VASCULAR DISEASE, VASCULITIS

PORCELAIN WHITE SKIN: PRETTY OR FATAL?

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Background: Degos disease is a rare, progressive vasculo-occlusive disease of unknown etiology affecting small and medium sized arteries characterized by multiple infarcts in the skin and internal organs. It is classified as benign variant and the malignant variant. Classic Degos disease is rare, with about 200 reported cases, majority of the cases being sporadic.

Observation: A 35 year old male presented to the emergency room with complaints of fever, vomiting and acute pain in abdomen since 4 days with a history of recurrent abdominal pain since 1 year. A dermatology opinion was called for lesions on the trunk. On cutaneous examination, multiple skin coloured and erythematous papules with umbilication along with multiple porcelain white atrophic macules with surrounding erythema and telangiectasias were present on the trunk and proximal extremities.
A skin biopsy taken from the papule revealed wedge shaped zone of necrosis extending from the epidermis to the reticular dermis with mucin deposition. Deep dermal vessels were thrombosed with lymphocytes in the damaged vessel wall.
A plain chest X-ray revealed gas under the diaphragm following which the patient was taken up for an exploratory laparotomy. It revealed a perforation in the ileum. Ilectomy was done and histopathology revealed fibrin thrombi in the vessels with focal areas of necrosis. The patient developed acute respiratory distress syndrome and succumbed after 5 days.

Key message: Degos is a fatal disease with no consistently effective therapy. A high index of suspicion is needed to diagnose and manage the disease in early stages.