Actas Dermosifiliogr. 2007;98:545-8

CASE REPORTS

Stewart-Bluefarb Syndrome

L Hueso,[a] B Llombart,[a] A Alfaro-Rubio,[a] C Serra-Guillén,[a] C Requena,[a] M González,[b] B Cano,[b] E Nagore,[a] O Sanmartín,[a] R Botella-Estrada,[a] and C Guillén[a]

[a]Servicio de Dermatología and [b]Servicio de Radiodiagnóstico, Instituto Valenciano de Oncología, Valencia, Spain

Abstract. Stewart–Bluefarb syndrome is a rare condition involving skin lesions that share clinical features with Kaposi sarcoma and that are secondary to an underlying arteriovenous fistula. We report the case of a 24-year-old man with progressive growth of skin lesions on the lower third of his left leg. Diagnosis of Stewart–Bluefarb syndrome was confirmed histologically and with detection of an arteriovenous fistula.

Key words: pseudo-Kaposi sarcoma, Stewart–Bluefarb, arteriovenous fistula, Kaposi sarcoma.

SÍNDROME DE STEWART-BLUEFARB

Resumen. El pseudosarcoma de Kaposi tipo síndrome de Stewart-Bluefarb es una entidad poco frecuente que presenta lesiones cutáneas semejantes clínicamente al sarcoma de Kaposi, secundarias a una comunicación arteriovenosa subyacente. Presentamos el caso de un varón de 24 años con unas lesiones cutáneas de crecimiento progresivo en el tercio distal de su pierna izquierda. La histología y la demostración de una comunicación arteriovenosa patológica confirmaron el diagnóstico de síndrome de Stewart-Bluefarb.

Palabras clave: pseudosarcoma de Kaposi, Stewart-Bluefarb, comunicación arteriovenosa, sarcoma de Kaposi.

Introduction

Acroangiodermatitis or pseudo-Kaposi sarcoma are synonymous terms encompassing 2 different conditions: acroangiodermatitis of Mali and Stewart–Bluefarb syndrome. Their name arises from the clinical and histological similarities both entities share with classic Kaposi sarcoma.[1]

Acroangiodermatitis of Mali is a common entity that mainly develops bilaterally on the limbs of elderly patients as a result of chronic venous insufficiency and is an extreme form of stasis dermatitis.[2] Stewart–Bluefarb syndrome, however, is an uncommon condition that mainly affects the limbs of young patients unilaterally secondary to an underlying arteriovenous malformation.[3,4]

We present the case of a 24-year-old man with progressive lesions on the distal third of the left leg. Histology and the demonstration of an arteriovenous fistula confirmed the diagnosis of Stewart–Bluefarb syndrome.

Correspondence:
Luis Hueso Gabriel
Servicio de Dermatología
Instituto Valenciano de Oncología
Profesor Beltrán Báguena, 8
46009 Valencia, Spain
luishueso@msn.com

Accepted for publication October 27, 2006.

Case Description

A 24-year-old man was referred to our department for pulsed dye laser treatment of a cutaneous lesion on the left leg that had previously been diagnosed as a vascular malformation. The patient had a small, brown papule on the internal aspect of the left ankle that appeared at the age of 2 and grew progressively and increased in volume. Examination revealed a well-delimited brownish violaceous tumor-like lesion measuring 4 × 5.5 cm and 2 similar lesions that were smaller and less raised on the dorsum of the foot and on the left calf (Figures 1 and 2). Magnetic resonance imaging of the area ruled out bone and tendon involvement. Routine laboratory workup was normal, as was serology for the human immunodeficiency virus. Based on clinical suspicion of pseudo-Kaposi sarcoma, biopsy and Doppler ultrasound were carried out. Histology revealed the presence of multiple thick-walled capillaries covered by diffuse globular cells. These structures were distributed diffusely throughout the papillary dermis and formed lobular structures in the reticular dermis, and were accompanied by red-cell extravasation and

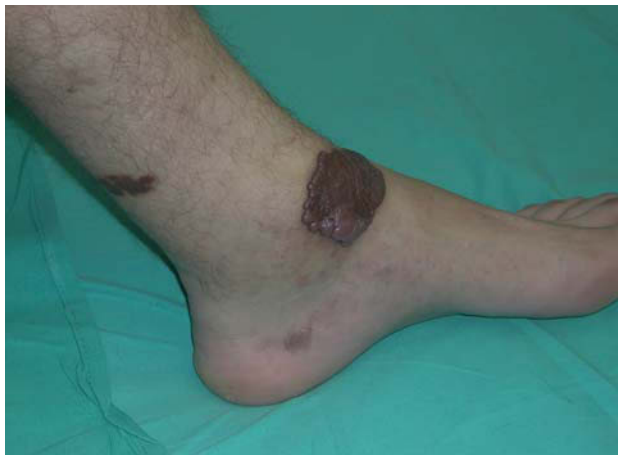

Figura 1. Papular and tumor-like lesions on the distal third of the leg.

