Diffuse atrophic papules and plaques, intermittent abdominal pain, paresthesias, and cardiac abnormalities in a 55-year-old woman

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CASE SUMMARY

History

A 55-year-old Caucasian woman was referred to the National Institutes of Health (NIH) for evaluation of widespread white papules and plaques, intermittent abdominal pain, cardiac abnormalities, and paresthesias. Symptoms began at age 50 years with painful skin lesions on her extremities. Additional lesions later appeared on her chest, back, and abdomen over 3 to 4 months. Treatment with topical corticosteroids was initiated without benefit.

The following year, at age 51 years, the patient experienced new-onset severe, crampy abdominal pain. Abdominal computed tomography revealed evidence of small bowel inflammation. The pain subsided with bowel rest. She continued to experience occasional, mild abdominal pain for the next 2 years, during which time the skin lesions improved in appearance but never resolved entirely.

At age 54 years, the patient's abdominal symptoms worsened with near-weekly severe bouts of pain for 2 to 3 months. Abdominal computed tomography showed changes suggestive of inflammation in the duodenum and proximal jejunum with surrounding mesenteric fat stranding. Exploratory laparoscopy revealed widespread lesions on the surface of the liver, peritoneum, and large and small bowel; these were similar in appearance to the lesions on her skin.

This episodic abdominal pain was accompanied by the development of 10 to 20 painful new skin lesions and intermittent right-sided paresthesias of the arm and leg. Head and neck magnetic resonance imaging were read as normal, failing to reveal any neurologic defects. Electrocardiogram revealed asymptomatic atrial fibrillation; echocardiogram showed moderate tricuspid regurgitation deemed to be clinically insignificant. Already on daily 81 mg of aspirin, the patient was also prescribed twice daily apixaban (5 mg) and metoprolol (25 mg) to reduce the risk of thromboembolic complications.

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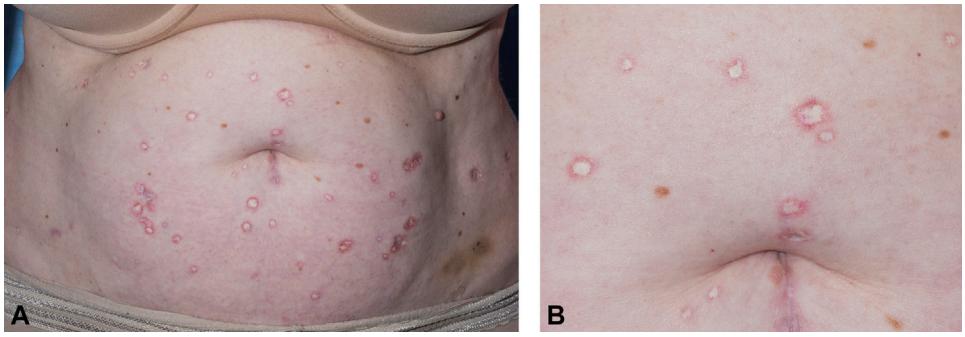

Fig 1. Malignant atrophic papulosis. **A**, Porcelain-white, irregularly shaped atrophic papules and plaques with erythematous, telangiectatic rim on the abdomen. **B**, Close-up view of atrophic papules.

Family history was significant for cerebrovascular accidents in both parents. There was no family history of skin or gastrointestinal disease. Approximately 5 years after disease onset, the patient was seen in the NIH dermatology clinic for further assessment.

Physical examination

Skin examination at the NIH showed numerous porcelain-white, irregularly shaped, pinpoint to 6- to 8-mm atrophic papules on the body with sparing of the face and neck. A distinct rim of intense erythema with telangiectasia characterized all lesions. Smaller lesions were present on the thenar and hypothenar aspect of the palms. In several areas, including the abdomen, back, and proximal aspect of her thighs, adjacent lesions formed larger atrophic plaques (Fig 1). A dark red, irregular 4-mm papule with slight overlying scale was noted on the medial aspect of her left knee.

Dermatopathology

Histopathologic examination of a skin punch biopsy specimen obtained from the medial aspect of her left knee revealed focal lichenoid dermatitis with hyperkeratosis, thickened basement membrane, superficial and deep perivascular chronic inflammation, hemosiderin deposition, and increased dermal mucin (Fig 2). No vascular immune complexes were seen on direct immunofluorescence.

Significant diagnostic studies

Laboratory studies were significant for the following: white blood cell count 3.04 K/μL (reference range, 3.98-10.04 K/μL), platelet count 165 K/μL (173-369 K/μL), and absolute lymphocytes 0.71 K/μL (1.18-3.74 K/μL). Lupus anticoagulant, antinuclear antibody, antidouble-stranded DNA, and anti-extractable nuclear antigen panel were within normal limits. IgM anticardiolipin antibody was 14 MPL (0—12 MPL); C3 and C4 complement were at the lower limit of normal. Tests for erythrocyte sedimentation rate, high-sensitivity C-reactive protein, fibrinogen, D-dimer, lactate dehydrogenase, immunoglobulins, and liver function revealed normal findings. Lipid panel was significant for total cholesterol of 229 mg/dL, low-density lipoprotein of 142 mg/dL, triglycerides of 212 mg/dL, and high-density lipoprotein of 45 mg/dL. Prothrombin time, partial thromboplastin time, and international normalized ratio were unremarkable with normal von Willebrand and factor VIII activity. Electrocardiogram showed sinus rhythm and was within normal limits. Echocardiogram revealed a mildly dilated left atrium and the presence of atrial fibrillation. There was no evidence of pulmonary hypertension.

Diagnosis

The diagnosis was adult-onset malignant atrophic papulosis (MAP)/systemic Degos disease (Online Mendelian Inheritance in Man no. 602248).

Follow-up

After the patient's exploratory laparoscopy revealed widespread visceral involvement, intravenous eculizumab 990 mg weekly for 4 weeks followed by 1200 mg every other week was added to her ongoing drug regimen. At the time of her evaluation at NIH, the patient had been treated with eculizumab for 6 months. She reported resolution of abdominal symptoms within 2 weeks of initiation of eculizumab with the exception of an isolated episode of pain that occurred 5 months into treatment and resolved within 12 hours. There has been minimal formation of new skin lesions and no new cardiac or neurologic symptoms developed since eculizumab was initiated, although paresthesias still occur intermittently. The patient continues to be treated with daily aspirin, twice daily apixaban and metoprolol, and 1200 mg of eculizumab twice monthly.