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Degos' Disease Mimicking Vasculitis

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Introduction

Degos' disease is a rare disorder with multisystem involvement and unknown etiology. This entity was first described by Degos in 1942 (1,2). Other synonyms for this disease are malignant atrophic papulosis, atrophic papulosquamous dermatitis, fatal cutaneous-intestinal syndrome, and thromboangiitis cutaneointestinalis disseminata (3). It has been more commonly reported in whites, men, and those in the third decade of life, although onset age ranges from 3 weeks to 67 years (4). The average course of the disease is reported to be around 2 years (1), but case reports of patients with a benign variant have been reported with survival of ~14 years (5). Death is most commonly due to intestinal perforation or cerebral infarction. We report a fatal case of Degos' disease that presented with abdominal pain and intestinal perforations, which clinically mimicked vasculitis.

Case report

A 49-year-old white woman initially developed painful oral ulcers and a maculopapular rash over her back, extremities, and trunk in April 1999. She then developed abdominal pain in March 2000 and underwent an evaluation that included colonoscopy and gastroscopy with biopsies, both of which were nondiagnostic. Five months later, the patient presented with a temperature of 103.5°F and an acute abdomen. Multiple maculopapular lesions with central pallor and an erythematous margin were observed on extremities, chest, and back. An abdominal computed tomography scan demonstrated free fluid, so she underwent exploratory laparotomy and small bowel resection due to perforating ulcers in the jejunum. Three weeks later, she had recurrent abdominal pain and underwent another exploratory laparotomy and bowel resection for 3 perforations involving the jejunum and ileum. Two days later, she underwent laparotomy again, this time a cyanotic distal ileum was revealed and resected. Histopathology showed severe necrotizing enteritis with interstitial hemorrhage, possibly ischemic, and acute serositis. Multiple vessels with thrombosis were seen, but inflammation was rare, and suggested the possibility of vasculitis. Cyclophosphamide and methylprednisolone were administered for presumed vasculitis. Infliximab therapy was also initiated. Enoxaparin was initiated due to concern of mesenteric thrombosis. The patient subsequently underwent revisions of the enterostomies. The biopsies and skin lesions were then found to be consistent with Degos' disease (Figure 1) and immunosuppressants were discontinued. She was then transferred to our institution for management in October 2000.

Past medical history included nonmetastatic breast cancer treated in 1990 with modified radical mastectomy, chemotherapy, and radiation therapy; hypertension; gastroesophageal reflux; atrial fibrillation; depression; and osteoporosis.

Relevant laboratory results were elevated C-reactive protein at 8.7 mg/dl (0–0.6) and rheumatoid factor at 85 IU/ml (0–30). Antiphospholipid antibody, dilute Russell viper venom time, protein C antigen, protein S antigen, angiotensin converting enzyme, antinuclear antibody, and antineutrophil cytoplasmic antibody were all negative or within normal ranges.

The patient's skin lesions and abdominal symptoms improved with intravenous dipyramidole (25 mg every 12 hours) and indomethacin (50 mg twice daily per rectum). Enoxaparin was discontinued because gastrointestinal bleeding was observed. She had no further recurrence of intestinal perforations and, being clinically stable, was discharged in November 2000. She subsequently moved out of state. However 3 months later, the patient developed pleural effusions, acute respiratory distress, and was given vasopressors. She developed abdominal symptoms similar to those experienced before and she died. An autopsy was not performed.

Discussion

Degos' disease is a rare vasculopathy involving multiple organ systems. The internal organs most commonly involved in the order of decreasing frequency are the gastrointestinal tract, central nervous system, thoracic organs, eyes, oral mucosa, and kidneys (6). Involvement of the bladder, pleura, and pancreas have also been reported (1). The most common initial presentation is the classic skin

