Pigmented purpuric dermatosis (PPD)-like mycosis fungoides on the leg of a Filipino male

Mycosis fungoides (MF) is an epidermotropic, primary cutaneous T-cell lymphoma (CTCL) composed of small to medium-sized T-lymphocytes with cerebriform nuclei and with a T-helper phenotype.[1] This is the most common type of cutaneous lymphoma, which represents almost 50% of all lymphomas arising in the skin.[1-3] The incidence of MF is approximately 0.36 per 100,000 person-years in the United States.[4-5]

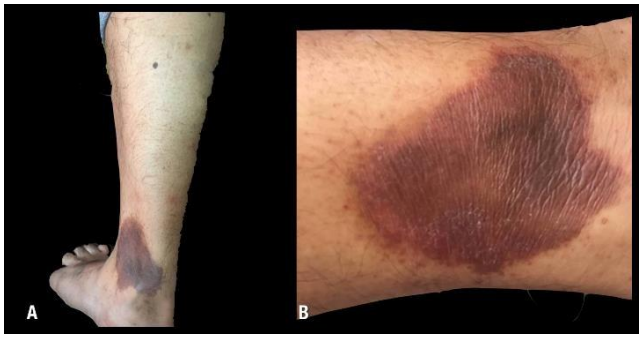

Figure 1A-1B. Solitary reddish brown plaque over the medial aspect of the right foot measuring 6 x 8 cms. Petechiae and purpuric macules are observed at the periphery of the plaque

Pigmented purpuric dermatosis (PPD) is a general term that is used to describe a group of chronic and relapsing cutaneous lesions of unknown etiology. This is characterized by petechiae and pigmentary macules of the lower limbs.[6, 7]

Mycosis fungoides may present in various forms as there are atypical variants that have been described in literature, one of the rare forms is the pigmented-purpura like MF.[8] Mycosis fungoides clinically presenting as PPD was also described in a renal transplant patient.[9]

We present a case of a 44-year-old Filipino male from Bicol, with a one-year history of an asymptomatic reddish-brown plaque on the medial aspect of the right foot. Review of systems, past medical and family history were unremarkable. Physical examination revealed a solitary, well-defined, irregularly-shaped reddish-brown plaque on the medial aspect of the right foot measuring 6 X 8 cms. Purpura and petechiae were noted at the periphery of the plaque (Figures 1A & 1B). The differential diagnosis at the time of consult were lichen simplex chronicus, pigmented purpuric dermatosis, and acroangiodermatitis. A 4-mm skin punch biopsy of the plaque showed focal vacuolar alteration of the basal layer, mild exocytosis of lymphocytes, and epidermotropism of medium-sized lymphocytes with perinuclear halo. The dermis revealed a focal lichenoid inflammatory infiltrate of lymphocytes and histiocytes. Dilated blood vessels and deposits of hemosiderin were also seen (Figure 2A & 2B). Immunohistochemistry showed positive staining of the epidermotropic lymphocytes with the immunohistochemical marker CD3 (Figure 2C) which confirmed the diagnosis of mycosis fungoides. He was started on methotrexate 15mg/week which showed significant improvement after 6 months of treatment (Figure

