Degos’ disease: acute abdomen with skin rash

A 32-year-old man developed fecal discharge from his wound following appendectomy for presumed acute appendicitis. He was re-operated on ten days later and seven large ileal perforations (1-3 cm each) were identified. The diseased ileum was resected and primary anastomosis done, following which he developed enterocutaneous fistula in the right iliac fossa and was referred to our hospital. Examination showed multiple erythematous, papular lesions with central umbilication over his trunk and extremities. Some of the papules had healed with central porcelain-colored scar formation (Fig 1a). On enquiring, he revealed that the rash had appeared in crops over the last 6 months. The right bulbar conjunctiva showed dilated vessels with a fornix-based wedge of circumscribed conjunctival congestion surrounding an area of scleral thinning evident by the bluish tinge due to visibility of the choroidal vasculature (Fig 1b).

Hemogram and liver function tests were normal. Serological tests for HIV and systemic lupus were negative. CT fistulogram suggested distal ileo-cutaneous fistula. Histology of the skin lesions revealed papular mucinosis. Histology of the resected ileum revealed fibrinoid necrosis with lymphocytic and neutrophilic infiltration and stenotic endothelial proliferation of the submucosal arterioles. A diagnosis of Degos’ disease was made. The patient had a favorable outcome with enteral and parenteral nutritional support and octreotide injections.

Degos’ disease (malignant atrophic papulosis) is a rare vasculitis syndrome described in Caucasians, characterized by infarction of small-caliber arterioles of the dermis, central nervous system and gastrointestinal submucosa. Infarction of subconjunctival vessels may lead to focal scleral thinning. The majority present with widespread asymptomatic skin rash, followed within days, weeks or even years by an acute abdomen or cerebrovascular accident. Failure to recognize the rash before laparotomy may lead to failure to recognize the cause of acute abdomen.

References


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