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CASE REPORTS

Bilateral Pseudo-Kaposi Sarcoma in Upper Limbs

R Fernández,[a] MM Verea,[a] W Martínez,[a] MT Yebra-Pimentel,[b] and E Fonseca[a]

[a]Servicios de Dermatología, and [b]Anatomía Patológica, Complejo Hospitalario Universitario Juan Canalejo, La Coruña, Spain

Abstract. Acroangiodermatitis or pseudo-Kaposi sarcoma is an angioproliferative, self-limited entity that includes a group of diseases, congenital or acquired, with cutaneous lesions similar to Kaposi sarcoma (KS). This term can lead to confusion because it comprises several entities that are completely different, nonetheless, it has an important clinical value as it guides the diagnosis and management of these patients. We report the case of a 67-year-old patient with lesions of acroangiodermatitis in both forearms secondary to arteriovenous shunts from hemodialysis. Doppler ultrasound showed a former arteriovenous fistula in addition to the one already known. Immunohistochemical study showed CD34+ staining in endothelial cells and absence of HHV-8 expression.

Key words:pseudo-Kaposi sarcoma, acroangiodermatitis, arteriovenous fistula.

PSEUDOSARCOMA DE KAPOSI BILATERAL EN MIEMBROS SUPERIORES

Resumen. El término acroangiodermatitis o pseudosarcoma de Kaposi (PSK) se refiere a una entidad angioproliferativa autolimitada que incluye un grupo de enfermedades, congénitas o adquiridas, con lesiones cutáneas similares al sarcoma de Kaposi (SK). Este término puede llevar a confusión puesto que engloba varias entidades completamente diferentes, sin embargo, tiene un valor clínico importante, ya que orienta al diagnóstico y al manejo de estos enfermos. Presentamos el caso de un paciente de 67 años con lesiones de acroangiodermatitis en ambos antebrazos secundarias a fístulas arteriovenosas para hemodiálisis. La ecodoppler permitió descubrir una fístula arteriovenosa antigua, además de la que ya conocíamos. Las técnicas de inmunohistoquímica demostraron CD34 + en las células endoteliales y la expresión del herpesvirus 8 humano (HHV8) fue negativa.

Palabras clave: pseudosarcoma de Kaposi, acroangiodermatitis, fístula arteriovenosa.

Introduction

The term pseudo-Kaposi sarcoma was first used in French sources in 1969 to describe lesions clinically and histologically similar to Kaposi sarcoma in a patient.[1] It was later used by Earhart, in 1974, in the English scientific literature.[2]

Acroangiodermatitis or pseudo-Kaposi sarcoma is an angioproliferative, self-limiting entity that includes a group of diseases, congenital or acquired, with cutaneous lesions resembling those of Kaposi sarcoma (KS). It has been described in relation to chronic venous insufficiency (Mali type),[3] arteriovenous malformations due to congenital or iatrogenic causes (Stewart Bluefarb type),[4] and other vascular conditions such as damage to veins due to amputation, badly fitting protheses,[5] impaired vasomotor function in paralyzed limbs, or prothrombin 20210A mutation.[6]

Pseudo-Kaposi sarcoma lesions due to arteriovenous fistulas caused by hemodialysis are well documented, and we must firstly rule out KS immediately before attempting to identify a treatable cause.

Case Description

The patient was a 67-year-old man examined by our service in November 2001 for asymptomatic violaceous-colored macular and papular lesions, measuring 3-6 mm in diameter, with onset 4 months earlier. These were located on one forearm with a known arteriovenous fistula, and also on the other forearm, where we were unaware of the presence of another arteriovenous fistula (Figure 1). When resident in the United Kingdom, the patient had developed chronic renal insufficiency secondary to reflux nephropathy, rendering hemodialysis necessary. In 1989 he received a kidney transplant, but arterial stenosis of the transplanted organ led to graft loss. In July 2001, now resident in Spain, he

Correspondence:
Rosa Fernández
Servicio de Dermatología. Hospital Abente y Lago
C/ Sir John Moore, s/n
15001 A Coruña, Spain
rosaftorres1980@hotmail.com

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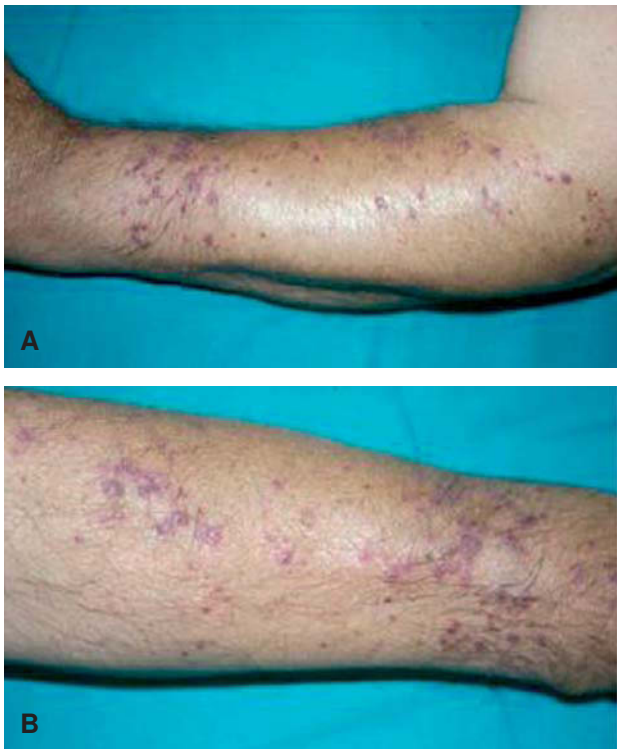

Figure 1. Dark-violaceous colored macular and papular lesions, with confluence to form plaques on the left (a) and right (b) forearms.

began hemodialysis again, and an arteriovenous brachiocephalic fistula occurred in the right forearm.

