Case for diagnosis*

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CASE REPORT

A 45-year-old white female presented with a 1-year history of papular lesions, ulcerations, and scarring on the abdomen, back, and limbs. Six months after symptom onset, she developed nausea, vomiting, and weight loss of 15 kg. The patient reported she had been admitted to hospital for abdominal pain and ascites at some time during the last 3 months. During this hospitalization, exploratory laparotomy and peritoneal biopsy were performed, with noncontributory findings.

Physical examination revealed scarring and ulcerations on the chest, back, and abdomen, as well as papules with a porcelain-white center, some with an erythematous halo, on the extremities (Figures 1 and 2).

Laboratory tests, including antinuclear and antiphospholipid antibody titers, were within normal limits. Histopathological examination of an upper extremity papula showed thrombosed vascular structures and scant inflammatory cells around the vessels (Figure 3). The patient was prescribed acetylsalicylic acid (300 mg/day), dipyridamole (150 mg/day), and enoxaparin (40 mg/day). Her condition improved during the first week of treatment, but she developed abdominal pain caused by a bowel perforation, and died of sepsis.

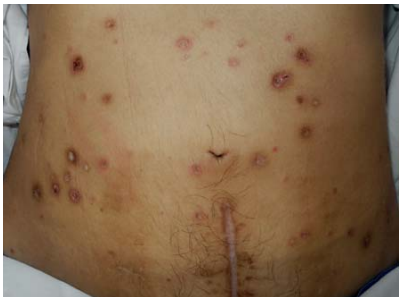

FIGURE 1: Multiple round scars and ulcerations on the abdomen, as well as an exploratory laparotomy scar

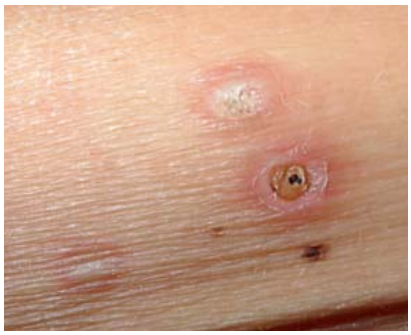

FIGURE 2: Scar with porcelain-white center and ulcerated crusted lesion on the forearm, each with a halo of erythema

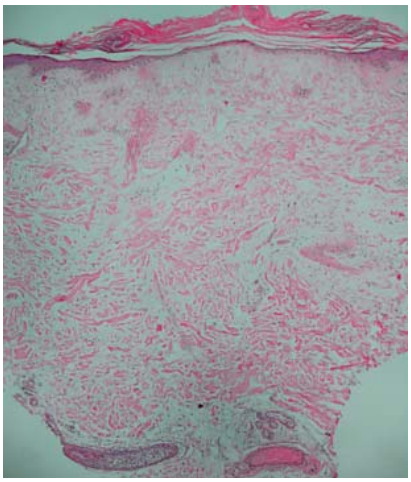

FIGURE 3: Skin biopsy specimen. Epidermis: hyperkeratosis and atrophy of the stratum spinosum. Dermis: localized necrobiosis with scant cells and absence of adnexal structures.Thrombosed blood vessels are visible deep in the dermis. (H & E stain, original magnification 40x)

DISCUSSION

In view of the clinical and laboratory findings, we established a diagnosis of Degos disease (DD). DD, also known as malignant atrophic papulosis, is a rare occlusive vasculopathy of unknown etiology, characterized by infarcts in the dermis, gastrointestinal tract, central nervous system, and other organs.[1,2,3,4] Two variants have been described: one benign, confined to the skin, and one malignant, with systemic involvement.[1,5] The first case was reported by Köhlmeier in 1941. DD affects individuals across all age ranges, but is most common in white young adults; there is a 3:1 male-to-female predominance.[1,2]

There are several theories about the pathogenesis of the disease: coagulopathy, vasculitis, primary

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endothelial cell disorder, and potential associations with collagen diseases, genetic factors, and viral infection (parvovirus B19).[2,4,5,6] Recent studies suggest it is an endotheliopathy, mediated by C5b-9 deposition and increased cell expression of interferon alpha leading to vascular changes.[7] Scheinfeld believes DD is a hematologic and/or endothelial condition, possibly attributable to an acquired, intrinsic genetic defect, and resembles paroxysmal nocturnal hemoglobinuria, a hematologic disease characterized by an alteration in the complement system that leads to intravascular hemolysis and thrombosis.[5]

The diagnosis of DD is mainly clinical. The appearance of the lesions varies according to the stage of disease progression. The initial lesions are round, pink papules, approximately 5 mm in diameter, which become umbilicated. In the chronic stage, these papules develop a porcelain-white central depression and a narrow, pink peripheral rim with fine telangiectasias. They are distributed predominantly across the trunk and limbs, and usually spare the scalp and palmoplantar regions.[2] The characteristic histological findings are wedge-shaped dermoepidermal necrosis and vascular thrombosis.[8]

DD affects several organs and systems, including the gastrointestinal tract, central nervous system, cardiopulmonary system, eyes, liver, and kidneys, usually after the onset of cutaneous lesions.[1] Approximately 50 to 60% of patients with systemic symptoms die within 2 to 3 years, most due to gastrointestinal perforation. Neurologic and ocular manifestations occur in 20% and 13% of patients respectively.[8]

Several drugs have been used in DD, including topical and systemic corticosteroids, azathioprine, methotrexate, ciclosporin, tacrolimus, mycophenolate mofetil, intravenous immunoglobulin, arsenic, sulfonamides, heparin, and warfarin, but none have produced satisfactory results.[1] Recent studies suggest that eculizumab, an anti-C5 monoclonal antibody, may be a promising alternative.[7] ❑

Abstract: Degos disease, also known as malignant atrophic papulosis, is a rare occlusive vasculopathy of unknown etiology characterized by infarcts in the dermis, gastrointestinal tract, central nervous system, and other organs. It is characterized by papules, which become umbilicated and evolve with a depressed porcelain-white central area, with an erythematous halo with telangiectasias. Histological findings include wedge-shaped dermoepidermal necrosis and blood vessel thrombosis. Approximately 50-60% of patients with systemic symptoms die within 2-3 years, most due to gastrointestinal perforation. We report a typical case, with lethal outcome, in a 45-year-old woman.
Keywords: Complement C5; Malignant atrophic papulosis; Vascular diseases

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