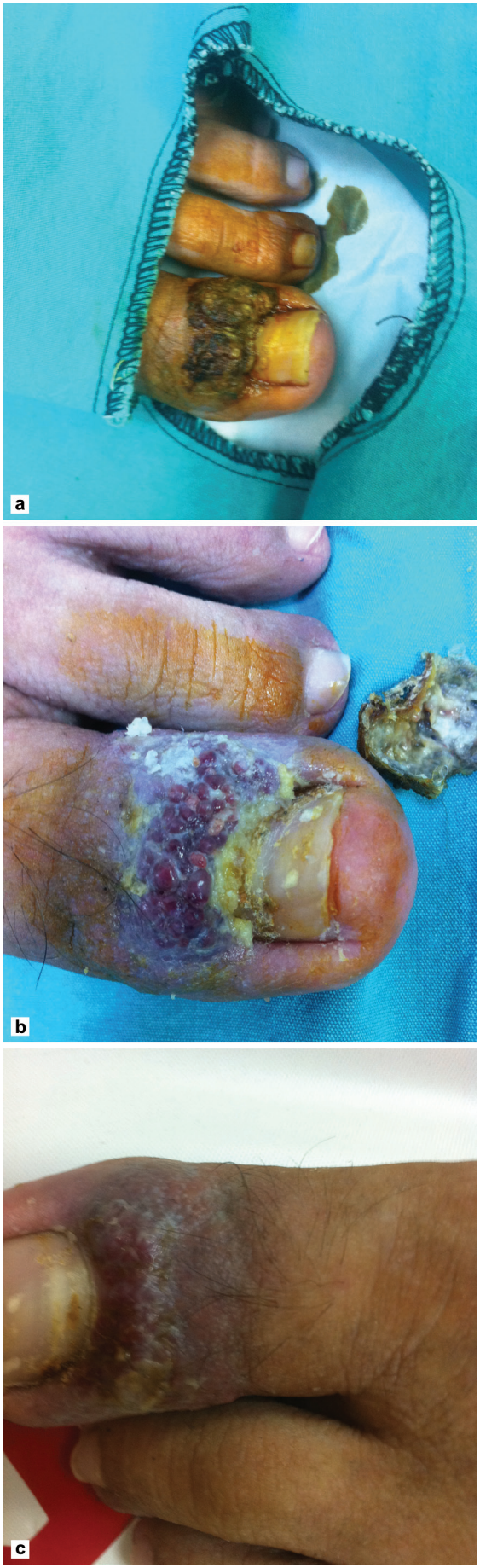

Figure 1 | a) Clinical presentation before the crust was removed; b) Clinical presentation after the crust was removed; c) Clinical presentation after treatment.

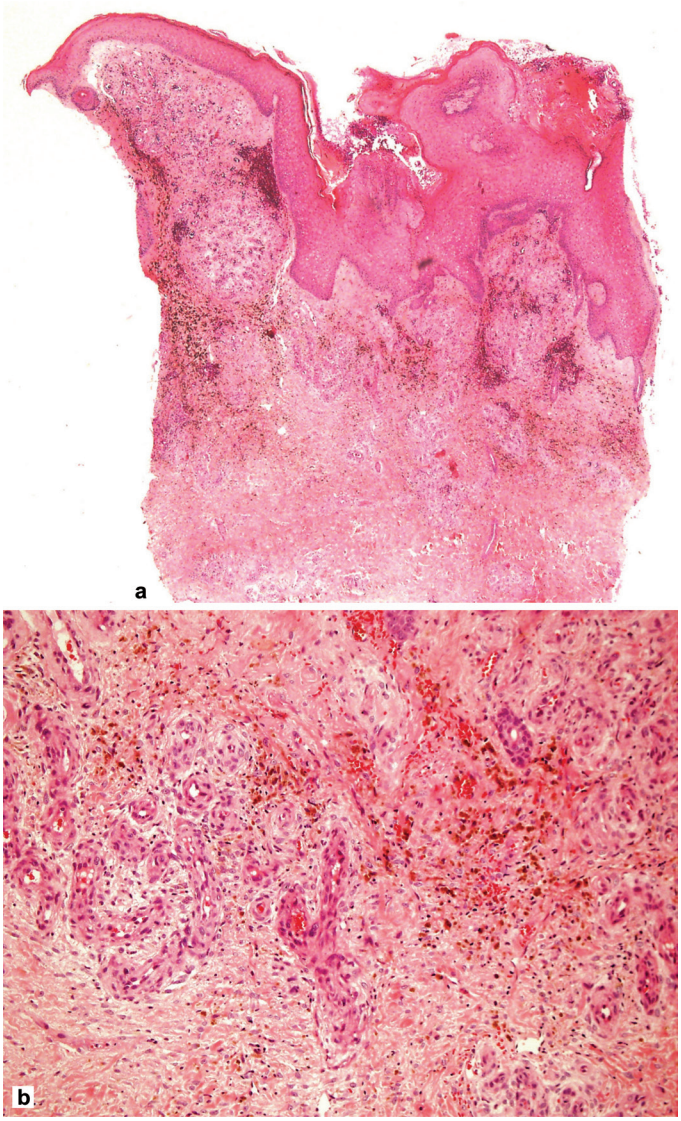

Figure 2 | a, b) Vascular proliferation on the papillary and reticular dermis, swelling of vascular endothelial, hemosiderin-laden macrophages, extravasated erythrocytes, increased fibroblastic activity (a: H&E, × 40, b: H&E, × 100).

by venous failure in elderly people and frequently occurs bilaterally on the foot dorsum, the hallux, the second toe, or the medial of the lower extremity. Mali-type acroangiodermatitis begins in the form of violaceus macules and patches, and it can transform into painless papulonodules or persistent plaques. Our patient had a case of Mali-type acroangiodermatitis beginning at a younger age in contrast to the most frequent cases, and it occurred unilaterally. In his medical history, he showed no symptoms other than tuberculosis and thrombophlebitis. Doppler ultrasonography detected venous failure. The type of AAD observed in the first gestation is accompanied by venous varicosities, appears on the dorsal part of the foot, and occurs in patients with chronic kidney failure and in those with arteriovenous shunts (3). Acroangiodermitis can be histopathologically and clinically confused with Kaposi's sarcoma. However, lack of jagged outline of the vessels, absence of promontory sign, and plump endothelial cells without atypia were the microscopic features that favored a diagnosis of AAD. The histopathology of AAD demonstrates similarities with chronic venous failure. Pigmented purpuric dermatosis, vasculitis, and lichen planus are other diseases that can be confused with AAD. In treatment of the Mali type, conservative approaches such as leg elevation, elastic bandage, protection from trauma,

and the use of antibiotics are recommended; if vascular malformation exists, surgery is required (7). Cases in which topical hydrocortisone, oral erythromycin, and dapsone treatment were effective have been reported (8–11). We obtained good results with application of cryotherapy, along with leg elevation and elastic bandage treatments, to papulonodular lesions. AAD is a rarely observed disease. Provided that an early diagnosis is established, AAD progression can be stopped with conservative treatments. Because AAD can be confused with malignant diseases such as Kaposi's sarcoma, and due to the differences in their treatments, a histopathologic diagnosis is required. We presented this case because of the unilaterally early onset of disease, delayed diagnosis, and rarity of the disease.

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