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DEGOS' DISEASE: A RARE CASE REPORT WITH REVIEW OF THE LITERATURE

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ABSTRACT
Degos' disease also known as malignant atrophic papulosis is an uncommon disease characterised by progressive vasculopathy leading to occlusion of small and medium sized arteries. It displays cutaneous as well as extracutaneous manifestations. Immediate causative factor may be disturbed endothelial function leading to abnormal coagulation. Effective treatment for the condition has not yet been found. We report a case of a 35 year old male presenting withhistory of generalised abdominal pain, vomiting and fever.Papular lesions associated with itching on the trunk noticed four days back. A diagnosis of Degos' disease with perforative peritonitis was given based on clinical and histological findings. We are reporting this case on account of the rarity of its occurrence.

INTRODUCTION

Degos' disease is a rare multisystem lymphocytic vasculitis in which there is widespread thrombosis of small vessels of skin, gastrointestinal, ocular and central nervous systems. However, usually the first feature is skin lesions and over many years, may be the only manifestation. The lesions in skin are usually erythematous, pink or red papules with central scar and peripheral telangiectatic rim [1]. In the systemic variant of Degos' disease, the most severe complication is intestinal perforation. There are less than 50 living patients of Degos' disease at present known worldwide, and less than 200 cases reported in medical literature [2].

CASE HISTORY

A 35 year old male patient presenting with fever since seven days, generalised on and off pain in abdomen since four to five days and two episodes of vomiting one day back. X-ray abdomen showed gas under diaphragm. Clinicoradiological diagnosis of perforation peritonitis in a suspected case of typhoid fever was given. Patient was admitted in intensive care unit.Biopsy tissue from the perforation site was sent for histopathological examination. Grossly it consisted of two tiny grey brown soft tissue bits each measuring 0.5 cm. On microscopy, mixed inflammatory cell infiltrate, dilated congested blood vessels, necrosis and haemorrhage were noted. Histopathological diagnosis consistent with the clinical diagnosis of perforation peritonitis was given.On further examination, he was also found to have papular lesions on the skin of trunk associated with itching. The papules had central white scars and peripheral telangiectatic rim. (Figure 1) Clinical diagnosis of Degos' disease was given. Skin biopsy from the lesion over chest was sent for histopathological examination. Grossly it consisted of a single skin covered grey white tissue bit measuring 0.4 cm. On microscopy, hyperkeratosis, finger like elongation of rete ridges, increased melanin in the epidermal basal layer, areas of mucin deposition in dermis and perivascular chronic inflammatory cell infiltrate were seen. (Figure 2 and 3) Histopathological diagnosis consistent with the clinical diagnosis of Degos' disease was given. Patient died on the fourth day due to septicemia.

Fig 1. Clinical photograph of skin lesion on chest showing papules with central white scars

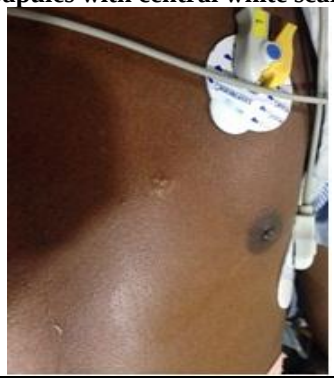


Fig 2. H&E stained section 4x showing skin with finger like elongation of rete ridges

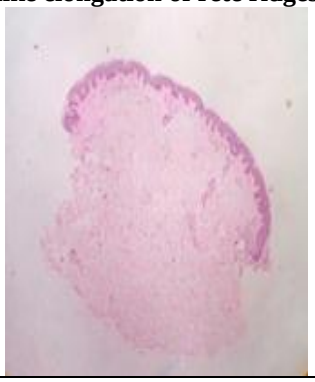
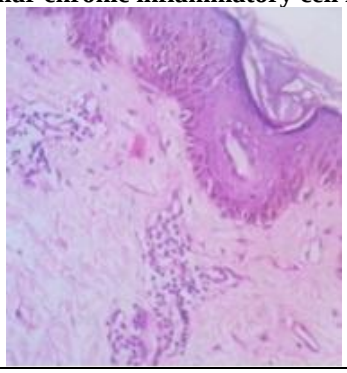


Fig 3. H&E stained section 10x showing skin with hyperkeratosis, increased melanin in the epidermal basal layer, and perivascular chronic inflammatory cell infiltrate.



