

treated with local topical steroids, to which he responded poorly.

All 3 patients were treated with local topical steroids, with a poor outcome in all cases. No systemic treatments were administered.

The clinical presentation of scalp sarcoidosis consists of indurated verrucous plaques and nodular lesions, often accompanied by alopecia, and can resemble that of discoid lupus erythematosus, necrobiosis lipoidica, organoid nevus, and cicatricial alopecia.<sup>1-5</sup> In general, scalp sarcoidosis is accompanied by sarcoid skin lesions in other locations.<sup>1,2</sup> Our 3 patients had infiltrated plaques with alopecia. Unfortunately, a trichoscopic examination, which is necessary to identify the trichoscopic features of scalp sarcoidosis, was not performed prior to biopsy. The alopecia may have been a consequence of the effects of the sarcoid granuloma on the follicles or of follicular replacement by the granuloma. All 3 patients had cutaneous sarcoidosis on the legs as well as the scalp. Sarcoidosis of other organs is common in patients with scalp sarcoidosis, and pulmonary and ophthalmologic sarcoidosis was observed in all 3 patients. In most cases, scalp sarcoidosis affects patients with active systemic sarcoidosis. We have previous experience with 2 other cases of scalp sarcoidosis in which biopsies were taken from locations other than the scalp. In both cases the patients' alopecia was strongly suggestive of sarcoidosis. Scalp involvement is common in cases of cutaneous sarcoidosis, and therefore a careful examination of the scalp should be performed if sarcoidosis is suspected. Indeed, scalp sarcoidosis may be less rare than previously thought. It is therefore important to include a scalp examination in the clinical examination of sarcoidosis patients.

## Conflicts of Interest

The authors declare that they have no conflicts of interest.

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## Stewart-Bluefarb Syndrome: Case Report and Review of the Literature<sup>☆</sup>



### Síndrome de Stewart-Bluefarb: caso clínico y revisión de la literatura

*To the Editor:*

Stewart-Bluefarb syndrome (SBS), also known as pseudo-Kaposi sarcoma, is a very rare subtype of angiodermatitis associated with congenital arteriovenous malformations (AVMs). We report a case of SBS and review the literature on this disease.

A 46-year-old man was referred to our unit for assessment of a painful, slow-growing ulcer on the lower right limb that had first appeared 3 months earlier. The patient had dyslipidemia and was an active smoker. The patient presented

with raised, self-limiting brownish papular lesions that had recurred since childhood and had never been diagnosed. Physical examination revealed symmetrical, bilateral palpable pulses without bruit or thrill, raised brownish-violaceous tumors in the pretibial and outermost supramalleolar region, perilesional eczema, and an ulcer measuring 3 × 2 cm with irregular borders, a fibrinous base, and mild signs of infection (Fig. 1), with no dysmetria of the lower limbs. Complete blood count, biochemistry, and serologic tests were negative or normal. Given the presence of atypical skin lesions, a complete Doppler ultrasound examination was performed on both limbs and a skin biopsy was carried out.

Histologic examination revealed the proliferation of capillaries in the deep dermis and papillary dermis, fibrosis, extravasated red blood cells, hemosiderin deposits, and tortuous vessels consistent with SBS. Doppler ultrasound confirmed the presence of underlying arteriovenous fistulas of the anterior and posterior tibial arteries with arterialized venous flow distal to the fistulas (Fig. 2).

Conservative treatment with 10 days of targeted antibiotic therapy was initiated after microbiologic culture detected the presence of *Staphylococcus aureus*. Compression therapy was also initiated with 2-in-1 compression stockings with a lateral zipper, as well as local wound care with a lipido-colloid dressing. At 3 months, the ulcer had healed and the pain had disappeared. The patient

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**Figure 1** Image showing raised lesions with perilesional eczema in the anterior pretibial surface and outermost supramalleolar region. An ulcer with perilesional cutaneous hyperpigmentation is visible at the center of the image.

has continued to use compression therapy and remains asymptomatic after 1 year (Fig. 3).

Angiodermatitis is a group of angioproliferative diseases that manifest as skin lesions resembling Kaposi sarcoma.<sup>1-3</sup> These entities consist of benign hyperplasia of preexisting vascular structures and can be associated with AVMs, as in the case of SBS.

SBS is a rare benign angioproliferative entity with histologic similarities to Kaposi sarcoma. It was first described by

