

Erythema Annulare Centrifugum in a Patient With Budd-Chiari Syndrome[☆]



Eritema anular centrífugo en un paciente con síndrome de Budd-Chiari

To the Editor:

A 56-year-old woman was observed for non-pruritic erythematous annular plaques, located symmetrically on knees, which have started 3 months before and had been progressing with peripheral extension and central clearing (Fig. 1). Past medical history was unremarkable until 1 week before, when she presented with abdominal pain, nausea, hepatomegaly, ascites, decreased urinary output and asthenia. Family history was irrelevant and the patient denied taking any medication.

Histopathologic examination of skin lesions biopsy showed a perivascular lymphocytic infiltrate in the superficial and deep dermis, suggestive of erythema annulare centrifugum (EAC), based on the correlation between clinical and histological findings (Fig. 2) on hematoxylin and eosin stain. Periodic acid-Schiff and Pearls colorations did not show remarkable findings.

Laboratory tests detected thrombocytosis; negative viral hepatitis serology, negative HIV, CMV and EBV serology; negative alfa fetoprotein; negative autoantibodies (ANA, anti-sDNA, antiphospholipid); undetected rheumatoid factor; normal levels of protein C, S, antithrombin II and homocysteine, no resistance to activate protein C. The search for prothrombin G20210A mutation was negative.

Doppler ultrasonography and computed tomography examination revealed thrombosis of the sub-hepatic veins

and portal hypertension with the diagnostic of Budd Chiari-Syndrome (BCS). Further investigation led to the diagnosis of essential thrombocythemia (ET) JAK2 negative, confirmed by bone marrow examination.

Despite anticoagulation and optimized diuretic therapy, renal function worsened and ascites persisted, and the patient was proposed for liver transplantation. Few weeks after transplantation, spontaneous resolution of cutaneous lesions was noticed, without relapse during 2 years of follow-up. Concerning her hematologic disease, she has been treated with hydroxyurea, however with poor results.

Discussion

Erythema annulare centrifugum (EAC) is a rare inflammatory skin disease presenting with erythematous papules or plaques that expand centrifugally with central clearing resulting in an annular shape.^{1,2} There are two types, superficial and deep, but this classification is not completely accepted and according to some authors the designation EAC should be reserved for the superficial type only.³

EAC has been considered a hypersensitivity reaction, rather than a specific clinicopathologic entity.⁴ When a trigger is identified, the treatment of the underlying condition mostly results in spontaneous resolution of the skin lesions.⁵ EAC has been associated with many different entities, including infections, malignant neoplasms,⁶ various autoimmune diseases^{2,3,7} and drugs,⁸ but its association with BCS or liver failure had not been previously described.

BCS is a rare vascular liver disease resulting from the obstruction of the hepatic venous outflow tract that can result in liver failure.⁹ The most common underlying



Figure 1 (a,b) Physical examination revealed well-defined erythematous annular plaques, located symmetrically on knees arranged in an annular shape.

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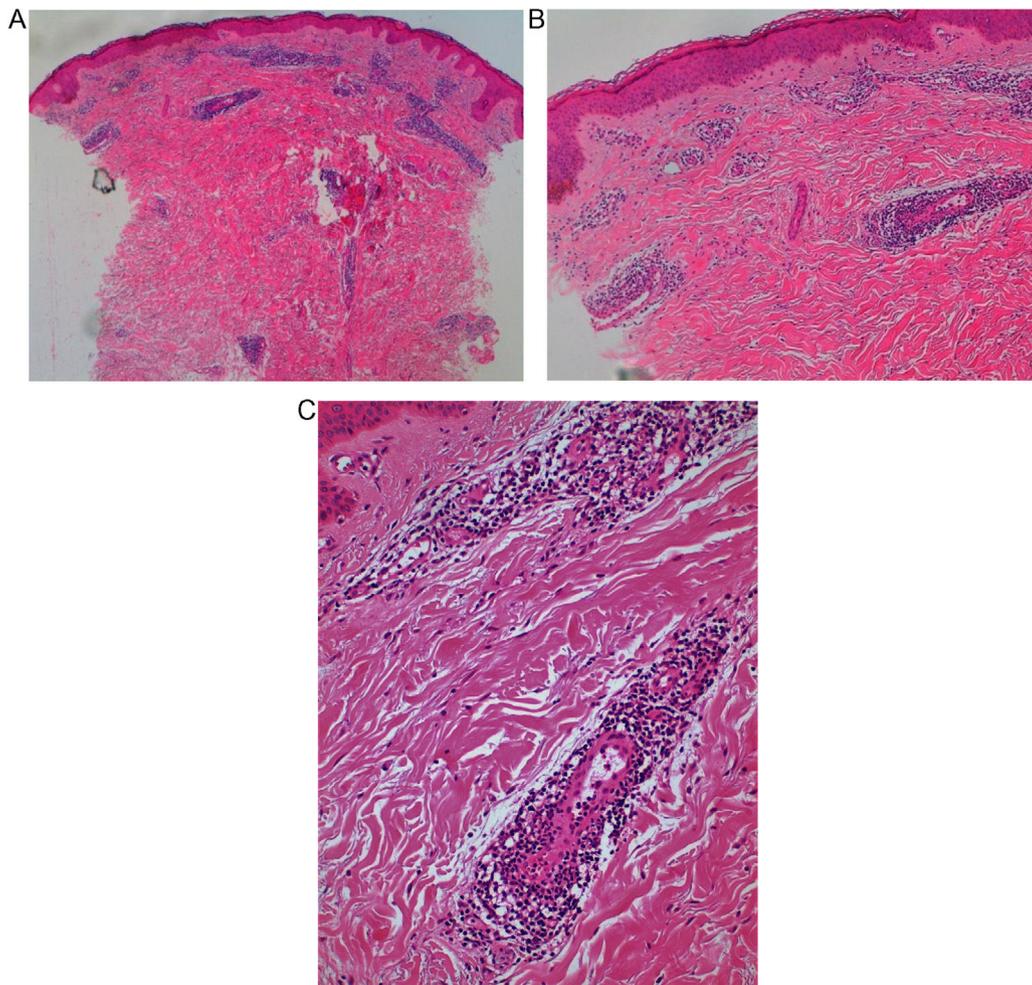