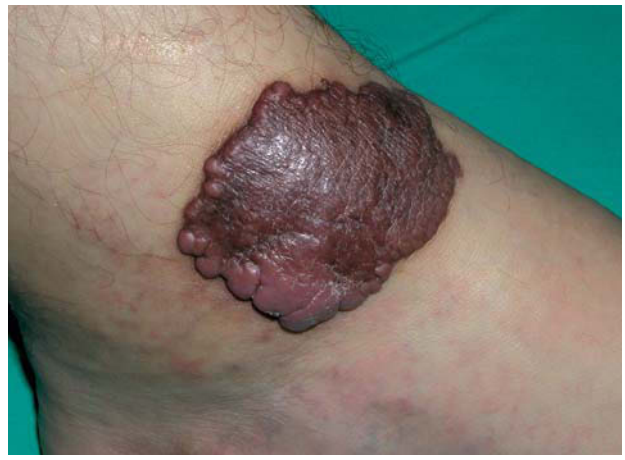

Figure 2. Detail of the largest tumor-like lesion on the left leg.

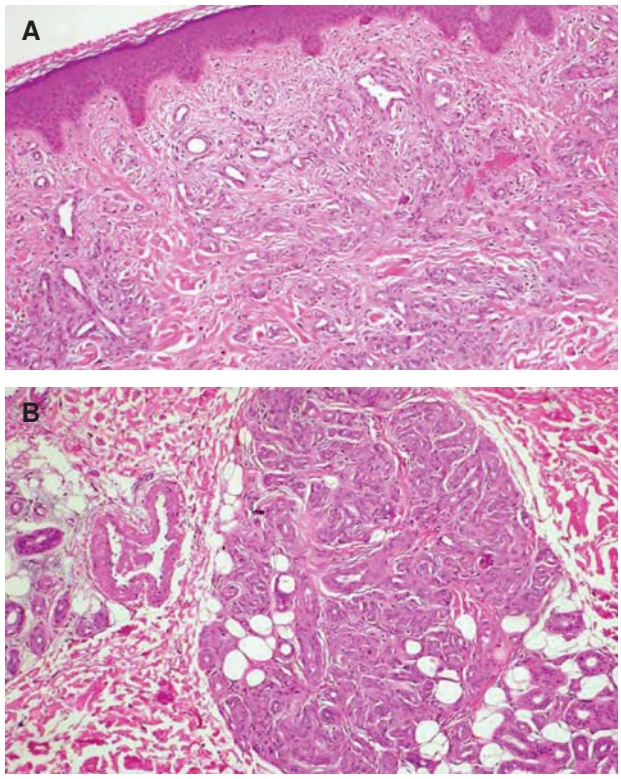

Figure 3. Proliferation of capillaries throughout the dermis. A, Diffusely distributed capillaries in the superficial dermis (hematoxylin-eosin, ×10). B, Lobular formations of capillaries in the middle dermis (hematoxylin-eosin, ×20).

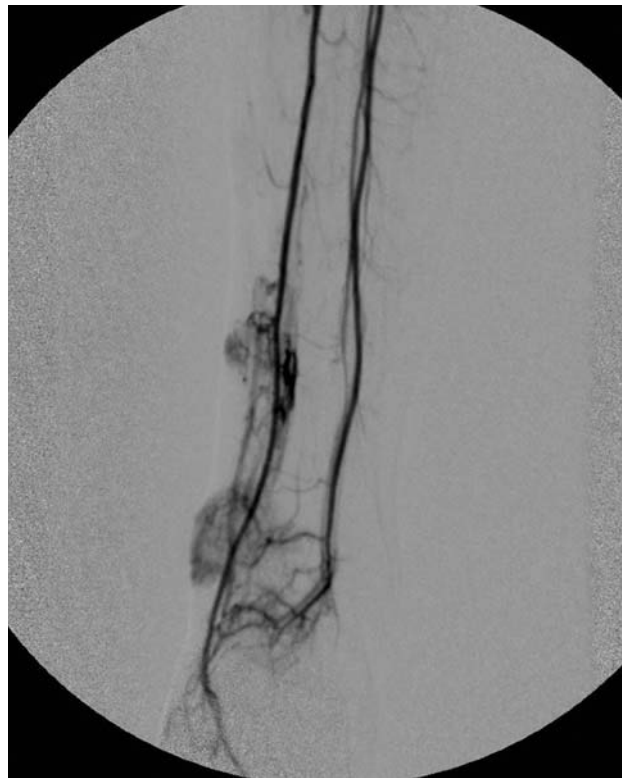

Figure 4. Arteriogram showing arteriovenous fistulas in the distal third of the left leg.

