Degos Disease: A Murderous Menace

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Sir,

A 56-year-old male was admitted in dermatology ward with multiple, widespread, nonpruritic, painless eruptions along with intermittent abdominal pain. He complained of constipation, nausea, vomiting, and weight loss of more than 10 kg in the past 6 months. He had undergone exploratory laparotomy for ileal perforation 2 months ago. There was no history of headache, dyspnea, fever, joint pain, diarrhea, blurred vision, hemiparesis, or any other systemic illnesses. Cutaneous examination revealed multiple, erythematous papules on trunk, neck, scalp, extremities, genitalia with sparing of palms and soles [Figures 1 and 2]. These were necrotic or showed white porcelain-like center with telangiectatic rim [Figure 3]. Systemic examination was normal. Differential diagnosis included Degos disease, lupus erythematosus, papulonecrotic tuberculid, and pityriasis lichenoides et varioliformis acuta. Routine blood and radiological investigations were normal. Laboratory investigations showed negative results for antinuclear antibodies, human immunodeficiency virus, hepatitis B surface antigen, tuberculosis, and syphilis. Coagulation profile showed raised plasma fibrinogen, D-dimer levels, and prothrombin time of 1.28 INR.

Four days after admission, he developed acute abdominal pain and vomiting. Per abdomen examination revealed guarding, rigidity, tenderness, and distension. X-ray abdomen showed gas under the diaphragm, ultrasound showed abdominal free fluid with internal echoes indicating perforating peritonitis. Emergency exploratory laparotomy revealed jejunal and ileal perforations with Grade 3 adhesions. Surgical resection of perforated bowel segments with end-to-end anastomosis was performed. Skin biopsy showed epidermal atrophy, thrombosis of vessels, perivascular lymphocytic infiltrate, and dermal sclerosis. There was no granuloma formation, vasculitis, or atypical cells [Figure 4]. Intestinal biopsy showed hemorrhagic necrosis with inflammation. A final diagnosis of Degos disease was made. Recurrent perforation on 22nd postoperative day warranted a repeat laparotomy with surgical resection-anastomosis. Broad spectrum antibiotics, subcutaneous low molecular weight heparin, intravenous pentoxifylline, supportive care was instituted. However, he succumbed to the disease. The patient died within 6 months of the appearance of skin lesions and 2 months from the third laparotomy.

Degos disease is an extremely rare endotheliopathy. Most cases are sporadic, but few are familial with autosomal dominant inheritance. Etiopathogenesis remains unclear. Endotheliopathy due to the involvement of stromal cell-derived factor-1/CXCL-12 leading to platelet activation and thrombosis of vessels is a proposed mechanism. Other mechanisms include coagulation defects, autoimmunity, poststreptococcal vasculopathy, and parvovirus B19-induced catastrophic endothelitis.

Degos disease manifests as the classical type with systemic involvement (malignant atrophic papulosis...
Correspondences with fatal outcome) or benign cutaneous type without any systemic manifestation for at least 3 years.[2] Classical Degos can be due to autoimmune, coagulopathy, or viral factors. Characteristic cutaneous lesions reveal structure-less nonpigmented center surrounded by interweaved hairpin and telangiectatic vessels in a “crown of thorns” arrangement on dermatoscopy.[1] Gastrointestinal, cardiopulmonary, renal, central, and peripheral nervous system can be affected.[4] Anti-platelet agents dipyridamole and aspirin are helpful in cutaneous disease. In systemic disease heparin, warfarin, azathioprine, methotrexate, cyclosporine, and pentoxifylline have been tried, but fail to halt disease progression. Newer modalities include eculizumab (monoclonal antibody against C5 complement) and treprostinil.[5,4] To conclude, a classical Degos disease (typical cutaneous morphology, genital lesions, gastrointestinal involvement, coagulopathy as a cause, and a fatal outcome) has been reported for its rarity.

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Conflicts of interest
There are no conflicts of interest.

References
1. Meephansan J, Komine M, Hosoda S, Tsuda H, Karakawa M, Murata S, et al. Possible involvement of SDF-1/CXCL12 in the pathogenesis of Degos disease. J Am Acad Dermatol 2013;68:138-43.
2. Heymann WR. Degos disease: Considerations for reclassification. J Am Acad Dermatol 2009;61:505-6.
3. Darwich E, Guilabert A, Mascaró JM Jr., Malvehy J, Puig S, Fernandez-Figueras M, et al. Dermoscopic description of a patient with thrombocytemia and factor V Leiden mutation-associated Degos’ disease. Int J Dermatol 2011;50:604-6.
4. Ali YN, Hamed M, Azita N. Lethal systemic degos disease with prominent cardio-pulmonary involvement. Indian J Dermatol 2011;56:564-7.
5. Magro CM, Wang X, Garrett-Bakelman F, Laurence J,
Shapiro LS, DeSancho MT. The effects of eculizumab on the pathology of malignant atrophic papulosis. Orphanet J Rare Dis 2013;8:185.

6. Shapiro LS, Toledo-Garcia AE, Farrell JF. Effective treatment of malignant atrophic papulosis (Köhlemeier-Degos disease) with treprostinil – Early experience. Orphanet J Rare Dis 2013;8:52.

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