

Case report

Degos disease in a 24-year-old Jordanian male

R.M. Al-Smadi¹, F. Abu-Jamous¹ and I. Omeish¹

Introduction

Degos disease (malignant atrophic papulosis) is a rare disease that affects skin and the gastrointestinal, ocular and central nervous systems. The disease is characterized by papules that develop porcelain-white centres and telangiectatic borders.

The etiology of the disease is unknown; however, the immediate cause may lie in impaired endothelial function or abnormal coagulation (fibrinolytic activity). Viral causes have been implicated. Histopathological findings are wedge-shaped areas of dermal necrosis covered by markedly atrophic malpighian layers and collagen with a smudged appearance.

Complications of the disease include peritonitis, intestinal perforation and, less frequently, cerebral infarction. There is no effective treatment; however, antiplatelet therapy may be beneficial for patients in whom platelet aggregation is impaired.

Prognosis is poor when systemic involvement is found, but seems to be good in benign forms.

Case history

A 24-year-old Jordanian male presented with papular skin lesions distributed over his trunk and proximal parts of extremities,

while his face, scalp and palms were not affected. The lesions had been present for 4 years with no other symptoms of the disease and no family history of similar illness. No other systems were found to be involved. Physical examination revealed erythematous papular lesions with atrophic porcelain-white centres distributed over the trunk and extremities (Figures 1 and 2).



Figure 1 Erythematous papular lesions on the trunk of the patient

¹Department of Dermatology, King Hussein Medical Centre, Amman, Jordan.

Received: 13/01/98; accepted: 25/01/99

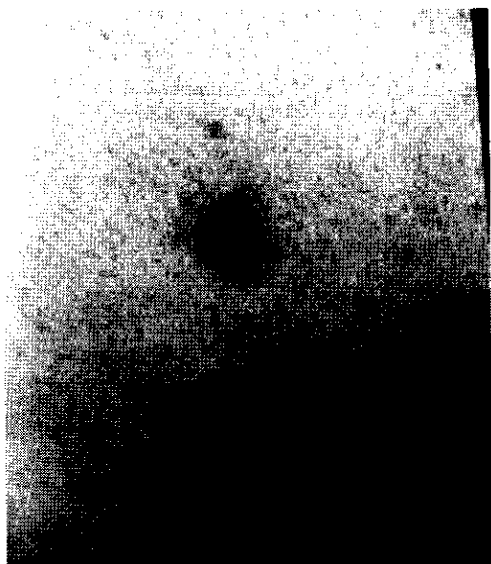


Figure 2 Closer view of erythematous papular lesion showing atrophic porcelain-white centre

Skin biopsy showed wedge-shaped areas of necrosis of the dermis and atrophic malpighian layers (Figure 3). In laboratory investigations, stool for occult blood was negative, and endoscopy and coloscopy showed no vascular lesions.

Discussion and conclusion

Degos disease was first described by Kohlmeier in 1941 [1] and was recognized as a specific entity by Degos and his associates in 1942 [2]. Only 114 cases of this rare disorder have been reported up to 1993 [1]. The disease affects the skin, gastrointestinal tract and, less commonly, the ocular and central nervous system. Local ischaemia may involve organs other than skin, such as the lungs, heart and kidneys. Intestinal lesions occur in 64% of cases and may manifest themselves from 3

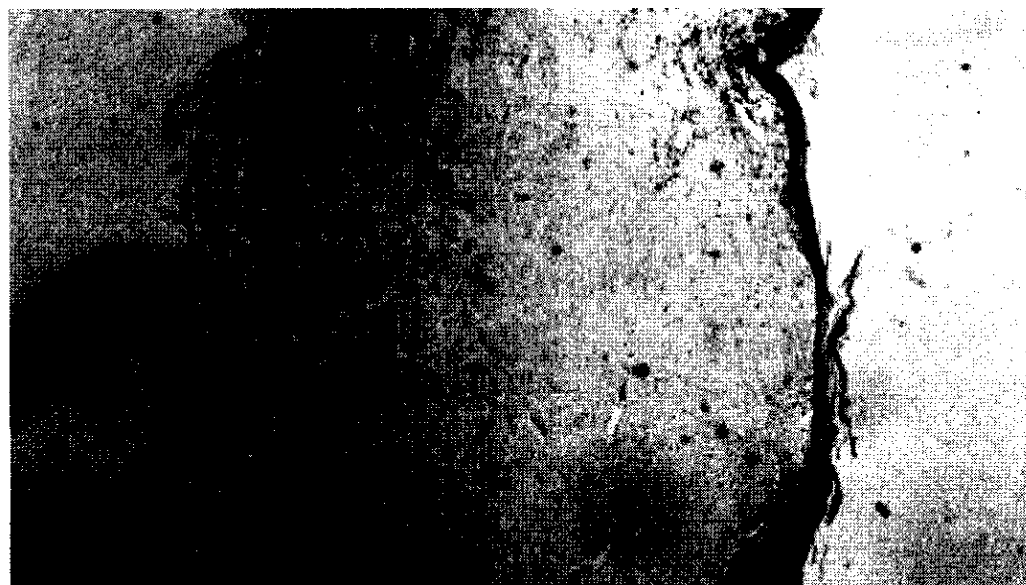


Figure 3 Histopathology of skin biopsy showing wedge-shaped areas of necrosis of the dermis and atrophic malpighian layers

weeks to 3 years after skin manifestation. A reported occurrence of Degos disease with non-systemic manifestation in three generations of one family argues for a genetic cause [3]. There are also reports of two cases involving impairment of blood fibrinolytic activity and alterations in platelet function that have responded to antiplatelet therapy [4]. Recently there has been a reported case of Degos disease with no visceral involvement in an AIDS patient [5].

Although our patient had had a history of skin lesions for 4 years, he had not yet developed systemic complications. He has been advised to report if any complications develop.

Degos disease is usually found to have systemic manifestation and a fatal outcome. However, there have been rare cases with isolated skin manifestation and good prognosis (benign form). Further studies are necessary to investigate triggering factors or genetic causes of this disease.

References

1. Dubertret L. Malignant atrophic papulosis (Degos' disease). In: Fitzpatrick TB et al. *Dermatology in general medicine*, 4th ed. New York, McGraw-Hill, 1993:1192-5.
2. Arnold HL, Odom RB, James WD. *Andrew's diseases of the skin: clinical dermatology*, 8th ed. Philadelphia, WB Saunders and Company, 1990:972-3.
3. Katz SK, Mudd LJ, Roenigk HH. Malignant atrophic papulosis (Degos' disease) involving three generations of a family. *Journal of the American Academy of Dermatology*, 1997, 37:480-4.
4. Vazquez-Doval FJ et al. Malignant atrophic papulosis: a report of two cases with altered fibrinolysis and platelet function. *Clinical and experimental dermatology*, 1993, 18:441-4.
5. Requena L, Farina K, Barat A. Degos disease in a patient with acquired immune deficiency syndrome. *Journal of the American Academy of Dermatology*, 1998, 38:852-6.