Benign cutaneous Degos disease

Dear Editor,

A 41-year-old woman presented with a 2-year history of multiple asymptomatic pea-sized papules with an erythematous, telangiectatic rim surrounding an atrophic, porcelain-white center on the trunk and extremities (Figure 1). She had experienced abdominal fullness and vomiting for 1 year. Histopathologic examination of a skin biopsy from the left forearm revealed hyperkeratosis, epidermal atrophy, vascular interface change, and wedge-shaped dermal infarct in which nuclear dust and extravasation of erythrocytes were present (Figure 2A and B). The vessels in the infarcted area showed focal obliteration and hyalinization (Figure 2C). Alcian blue staining demonstrated abundant mucin deposition outlining the wedge-shaped area of collagen degeneration (Figure 2D). Direct immunofluorescence was negative for IgG and C3. The clinical and pathological manifestations were compatible with Degos disease.

Complete blood count with differential, hepatorenal, and coagulation profiles, urinalysis and immunological fecal occult blood test were all within normal limits except for an elevated erythrocyte sedimentation rate. Results of other laboratory examinations were also within normal limits, including profiles of systemic lupus erythematosus, systemic sclerosis, rheumatoid arthritis, dermatomyositis, antiphospholipid syndrome, and vasculopathy. The ophthalmologic examination revealed no conjunctival avascular patches or telangiectasia. Magnetic resonance of the brain showed no abnormalities. Esophagogastroduodenoscopy revealed reflux esophagitis. Upper gastrointestinal and small intestine series showed no evidence of bowel perforation, obstruction or abnormal mucosal changes. A diagnosis of Degos disease, probably the benign form, was made based on the clinical and pathological features. The patient was treated with metoclopramide, aspirin, and dipyridamole in outpatient follow-up. Some new cutaneous lesions occurred and the old ones healed with atrophic scars. Although she still suffered from abdominal fullness and vomiting, no intestinal perforation, cerebrovascular accident, or other life-threatening complications occurred during the following 6 months.

Degos disease is a rare thrombo-obliterative vasculopathy characterized by pathognomonic atrophic porcelain-white papules with a surrounding telangiectatic rose-colored rim. Skin lesions usually precede systemic manifestations with the most common affected organ systems being, in order, gastrointestinal, neurologic, cardiac, and pulmonary systems. Ocular involvement, including avascular conjunctival patches, telangiectatic vessels, and scleral thinning, has also been described. The pathogenesis of the occlusive vasculopathy with eventual tissue infarction, although not clearly understood, may be related to coagulopathy, primary dysfunction of the endothelial cells, and dysregulation of interferon-α and the membranolytic attack complex of the complement system.

Degos disease is also known as malignant atrophic papulosis, nomenclature of which puts emphasis on characteristic skin eruptions and potentially fatal course in most patients. However, a minority of patients have a rather benign course with skin-limited disease, no systemic involvement and long-term survival.
be a hallmark of benign cutaneous Degos disease, but not all histopathologic reports of cases support this hypothesis.

Due to the rarity of Degos disease and the high rates of treatment failure for its classical variant, evidence-based efficacy of therapeutic modalities has been demonstrated in individual case reports, but not in randomized controlled trials. The combination of aspirin and dipyridamole or each alone has shown variable effectiveness. In patients with progressive malignant atrophic papulosis, treatment with eculizumab, the monoclonal antibody to the complement protein C5, demonstrated immediate efficacy but could not prevent further progression. Corticosteroids, generally thought to be contraindicated, may worsen skin eruptions and hasten complications. Surgical intervention including laparotomy for abdominal involvement has not been successful.

Our patient, who had a 2-year history of characteristic skin eruptions without significant evidence of systemic involvement, was regarded as benign cutaneous Degos disease. Therefore, dermatologists should be alert to the possibility of Degos disease in patients who present with pathognomonic skin lesions. Patients with benign cutaneous Degos disease may have a fair prognosis with long-term survival, but they require regular follow-up due to the possibility of future systemic involvement.

References