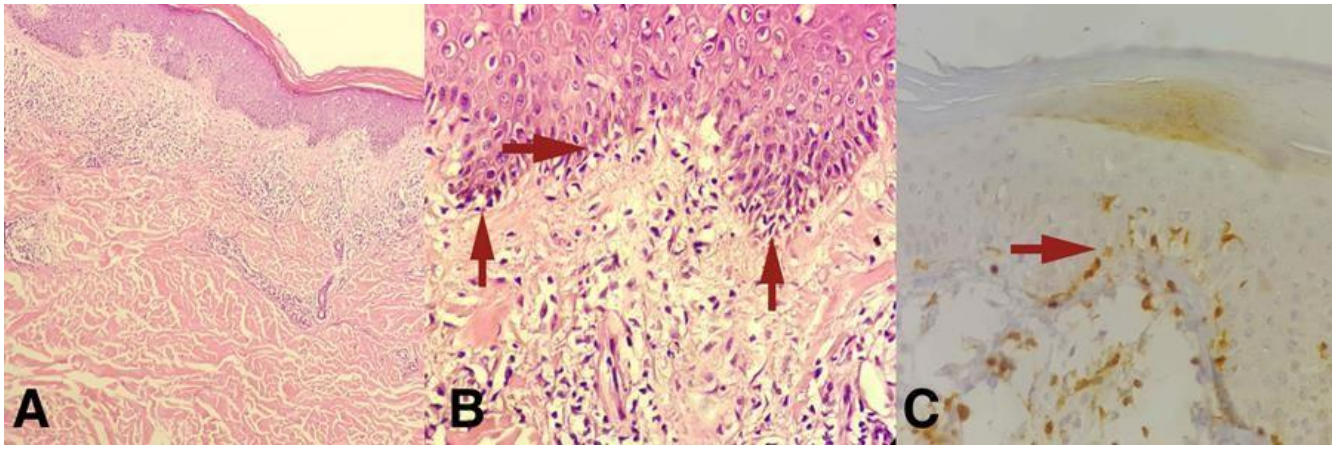

Figure 2A-2C. Figure 2: Histopathology shows mild acanthosis of the epidermis, epidermotropism of medium-sized lymphocytes with irregularly-shaped nuclei and perinuclear halo (red arrows). The dermis reveals wiry collagen bundles in the papillae and a focally lichenoid and superficial perivascular inflammatory infiltrate of lymphocytes (H & E) (A) x100, B) x400). Immunohistochemistry shows epidermotropism of atypical lymphocytes (red arrow) and papillary dermal lymphocytes positive for CD3 (C) x400)

3A & 3B).

Mycosis fungoides classically presents on the face, buttocks, intertriginous areas, and breasts. Atypical locations include the distal extremities, palms, soles and single lesions anywhere in the body.[10] In patients with dark skin, patches and plaques of MF appear less erythematous and have a greyish or silver hue.[11]

Our patient presented with an asymptomatic, reddish-brown plaque on the right lower leg. This clinical presentation may be mistaken for pigmented purpuric dermatosis as this usually presents as brown patches and plaques with few scattered purpura and petechiae over the lower extremities.[6] Histopathologic examination usually shows a focal lichenoid inflammatory infiltrate of lymphocytes and histiocytes with dilated blood vessels and deposits of hemosiderin.[12,13] However, the presence of epidermotropism of medium-sized to large lymphocytes with irregular nuclei and perinuclear halo favor a diagnosis of MF. [14] Immunostaining with CD 3, a T-cell marker usually confirms the diagnosis of MF. In the literature, MF may present as PPD-like lesions, as has been highlighted in various case reports. [8, 9, 15]

There are different treatment options for mycosis fungoides such as topical corticosteroids, imiquimod, tacrolimus, bexarotene, nitrogen mustard, phototherapy, radiotherapy and chemotherapy with methotrexate.[14] Another treatment option is the use of low dose methotrexate for the treatment of patch stage or focal plaque-stage mycosis fungoides.[16, 17] In this case, we started our patient on low dose methotrexate which showed significant improvement (Figure 3A-3B).

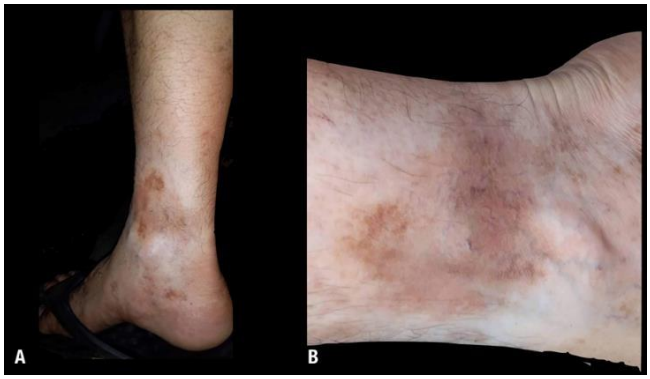

Figure 3A-3B. 6 months post-treatment with oral methotrexate showing significant resolution of the plaque with residual faint hyperpigmentation and telangiectasia.

In conclusion, we present a case of mycosis fungoides mimicking pigmented purpuric dermatosis which highlights the importance of a skin biopsy, immunohistochemistry, and clinicopathologic correlation. In rural areas where phototherapy is not available, oral methotrexate can be considered as a primary therapeutic option.

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