Degos disease-like presentation in systemic lupus erythematosus

Sir,

Degos disease (DD) or malignant atrophic papulosis or Kohlmeier-Degos-Delort-Tricort syndrome was first described in 1941 by Kohlmeier, and Robert Degos recognized it as a distinct entity in 1942.[1,2] This rare disorder is probably an endovasculitis affecting small- and medium-sized arteries leading to tissue infarction mainly in the skin, gastrointestinal system, and central nervous system. It is characterized by porcelain white atrophic papules with infarctive lesions. Variants of DD with only cutaneous involvement and association with collagen vascular disease have been described.[1,3] Hence, DD is considered as a distinctive reaction pattern rather than a specific disease.[4] We report a patient with the benign cutaneous form of DD whose immunological profile was suggestive of systemic lupus erythematosus (SLE).

A 32-year-old male presented with multiple erythematous papules and nodules mainly over the back of trunk and proximal extremities of 2 weeks duration. These lesions rapidly developed necrosis at the center which eventually healed leaving behind porcelain-white, atrophic scars within a week. He gave history of photosensitivity and erythema over the malar area.

Cutaneous examination revealed a faint malar rash with multiple skin color and erythematous papules and nodules of size 0.5-1 cm over the trunk. Some papules had central necrosis. Multiple round and linear atrophic porcelain white scars were seen over both arms and forearms, some having a rim of erythema [Figure 1]. Atrophic telangiectatic lesions were seen on the tips of thumb, index, and middle fingers.

Histopathology from an erythematous papule with central depression revealed dermal edema, mucin deposition and lymphocytic vasculitis consistent with DD [Figure 2]. A biopsy from a nodule showed

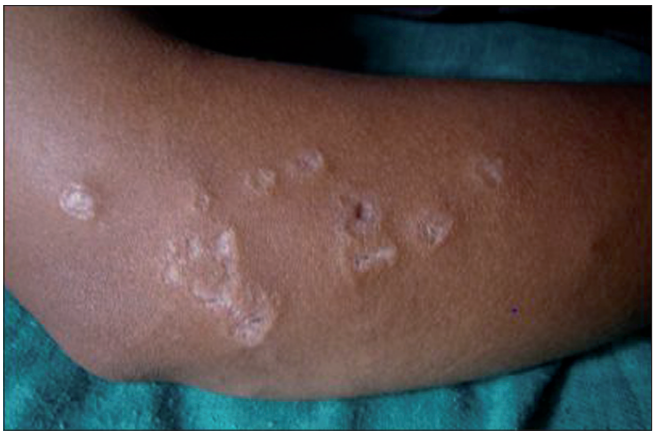

Figure 1: Papule with porcelain white atrophic centre

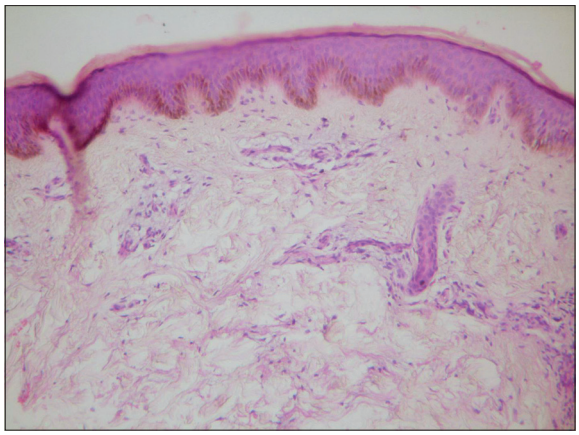

Figure 2: Dermal edema and mucin deposition with lymphocytic vasculitis, consistent with Degos disease (H and E, ×40)

basal cell degeneration, interface dermatitis, and periappendageal and perivascular infiltrate suggestive of SLE. The immunological profile revealed positive ANA, RA factor, anti-Sm, anti-u_1-RNP, anti-SS-A, and antiplatelet antibodies. With these findings we came to a diagnosis of benign cutaneous form of DD with SLE. He was treated with hydroxychloroquine and low dose aspirin.

DD is an uncommon condition characterized by asymptomatic papules which over a few days to weeks develop porcelain white depressed scars with rosy rim partly due to delicate telangiectases. A fatal outcome can occur within 1-2 years due to gastrointestinal and central nervous systems involvement.

Our patient had only cutaneous features suggestive of the benign cutaneous form of DD. Harvell *et al* have reported cases with only cutaneous involvement. About 4-15% of patients could have a benign course. Our patient also had malar rash, photosensitivity, positive ANA, and anti-Sm antibody, and past history of thrombocytopenia, suggestive of SLE. A review of the literature showed that until 2000, 16 patients with typical lesions of DD seemed to have other diseases concurrent with it, mainly collagen vascular diseases,[4] of which 11 had systemic lupus erythematosus and one each had rheumatoid arthritis, dermatomyositis, progressive systemic sclerosis, drug-induced immunosuppression, and AIDS. Dubin[5] *et al* described a 29-year-old man with clinical and histological features of DD and immunological features suggestive of SLE similar to our case. Black reported two females with SLE who developed atrophic papules of DD.[1] Histopathology of the early papule had shown abundant mucin which was similar to our finding. A biopsy from another lesion was suggestive of SLE. Doutre *et al*[3] have also reported similar findings.

According to Ball *et al*,[4] DD is a distinctive morphologic pattern induced by many different factors, rather than a disease with a single specific cause.

Since our patient had only cutaneous lesions and no systemic involvement, he was treated with hydroxychloroquine.

This case is reported for its exceptionally interesting and rare presentation with a varied immunological profile. Patients with skin lesions typical of DD should be assessed for other associated diseases like SLE, other collagen vascular diseases, and immunosuppressive disorders.

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