Figure 2 (a, x40; b, x100; c, x200) Histopathological examination (H&E) revealed an epidermis without significant changes and a perivascular histolymphocytic infiltrate in the superficial and deep dermis. There is no evidence of vasculitis.

prothrombotic risk factor is a myeloproliferative disorder as ET, although it is now recognized that almost half of patients have multiple underlying prothrombotic risk factors.¹⁰ Some chemical mediators released from activated platelets could explain some cutaneous findings associated with thrombocytosis such as erythromelalgia, acute febrile neutrophilic dermatosis and arterial thrombotic diseases¹¹ but not EAC.

In our case, the skin lesion resolved without recurrence after liver transplantation. On the other hand, there was not an haematological improvement despite preconized therapy making the association between EAC and BCS/liver disease more likely than with ET.

To the best of our knowledge, we report the first case of EAC probably associated with BCS in a patient with ET. Our report shows a new association of EAC with a systemic disease, thus emphasizing the importance of clinical and complementary evaluation in a patient with EAC, as well as the importance of more studies to clarify the physiopathology of this clinical identity.

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Successful Suppression of Recurrent Zosteriform Mycosis Fungoides With Maintenance Valacyclovir[☆]



Supresión de un cuadro de micosis fungoide recurrente con distribución zosteriforme mediante tratamiento de mantenimiento con valaciclovir

To the Editor

Mycosis fungoides (MF) is a cutaneous T-cell lymphoma (CTCL) that typically presents as patches or plaques in sun-protected areas. However, MF frequently exhibits clinical variability, and its atypical presentations can make the diagnosis difficult. Histopathology classically shows an atypical, superficial lymphoid infiltrate with epidermotropism. In addition, molecular assays can demonstrate a dominant T-cell clone in the skin, and flow cytometry can illustrate immunophenotypic abnormalities characteristic of MF. Zosteriform MF is an exceedingly rare variant in which lesions occur in a dermatomal distribution.^{1–3} We describe the first case of zosteriform MF that responded to antiviral therapy.

Interestingly, the patient's MF recurred on four occasions when antiviral therapy was discontinued or the dose was reduced.

A 69-year-old African-American female presented in October 2002 with a one-month history of erythema, pruritus, and hypo- and hyperpigmentation that began on her left upper back and spread down her arm onto her left chest in a dermatomal distribution. She denied pain, anesthesia, or history of herpes zoster. Physical examination revealed hypo- and hyperpigmented patches on left upper back (9 × 8 cm), left upper chest (8 × 7 cm), and left arm (9 × 4 cm) with 3% total body surface area (BSA) involvement (Fig. 1A; Fig. 2A). There were no bullae present. Wood's light examination showed depigmentation consistent with vitiligo.

Biopsy of the left upper back showed a dermal CD4+ and CD8+ atypical lymphoid infiltrate with focal epidermotropism (Fig. 3A–B). Immunohistochemical studies (Fig. 3C–F) demonstrated CD3+ T cells in the epidermis and dermis with predominance of CD4 over CD8 in the dermis, with a CD4:CD8 ratio of approximately 4:1. In the epidermis, there was a subset of atypical lymphocytes negative for CD4 and CD8. There is loss of CD7 expression. Rare cells were reactive for CD30. The majority of the lymphocytes were positive for TCR beta (BF1) and negative for TCR gamma. Monoclonal T-cell receptor gamma-chain gene

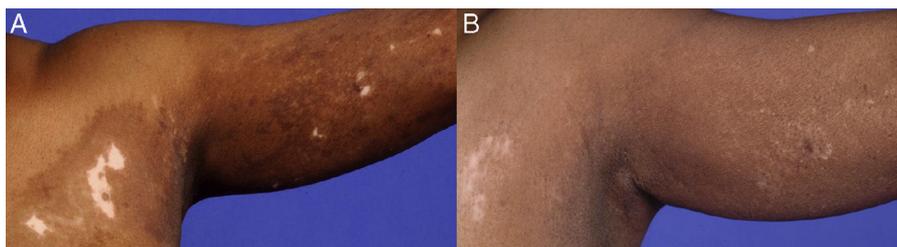


Figure 1 Área con hypo- e hiperpigmentación en región superior del torso y brazo A) como presentación inicial B) y pigmentada en el 90% con tratamiento con valaciclovir.

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