hemosiderin deposits (Figure 3). Doppler ultrasound revealed an underlying arteriovenous fistula, with low flow resistance and long diastoles in the popliteal artery afferent to the malformation, and an arterialized venous flow in a venous collector. Subsequent angiography revealed 2 affected areas on the distal third of the leg, in both the arterial phase and the venous phase. The larger and more distal was fed by 2 branches of the anterior tibial artery, whereas the smaller and more proximal was fed by multiple small branches of the posterior tibial artery (Figure 4).

Demonstration of this arteriovenous fistula confirmed the diagnosis of pseudo-Kaposi sarcoma (Stewart–Bluefarb type).

The absence of symptoms and the possible complications of surgery to correct the arteriovenous fistula meant that conservative measures (hygiene and rest) were taken in this limb along with programming of clinical follow-up visits. The lesions remained stable after 1 year of follow-up.

Discussion

Pseudo-Kaposi sarcoma is an unfortunate term that makes reference to the clinical and histological similarities between 2 different entities (acroangiodermatitis of Mali and Stewart–Bluefarb syndrome) and classic Kaposi sarcoma. The term was coined by Earhart et al[1] in 1974. In 1965, Mali et al[2] described kaposiform lesions in patients with chronic vascular insufficiency. In 1967, independently and almost simultaneously, Bluefarb and Adams[3] and Stewart[4] described similar lesions in a patient with an arteriovenous malformation.

Acroangiodermatitis of Mali generally appears in elderly patients with chronic vascular insufficiency. It usually affects the distal third of both limbs, initially as violaceous macules before progressing to papules and nodules in the setting of stasis dermatitis.[2]

Stewart–Bluefarb syndrome is an uncommon entity that usually occurs in young patients who present an underlying arteriovenous malformation. It generally appears unilaterally on the dorsum of the foot, on the ankle, and on the calf. The lesions include brown or violaceous plaques and macules that can grow progressively and develop verrucous lesions and ulcers. There may also be edema, local temperature increase, soft-tissue hypertrophy, varices, stasis changes, and hypertrichosis.[3,4]

There have been occasional reports of pseudo-Kaposi sarcoma associated with iatrogenic arteriovenous fistulas in dialysis patients,[5] in Klippel–Trenaunay–Weber syndrome,[6] in paralyzed limbs,[7] and in amputation stumps.[7,8]

Arteriovenous fistula may be clinically suspected during examination on palpation of a thrill, auscultation of a murmur, or detection of asymmetrical arterial pulses. Doppler ultrasound is a harmless and highly sensitive screening method, and arteriography is the complementary examination of choice in the case of a well-founded suspicion of arteriovenous fistula. The most typical angiographic sign is early venous filling, which is proportional to the length of the fistula.[9] Radioisotope scanning is less invasive than arteriography.[10]

The histological descriptions of published cases are not very specific and refer to a proliferation of capillaries in the superficial and middle dermis accompanied by fibroblasts, extravasated red cells, and hemosiderin deposits.[11-15] In acroangiodermatitis of Mali, these changes are limited to the superficial dermis. The absence of proliferation of spindle-shaped $CD34^+$ cells and of vascular slits, which are typical of Kaposi sarcoma, can help in the differential diagnosis with this condition.[16] In pseudo-Kaposi sarcoma, unlike classic Kaposi sarcoma, human herpes virus type 8 is not detected and the endothelial cells are positive for factor VIII.[17]

The pathogenesis of Stewart–Bluefarb syndrome is not well understood. It has been postulated that the increase in venous pressure resulting from the arteriovenous malformation may stimulate the proliferation of endothelial cells. A recent report suggested that the arteriovenous steal syndrome with distal ischemia may induce a local increase in vascular endothelial growth factor, which would lead to endothelial proliferation.[18] Similarly, the role of mast cells in the proliferation of endothelial and perivascular cells under conditions of ischemia has also been suggested.[19]

Treatment is usually conservative—compression, elevation of the limb, and care of ulcers, infections, and other complications. The ideal treatment is resolution of the underlying vascular malformation, although this is not often possible due to the common existence of very distal arteriovenous fistulas. Surgery can lead to ulceration or other complications and only resolves macroscopically detectable fistulas. Amputation may occasionally be necessary. Surgery is indicated in cases involving functional impotence, refractory pain, recurrent infection, bleeding, or cardiac decompensation.[13,14] Selective embolization with different particles (Gelfoam, Ivalon, acrylates, amino acids, alcohol, etc) may be a valid alternative.[17,20,21]

Conflicts of Interest
The authors declare no conflicts of interest.

