CASE REPORT

Acroangiodermatitis or Kaposi's Pseudosarcoma Mali Type Secondary to Chronic Venous Insufficiency due to May-Thurner Syndrome

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ABSTRACT

Acroangiodermatitis, or Kaposi's pseudosarcoma, is a rare angioproliferative dermopathy, generally found on the lower extremities. Among the four described types, the Mali type is the most common in adults, resulting from vascular disorders associated with chronic venous insufficiency.

We present the case of a patient with a history of deep vein thrombosis complicated by pulmonary embolism secondary to May-Thurner Syndrome, diagnosed through a study of skin lesions compatible with Mali-type acroangiodermatitis.

Although the relationship between venous insufficiency and this dermopathy is well-known, there is no published case of this presentation secondary to May-Thurner Syndrome, increasing the risk of underdiagnosing both conditions.

INTRODUCTION

Acroangiodermatitis (AAD), or Kaposi's pseudosarcoma, is a rare inflammatory proliferative vascular dermopathy, chronic, reactive, and benign in nature, which can resemble Kaposi's sarcoma (KS). It is observed in patients with chronic venous insufficiency, congenital arteriovenous malformation, acquired iatrogenic arteriovenous fistula, paralyzed limb, and amputation stumps.[1,4] Four variants are recognized: 1) Stewart-Bluefarb Syndrome, typically appearing unilaterally at a young age, resulting from congenital arteriovenous malformations and arteriovenous shunts; 2) Mali type, occurring in adults due to chronic venous insufficiency, often bilaterally, also associated with hypercoagulable states; 3) Favre dermatitis of the first pregnancy, manifesting as purpura on varicose lower limbs; and 4) AAD in arteriovenous fistulas for hemodialysis in chronic renal failure.[3,5,6]

The most accepted etiopathogenesis is hypoperfusion, where vascular alterations leading to venous plexus dysfunction cause reflux and venous hypertension, with consequent chronic edema and tissue hypoxia resulting

in skin inflammation and fibrosis, an increase in fibroblasts, hypertrophy, and reactive capillary proliferation.[2,5] The Mali type variant usually begins on the left side of the body and later becomes bilateral, affecting the lower part of the leg, especially the medial malleolus, the dorsal surface of the foot, and the toes with slow-developing red-violet macules, papules, or plaques, which can become verrucous or form painful ulcers.[3,5,7] Poroid and bullous forms have also been described.[8]

Histopathologically, there is a proliferation of new papillary dermal capillaries with thick walls in a lobular pattern, hypertrophic and tortuous venules lined by plump endothelial cells; fibroblast infiltration, luminal thrombi, hemosiderin deposits, and extravasated erythrocytes with a superficial perivascular infiltrate of lymphocytes and histiocytes. Differentiation from KS is necessary both clinically and histologically. AAD presents with parakeratosis, epidermal spongiosis, a greater deposit of hemosiderin, and absence of CD34+ staining in perivascular cells; while KS shows proliferation of irregular vascular channels with slit-like lumina, cellular atypia, CD34+ immunohistochemistry in endothelial and perivascular cells, and positive staining for human herpesvirus.[3,5,9]

Some patients with stasis dermatitis may develop AAD, and other differential diagnoses include lichen planus, pigmented purpura, and deep mycosis. To distinguish it from Stewart-Bluefarb syndrome, imaging studies are required. Treatment consists of correcting the underlying vascular pathology through surgical interventions, selective embolization, or sclerotherapy; symptomatically, compression and elevation help. Antibiotics (erythromycin) or dapsone combined with topical corticosteroids and compression measures have also been used.[2,5,8,10]

CLINICAL CASE

We present the case of a 33-year-old male patient with a history of deep vein thrombosis in the left leg nine years ago, complicated by pulmonary embolism. Referred from vascular surgery to dermatology due to a two-year history of an ochre-colored hyperpigmented macule on the medial malleolus of the left foot, extending to the dorsum of the foot as a violaceous plaque (Fig. 1). Within two months, the lesion progressed to an ulcer with irregular borders located from the first to the fourth toe, covered with meliceric crust, with seropurulent exudate, accompanied by edema and functional impairment (Fig. 2). Pedal pulses were present.

In the complementary tests, the secretion culture showed Enterococcus faecalis sensitive to vancomycin; the histopathological report of the lesion revealed a proliferation of dilated small-caliber vessels with prominent endothelium on edematous dermis, surrounded