DISCUSSION AND CONCLUSION

Kohlmeier first described malignant atrophic papulosis in 1941 [3]. Robert Degos recognized it as a separate entity in 1942 [4]. It is a vasculopathy or an endovasculitis. It is characterised by tissue infarction caused by progressive occlusion of small and medium sized arteries.It usually presents in adulthood with a 3:1 male predominance. In our case, the patient was an adult male.

Degos' disease may show two types of presentations- limited benign cutaneous form and a multiorgan, systemic, lethal form. The systemic involvement may involve organs of gastrointestinal system (50%), central nervous system (20%), ocular system, heart, lung, liver, kidneys [5]. Rarely, the systemic manifestations occur before the skin lesions while in most of the cases they develop weeks to year after the occurrence of skin lesions. In our case, the gastrointestinal system and skin were involved.

Gastrointestinal tract lesions may be asymptomatic or may produce symptoms like dyspepsia, nausea, vomiting, bowel movement and bleeding. It may also present as an abdominal emergency revealing the intestinal involvement and leading to perforation. In our case, the presenting symptoms were fever, vomiting and abdominal pain. Perforation peritonitis was proved in our patient.

Neurologic involvement causes manifestations like hemiparesis, aphasia, multiple cranial nerve involvement, monoplegia and seizures. Ocular manifestations include posterior subcapsular cataract, visual field defects, third cranial nerve palsies, papilledema and scleral plaques. In our case, no neurologic or ocular involvement was seen.

Skin lesions are usually pink or yellow papules healing with central porcelain white zone of atrophy. In our case, similar findings were seen.

In 2004, High et al. contended that Degos' disease may represent a common end point to a variety of vascular insults, many of which have not been fully elucidated rather than being a separate entity [6]. Bull *et al.* in 2003 proposed that it is just a variant of lupus erythematosus [7]. According to some authorities, it represents infarctive changes caused by primary endothelial cell defect with secondary thrombosis.

In 1997, Katz *et al.* described a familial variant of Degos' disease. Hence along with the patient, close follow up of family members is essential. To minimize the complications, signs of external and internal bleeding should be closely watched for early intervention. In our case, there was no familial involvement. No useful medical therapy is known for Degos' disease. In some patients with only cutaneous involvement, antiplatelet drugs may lessen the number of new lesions. Other treatment options are topical corticosteroids, sulphonamides, heparin,

methotrecate etc. In our case, no treatment was possible as the patient died in the hospital due to septicemia.

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Nil

CONFLICT OF INTEREST

The authors declare no conflicts of interest.

REFERENCES

1. Barriere H. (1978). Papulose atrophiantemaligne. *Ann Dermatol Venereol*, 105, 733.
2. De Breucker S, Vandergheynst F, Decaux G. (2008). Inefficacy of intravenous immunoglobulins and infliximab in Degos' disease. *Acta Clin Belg*, 63, 99-102.
3. Kohlmeier W. (1941). Multiple Hautnekrosenbeithrombangiitis obliterans. *Arch Dermatol et Syphilol*, 181, 783-4.
4. Degos R. Malignant atrophic papulosis. *Br J Dermatol*, 100, 1979, 21-36.
5. Waked M. (2008). Degos Disease: A Case Report and Review of Literature. *Egyptian Dermatology Online Journal*, 4, 5.
6. High WA, Aranda J, Patel SB, Cockerell CJ, Costner MI. (2004). Is Degos' disease a clinical and histological end point rather than a specific disease? *J Am AcadDermato,* 50, 895-9.
7. Ball E, Newburger A, Ackerman AB. (2003). Degos' disease: a distinctive pattern of disease, chiefly of lupus erythematosus, and not a specific disease per se. *Am J Dermatopatho*, 25, 308-20.