References

1. Earhart RN, Aeling JA, Nuss DD, Mellette JR. Pseudo-Kaposi sarcoma. A patient with arteriovenous malformation and skin lesions simulating Kaposi sarcoma. Arch Dermatol. 1974;110:907-10.
2. Mali JWH, Kuiper JP, Hamers AA. Acro-angiodermatitis of the foot. Arch Dermatol. 1965;92:515-8.
3. Bluefarb SM, Adams LA. Arteriovenous malformation with angiodermatitis. Stasis dermatitis simulating Kaposi's sarcoma. Arch Dermatol. 1967;96:176-81.
4. Stewart WM. Fausse angiosarcomatose de Kaposi par fistules arterioveinulaires multiples. Bull Soc Fr Dermatol Syphil. 1967;74:664-5.
5. Goldblum OM, Kraus E, Bronner AK. Pseudo-Kaposi's sarcoma of the hand associated with an acquired, iatrogenic arteriovenous fistula. Arch Dermatol. 1985;121:1038-40.
6. Lyle WG, Given KS. Acroangiodermatitis (pseudo-Kaposi's sarcoma) associated with Klippel-Trenaunay syndrome. Ann Plast Surg. 1996;37:654-6.

7. Hodl S, Kresbach H. Kaposiform angiodermatitis (pseudo-Kaposi's disease) on an amputation stump. A new entity. Hautarzt. 1988;39:302-3.
8. Sbano P, Miracco C, Risulo M, Fimiani M. Acroangiodermatitis (pseudo-Kaposi sarcoma) associated with verrucous hyperplasia induced by suction-socket lower limb prosthesis. J Cutan Pathol. 2005;32:429-32.
9. Smiddy PF, Molloy MP, Flanagan N, Barnes L. Pseudo-Kaposi's sarcoma: The association of arterio-venous malformations with skin lesions resembling Kaposi's sarcoma. Australas Radiol. 2001;45:225-7.
10. Rosen T, Martin S, Stern JK. Radionuclide scanning in pseudo-Kaposi's sarcoma. Arch Dermatol. 1979;115:747-8.
11. Requena L, Sangueza OP. Cutaneous vascular proliferations. Part II. Hyperplasias and benign neoplasm. J Am Acad Dermatol. 1997;37:887-919.
12. Rongioletti F, Rebora A. Cutaneous reactive angiomatoses: patterns and classification of reactive vascular proliferation. J Am Acad Dermatol. 2003;49:887.
13. Marcoval J, Pagerols X, Iñiguez D, Peyri J. Acroangiodermatitis asociada a malformación arteriovenosa (síndrome de Bluefarb-Stewart). Actas Dermosifiliogr. 1992;83:143-5.
14. Punsoda G, Ribera M, Ferrándiz C. Acroangiodermatitis por fístula arterio-venosa. Pseudosarcoma de Kaposi tipo Bluefarb-Stewart. Actas Dermosifiliogr. 1991;82:337-40.
15. Fernández V, Pereiro M, Cervantes R, Zulaica A, Peteiro C, Toribio J. Síndrome de Bluefarb-Stewart. Actas Dermosifiliogr. 1987;78:291-3.
16. Kanitakis J, Narváez D, Claudy A. Expression of the CD34 antigen distinguishes Kaposi's sarcoma from pseudo-Kaposi's sarcoma (acroangiodermatitis). Br J Dermatol. 1996;134:44-6.
17. Brenner S, Martínez de Morentin E. What's new in pseudo-Kaposi's sarcoma. J Eur Acad Dermatol Venereol. 2001; 15:382-4.
18. Requena L, Farina MC, Renedo G, Álvarez A, Yus ES, Sangueza OP. Intravascular and diffuse dermal reactive angioendotheliomatosis secondary to iatrogenic arteriovenous fistulas. J Cutan Pathol. 1999;26:159-64.
19. Ikeda E, Kano E, Baba S, Suzuki H. Mast cells in pseudo-Kaposi's sarcoma lesions. J Eur Acad Dermatol Venereol. 2001;15:487-9.
20. Larralde M, González V, Marietti R, Nussembaum D, Peirano M, Schroh R. Pseudo-Kaposi sarcoma with arteriovenous malformation. Pediatr Dermatol. 2001;18:325-7.
21. Agrawal S, Rizal A, Agrawal CS, Agrawal A. Pseudo-Kaposi's sarcoma (Bluefarb-Stewart type). Int J Dermatol. 2005;44:136-8.