Blood tests were normal; he was negative for human immunodeficiency virus and hepatitis B and C.

Two biopsies were taken a year apart, and both showed similar changes, consistent with the neoformation of vessels in the papillary dermis and mid dermis, some with capillary morphology and vascular rifts (Figure 2). The endothelial cells lining the vessels showed no abnormalities and extravasated erythrocytes and macrophages were present, along with hemosiderin deposits in the dermis, and inflammatory infiltration of lymphocytes, histiocytes, eosinophils, and some plasma cells (Figure 3). Endothelial cells were CD34+ but perivascular cells were CD34-. There was an absence of human herpes virus 8 (HHV-8) expression.

Doppler ultrasound showed an arteriovenous fistula in the right forearm, with no abnormal flow and another partially embolized fistula in the left forearm.

Discussion

Diagnosis in our patient was made difficult by the presence of bilateral lesions, as we were unaware that a previous arteriovenous fistula had been embolized outside Spain.

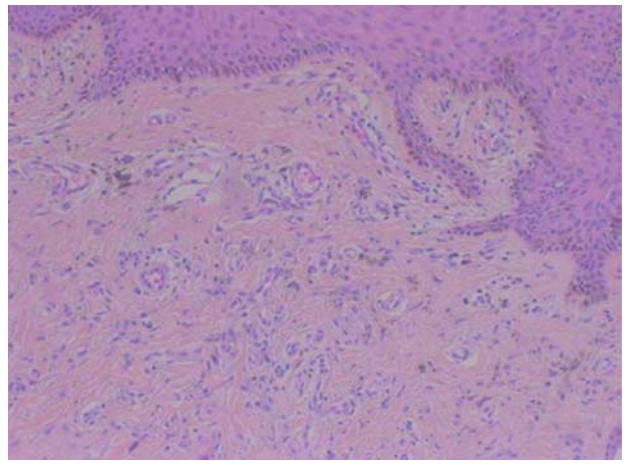

Figura 2. Neoformation of vessels in papillary dermis and mid dermis, some with capillary morphology and others with vascular clefts (Hematoxylin-eosin, ×40).

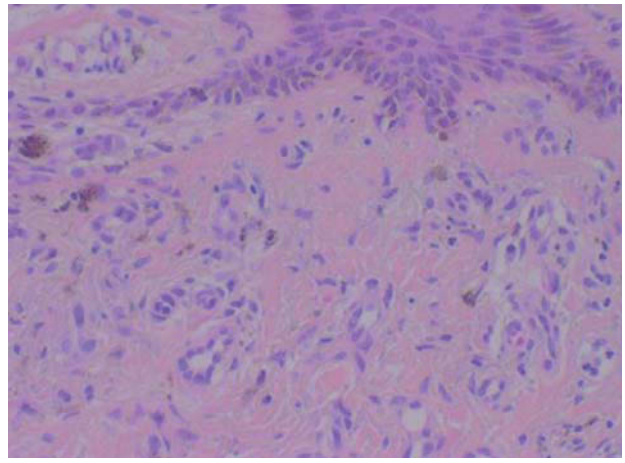

Figura 3. Prominent endothelial cells and hemosiderin deposits. Inflammatory infiltrate of lymphocytes, eosinophils, plasma cells, and histiocytes (Hematoxylin-eosin, ×200).

Pseudo-Kaposi sarcoma lesions resemble true KS, both clinically and histologically. Both are characterized by a proliferation of blood vessels, extravasated erythrocytes, and hemosiderin deposits. Histological differences, however, can help to distinguish them, including the presence of abnormal cells in KS and the low density of inflammatory infiltrate in pseudo-Kaposi sarcoma, which is also low in plasma cells. Likewise, pseudo-Kaposi sarcoma does not present irregular vascular clefts between collagen layers coated by endothelial cells with abnormal nuclei and mitoses, as occurs in true KS.[7]

Despite histological differences, it can be difficult to differentiate the two,[8] hence new diagnostic histopathological techniques have been developed.

Both vascular endothelial cells and stromal spindle cells of KS express the CD34 marker, while pseudo-Kaposi

sarcoma only tests positive for endothelial cells in the dermal capillaries, but not in the surrounding stroma.[9] Also, the HHV-8 expression is only found in KS, and the factor 8 antigen is negative in KS lesions and positive in pseudo-Kaposi sarcoma.

Radiological techniques can help to distinguish this entity from other arteriovenous malformations: ultrasound, Doppler ultrasound, computerized tomography, magnetic resonance imaging, angiography, and phlebography.[10]

Differential diagnosis is important due to the different biological behavior and treatment of the 2 entities. While pseudo-Kaposi sarcoma is limited to the skin and has an indolent course, KS can spread to other organs, and requires a more aggressive treatment.

The pathogenic mechanisms are not completely understood. Both in chronic venous insufficiency and in arteriovenous shunts, elevated vein and capillary pressure occurs with edema, in turn stimulating a proliferation of endothelial cells and fibroblasts. Distal hypoxia can also induce endothelial proliferation through a local increase of endothelial vascular growth factor.[11,12] In other cases, a minor trauma has been considered responsible for provoking the skin lesions.

Treatment tends to be conservative: elevation of the affected limb, surgical treatment of the arteriovenous fistula, sclerotherapy of arteriovenous malformations, etc. Correction of the arteriovenous fistula diminishes skin lesions and helps to prevent progression.

Conflicts of interest

The authors declare no conflicts of interest.

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