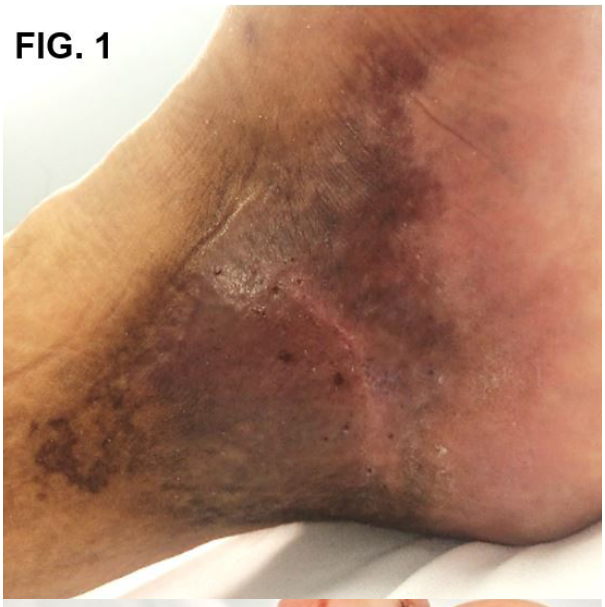
FIG. 1

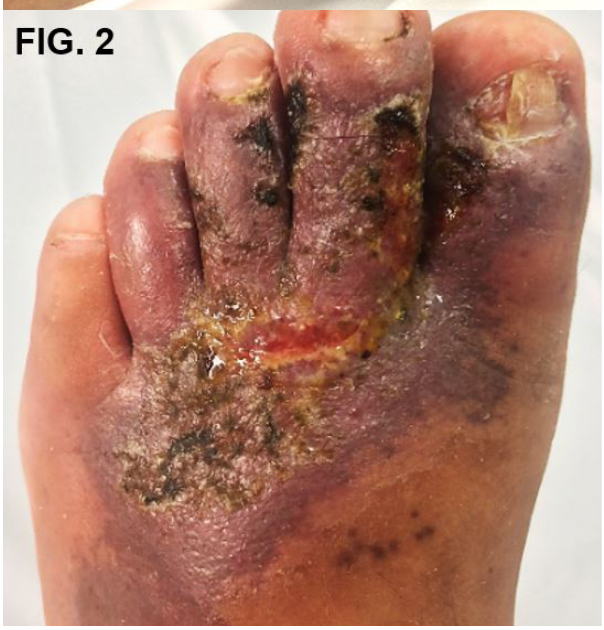
FIG. 2

by endothelial cells in areas where the vessels form nodules, with extravasated erythrocytes and hemosiderin deposits (Figs. 3 and 4). A new consultation with vascular surgery was conducted, and an angio-CT scan showed evidence of compression of the left iliac vein, positioned between the left iliac artery and the L5 vertebral body, leading to the diagnosis of May-Thurner Syndrome (MTS) (Fig. 5).

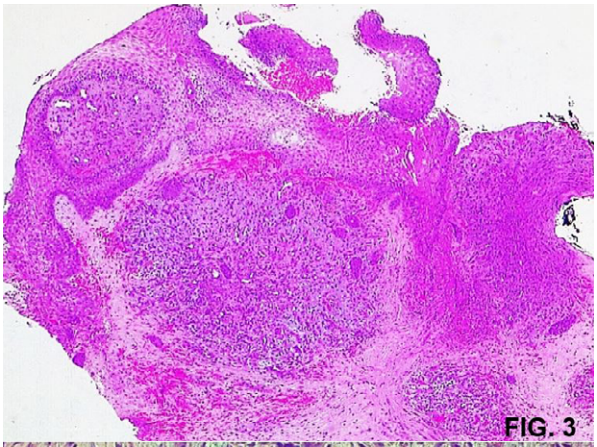
FIG. 3

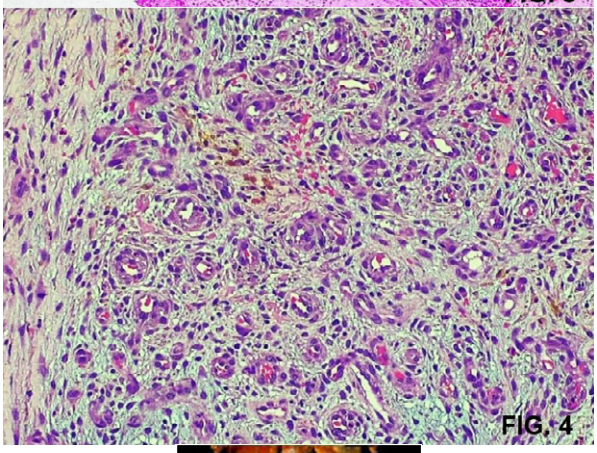
FIG. 4

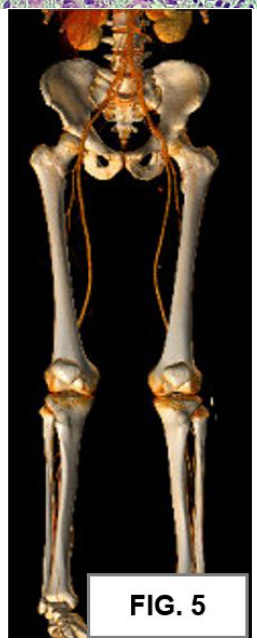
FIG. 5

The treatment consisted of wound care with debridement and parenteral antibiotics, which improved the infection, in addition to dapsone. Vascular surgery performed embolizations and placed a vena cava filter, with resolution of the lesion at the 3-month follow-up.