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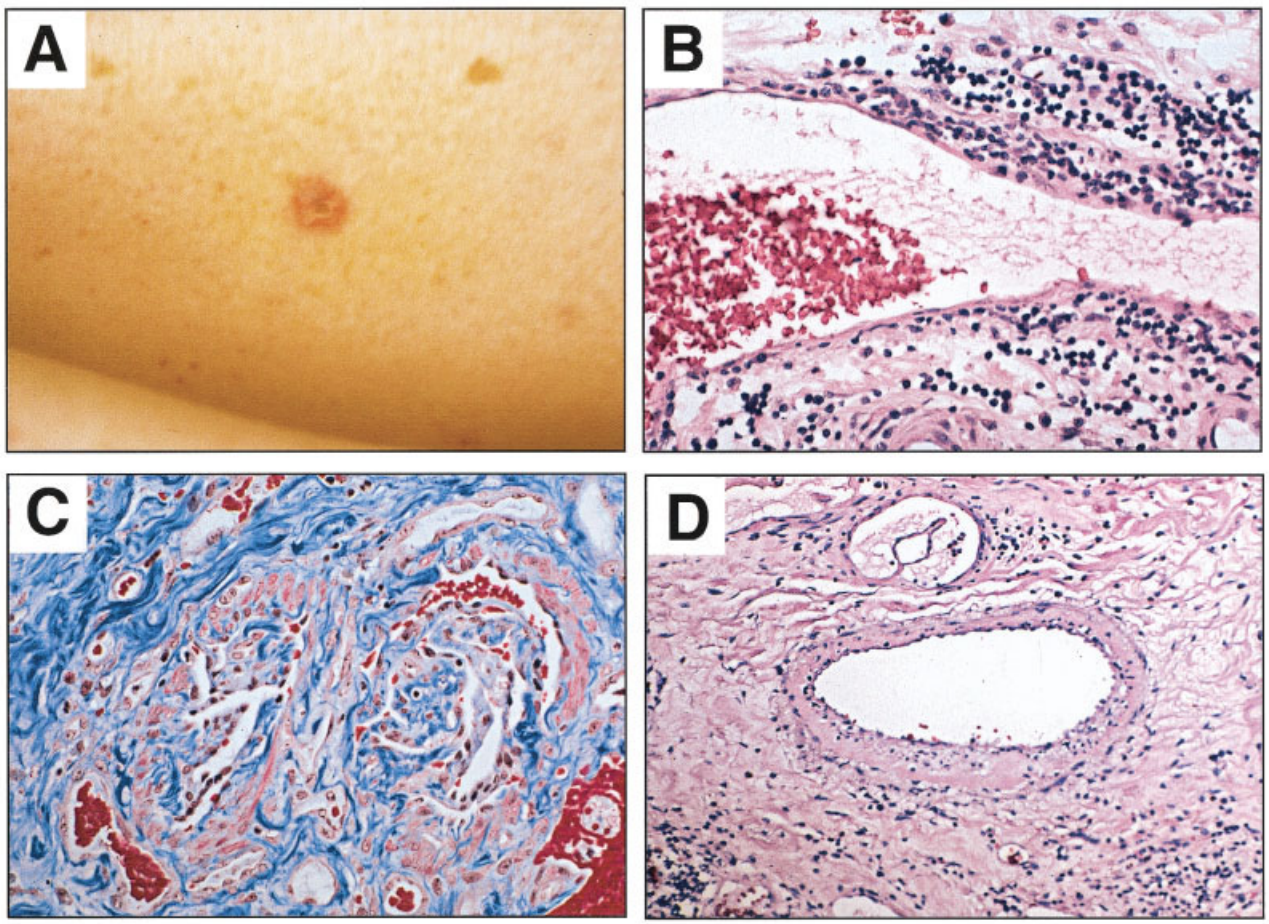

Figure 1. The typical skin lesion in Degos' disease, as seen in our patient, is described as maculopapular 2–5 mm, umbilicated, with a porcelain white center and erythematous margins (**Panel A**). Small intestine biopsy in this patient demonstrated vascular changes consistent with Degos' disease: rare inflamed vessels (**Panel B**), occlusion of vascular lumens by fibrointimal proliferation and organized fibrin thrombi (**Panel C**), and atrophy of vessel wall with fibrin deposition (**Panel D**).

eruption, followed after months to years by involvement of the internal organs. The typical skin lesion is seen primarily in the trunk and extremities and is described as maculopapular, umbilicated with a porcelain central area of necrosis and erythematous margins (3). Gastrointestinal tract involvement is seen in 47% of the patients (7) with clinical manifestations ranging from dyspepsia to an acute surgical abdomen with perforated viscus (4). It has also been suggested that endoscopy be performed routinely in all patients to detect asymptomatic silent perforations, even in those who do not have apparent gastrointestinal involvement (8). Central nervous system involvement is seen with hemiparesis, myelopathy, polyradiculopathy, visual disorders, and subsequently mental deterioration and cranial nerve palsies (9).

The etiology and pathogenesis of Degos' disease are still unknown. The etiology is speculated to be immunologic dysfunction, coagulation or fibrinolysis disturbances, or possibly viral infections (10). Histopathologically, involvement of small arteries, arterioles, and venous lesions have been described. There is intimal involvement with endothelial swelling, proliferation, infiltration of mononuclear cells, fibrosis, thrombus formation, and fibrin deposition (11). Multiple infarctive and thrombotic lesions may be seen and sometimes misdiagnosed as vasculitic lesions. However, the distinction is intimal proliferation with sparse inflammation around the affected vessels and the elastica, media, or adventitita being devoid of pathology (12). Immune complexes also have not been detected in most cases reported (13).

The diagnosis is mainly a clinicopathologic correlation. The typical skin lesions and internal organ involvement should alert a physician to this disease. Unfortunately, treatment options for Degos' disease are limited. Immunosuppressant therapy, anticoagulants, and plasma exchange have been tried but not proven to be effective (4). The antiplatelet agents aspirin and dipyramidole have been suggested to retard the process, but may contribute to risk of gastrointestinal and intracranial hemorrhage (14,15).

Thus it is important for physicians, especially rheumatologists, to be aware of the spectrum of the various clinical findings in Degos' disease and be vigilant because this can be mistaken for vasculitis. Furthermore, it is also important to consider this disease when vasculitis has been diagnosed but is not responding to immunosuppressive therapy.

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