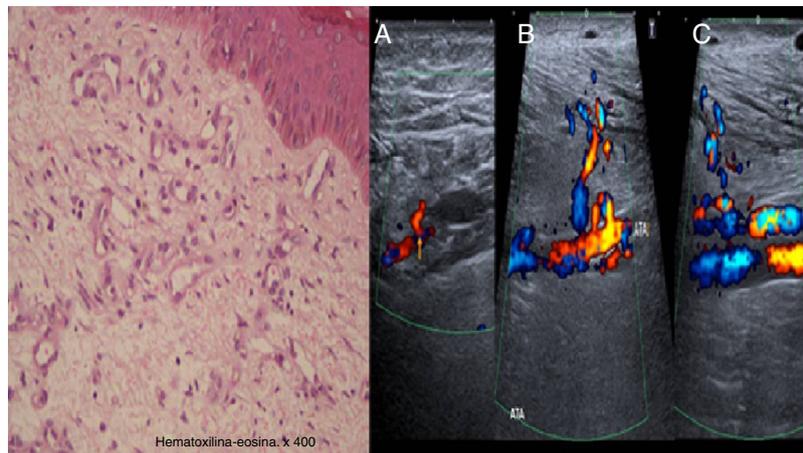


**Figure 3** Healed ulcer after conservative medical treatment and compression therapy.

Earhart et al.<sup>4</sup> in 1974 and fewer than 20 cases have been reported. SBS generally affects young men starting in the second decade of life<sup>5</sup> and is characterized by progressive brown or violaceous maculopapular skin lesions and underlying AVMs. The lesions appear unilaterally on the lower limbs—on the dorsum of the foot, on the ankle, and on the calf—and are accompanied by edema, local temperature increase, soft-tissue hypertrophy, and varicose veins as a consequence of venous stasis.<sup>2,6</sup> Complications can include ulcers and verrucous lesions.<sup>1,3</sup>

Differential diagnosis should include acroangiodermatitis of Mali, which typically appears in elderly patients as a consequence of chronic venous insufficiency and develops bilaterally, as well as Kaposi sarcoma, lichen planus, vasculitis, lymphoproliferative diseases characterized by congenital arteriovenous fistulas such as Klippel-Trenaunay-Weber syndrome, and iatrogenic diseases in patients who have hepatitis C or are undergoing dialysis.<sup>4,7,8</sup> Various theories have been proposed regarding the etiology and pathogenesis of SBS; an increase in venous pressure resulting from AVM or ischemia caused by arteriovenous steal can stimulate the proliferation of endothelial cells.<sup>7</sup>

Suspicion of SBS is purely clinical (atypical venous lesions, soft-tissue hypertrophy, palpation of thrill, or auscultation of bruit) and diagnosis is confirmed by histology; it is therefore important that dermatologists be familiar with this rare disease.<sup>8</sup> Histologically, SBS is characterized by the



**Figure 2** On the left, a histologic image showing the proliferation of capillaries in the deep dermis and papillary dermis, fibrosis, extravasated red blood cells, hemosiderin deposits, and tortuous vessels. On the right, Doppler ultrasound images showing an arteriovenous fistula of the posterior tibial artery (A), anterior tibial artery (B), and arterialized venous flow distal to the anterior tibial vein (C).

proliferation of endothelial cells, angiogenesis with a lobular pattern in the deep dermis and papillary dermis, and extravasated red blood cells, without fusiform neoplastic cells or proliferation independent of normal structures (features typical of Kaposi sarcoma).<sup>1,7</sup> Doppler ultrasound can detect the presence of arteriovenous communications,<sup>2,3</sup> although arteriography remains the gold standard<sup>1,7</sup> for diagnosis and treatment.<sup>3,5</sup>

Treatment of SBS remains controversial. Most experts recommend conservative treatment<sup>8</sup> and preventive treatment with compression stockings and limb elevation if edema is present.<sup>6</sup> Symptomatic treatment of local complications is crucial,<sup>1</sup> whereas specific treatment is rarely possible due to the presence of various distal communications.<sup>7,8</sup> Surgery is indicated in patients with functional impotence, refractory pain, recurrent infection, bleeding, or cardiac decompensation.<sup>5,8</sup> Amputation of the limb is sometimes the only option.<sup>9</sup> Selective embolization with different particles, intravenous laser, radiofrequency ablation, and ultrasound-guided sclerotherapy can be alternatives in complex and symptomatic cases.<sup>5</sup> A recent case report described the use of the glycosaminoglycan Caciqliq20 (heparin sulfate analogue) in a patient with SBS, with complete resolution of the ulcer.<sup>10</sup>

In conclusion, we have described a case of a rare syndrome of angiokeratosis that manifested as a chronic painful ulcer and skin lesions resembling Kaposi sarcoma, arising from an underlying arteriovenous communication. We have described a new vision for diagnosis using Doppler ultrasound and review the handful of studies on controversial therapeutic measures for this disease that have been published to date.

### Conflicts of Interest

The authors declare that they have no conflicts of interest.

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## Correlation Between Clinical, Histopathologic, and Ultrasound Findings in Focal Acral Hyperkeratosis<sup>☆</sup>

### Hiperqueratosis acral focal: correlación clínica, histopatológica y ecográfica



To the Editor:

We report the case of a 38-year-old woman (skin type V) with a history of tricuspid and mitral regurgitation who had undergone pulmonary bilobectomy for atypical carcinoid tumor. She was taking oral contraceptives. She attended our center with a flare-up of pruriginous lesions on her hands and forearms that had first appeared 1 year previously. The lesions appeared every month and did not remit completely between flare-ups, although they did improve with medium-potency topical corticosteroids. She reported no lesions at other sites and denied having had concomitant symptoms. Physical examination revealed poorly defined, apparently residual erythematous-brownish macules on the anterior and lateral surfaces of the wrists. Multiple hyperpigmented papules with a shiny surface measuring between

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