DISCUSSION

Acroangiodermatitis is a benign, rare cutaneous vascular disorder classified within reactive angioproliferative diseases. The Mali type variant is more common in adult men, affecting the distal lower extremities and associated with chronic venous insufficiency (CVI).[1,4] Since CVI is the most prevalent vascular disease, AAD is more likely to appear as a rare complication in patients with chronic venous stasis and venous hypertension in the lower limbs,[11] as in this patient, whose trigger was found to be May-Thurner Syndrome (MTS).

MTS, also known as Cockett Syndrome or iliac vein compression syndrome, most often refers to the phenomenon of left iliac vein compression by the right common iliac artery above and the fifth lumbar vertebra below, with the left side being more frequently affected. It predominantly occurs in women between the third and fifth decades of life.[10] Although the prevalence is relatively high, it is often underdiagnosed as most cases are asymptomatic;[11,12] however, in 2-5% of cases, it results in CVI (varicose veins, leg pigmentation, venous ulcers) or deep vein thrombosis (DVT) due to chronic venous tissue damage caused by constant arterial pulsation.[10,12,13] Optimal treatment includes thrombus removal (catheter-directed thrombolysis or mechanical aspiration) if DVT is present, followed by correction of the underlying iliac stenosis via balloon dilation, vena cava filter, or stent placement.[10,12]

Although the literature suggests that MTS should be considered in cases of left lower limb DVT,[10,12,13] our patient, who had this history complicated by pulmonary embolism at a young age, was not initially considered for this diagnosis and did not receive the corresponding

treatment. After dermatological assessment, with histopathology revealing Mali-type AAD, a new evaluation by vascular surgery was requested to investigate the underlying cause of CVI, ultimately revealing iliac compression, diagnosing, and optimally treating MTS, thereby preventing other thromboembolic complications and supporting the resolution of the cutaneous pathology.

CONCLUSION

AAD is a rare angioproliferative dermopathy, with the Mali type variant typically beginning in the left lower extremity and subsequently becoming bilateral in response to chronic venous insufficiency. In our patient, MTS and DVT were identified as factors contributing to CVI. In this case, as in the majority of published cases, the diagnosis of AAD is initially incorrect or delayed, either due to nonspecific symptoms that mimic other cutaneous conditions or lack of awareness of its existence, making it a challenge—especially if MTS is not considered as a potential trigger. Therefore, a high degree of suspicion is necessary, and AAD should be considered as a differential diagnosis in angioproliferative lesions resulting from venous insufficiency and stasis dermatitis, to avoid inadequate treatments and complications, address the underlying cause, and improve prognosis and quality of life.

REFERENCES

1. Chea EP, Rutt VL, Levin J, McClain R, Purcell SM. Acroangiodermatitis of Mali and Stewart-Bluefarb syndrome. Cutis. 2019;103(6).
2. Yosipovitch G, Nedorost ST, Silverberg JI, Friedman AJ, Canosa JM, Cha A. Stasis Dermatitis: An Overview of Its Clinical Presentation, Pathogenesis, and Management. Am J Clin Dermatol. 2023;24(2).
3. Yang C, Li D, Li Y, Li W, Zhang M, Yang X. Pseudo-Kaposi's Sarcoma: A Rare Case and Review. Clin Cosmet Investig Dermatol. 2023;16:1319–1323.
4. Rongioletti F, Rebora A. Cutaneous reactive angiomatoses: Patterns and classification of reactive vascular proliferation. J Am Acad Dermatol. 2003;49(5):887–96.
5. Lauck K, Nguyen QB, Klimas N, Rogge M. Acroangiodermatitis presenting as unilateral hypertrophic verrucous plaques. Dermatol Online J. 2022;28(2).
6. Mehta A, Pereira R, Nayak C, Dhurat R. Acroangiodermatitis of Mali: A rare vascular phenomenon. Indian J Dermatol Venereol Leprol. 2010;76(5)
7. Sun L, Duarte S, Soares-de-Almeida L. Acroangiodermatitis of Mali—An Unusual Cause of Painful Ulcer. Actas Dermosifiliogr. 2023;114(6).
8. Santosa A, Chandran NS. Novel manifestations of acroangiodermatitis: A report of two cases. Indian J Dermatol. 2020;65(3).
9. Lebwohl M, Kirsner RS, Margolis DJ, Barankin B, Hashimoto T, Canosa JM, et al. Stasis dermatitis: A challenging patient journey. JEADV Clin Pract. 2023;2(4).
10. Poyyamoli S, Mehta P, Cherian M, Anand RR, Patil SB, Kalva S, et al. May-Thurner syndrome. Cardiovasc Diagn Ther. 2021;11(5).
11. González Escudero M, Peraza Cruz D, Roque Pérez L. Pseudosarcoma de Kaposi tipo Mali unilateral en paciente con insuficiencia venosa crónica. Rev Habanera Ciencias Medicas. 2020;19(5):1–15.
12. Sun Y, Song S. Nonnegligible causes of symptoms of acute lower extremities—3 cases of May-Thurner syndrome with deep vein thrombosis. Thromb J. 2021;19(1).
13. Alkhater M, Jockenhöfer F, Stoffels I, Dissemond J. May-Thurner syndrome: an often overlooked cause for refractory venous leg ulcers. Int Wound J. 2017;14:578–82.