

# Cutaneous-limited Degos disease – benign variant or distinctive clinical entity?

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## Abstract

Degos disease is a very rare disorder of unknown aetiology. Pathognomonic cutaneous lesions are frequently complicated by systemic involvement that confers poor prognosis to the patient. Recently, more reports have been published on Degos disease with benign course and less unfavourable prognosis. We report another case of a young female patient presenting a cutaneous form of the disease with a nine-year course, which raises the question of whether systemic and benign, confined to the skin form of Degos disease represent two distinct entities or variants of one vascular disease.

**Key words:** benign Degos disease, malignant atrophic papulosis, prognosis.

## Introduction

Degos disease (DD), also known as malignant atrophic papulosis, is a rare systemic entity of unknown aetiology, characterized by an idiopathic vaso-occlusive process of small and medium-sized vessels leading to multiple infarctions in the skin and systemic organs, especially the gastrointestinal tract and nervous system. In such cases the risk of death is estimated to exceed 50% [1, 2]. Recently, more reports have been published suggesting that there may be a strictly cutaneous form of the disease without systemic involvement [3-8].

With this report, another case has been added to the group of benign, long-term Degos disease confirming the thesis of presence of a benign form of this disease.

## Case report

A 27-year-old Caucasian woman was admitted to the out-patient Ward of the Dermatology Department of the Medical University of Gdansk in 2006. In dermatological examination multiple (> 30), sharply separated, red-brownish papules 0.2-0.4 cm in diameter with a porcelain-white central atrophic scar were found. Lesions were observed on the trunk, thighs, buttocks and forearms (Fig. 1). First, single papules had appeared on the lower extremities five

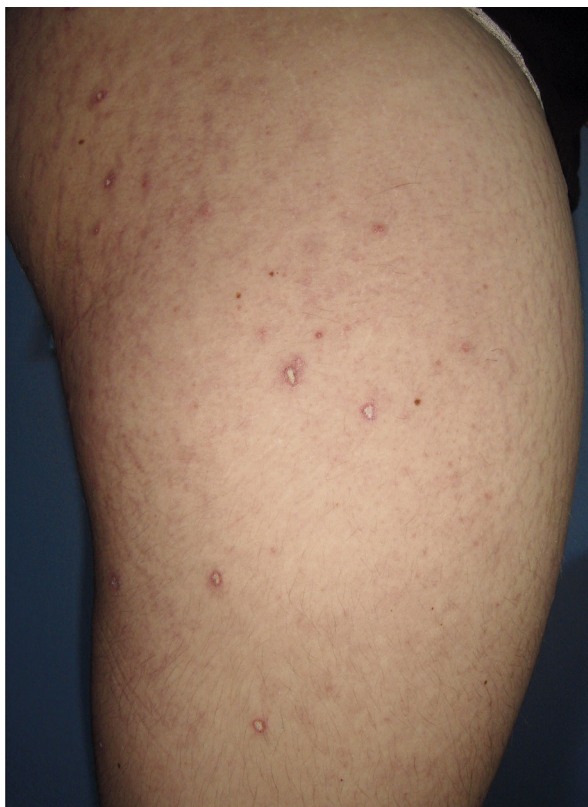
years earlier. Lesions located on sun-protected skin caused no subjective symptoms, which is why the patient refrained from seeking medical help. The patient's personal and family history was not contributory. She had no systemic complaints and did not take any medications. Biopsy of the papule from the forearm revealed a wedge-shaped area of sclerosis of superficial dermis resembling a reverse cone with slight lymphocytic infiltrate around. Flattened epidermis with marked hyperkeratosis, loss of basal cell layer and perivascular lymphocyte infiltrations within the dermis were observed (Fig. 2. A–C).

Based on the clinical and histopathological pattern of skin lesions of our patient the diagnosis of Degos disease was established.

Laboratory investigations including routine blood chemistry, fibrinogen, coagulation profile, full blood count, erythrocyte sedimentation rate, acute phase proteins, rheumatoid factor, cryoglobulins, antinuclear antibodies ANA Hep 2, ANA dsDNA, ANA SS-A/Ro, ANA SS-B/La anticardiolipin antibodies, antiphospholipid antibodies, anti-neutrophil cytoplasm antibodies (ANCA), serum immunoglobulins, complement components (C3, C4), *Treponema pallidum* and hepatitis serology, thyroid function test, and faecal blood test showed normal or negative findings. Direct immunofluorescent testing was negative.

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**Fig. 1.** Typical papules with porcelain-white atrophy surrounded by a rim of erythema on the right thigh

Chest X-ray, electrocardiogram, echocardiogram, and ultrasound of the abdomen were normal.

Our patient, still free of systemic symptoms, is aware of possible complications and is being closely reexamined for any progress of the disease. The patient has not received any medications for skin lesions. No new lesions have appeared during five years of follow-up and morphology of existing papules has not changed.

## Discussion

Degos disease was first described in 1941 by Köhlmeier, who believed that characteristic papular lesions are skin manifestation of Buerger's disease (thromboangiitis obliterans) [9]. One year later Robert Degos classified malignant atrophic papulosis as a distinct clinical entity and showed its systemic character [1]. Since that time the disease has been reported under the name of this author. Degos disease is an extremely rare entity; to date, approximately 200 cases have been reported in the literature [10]. The onset is usually between the second and fourth decade (mean age 35 years), with no gender predominance. Degos disease uncommonly affects children [1, 2, 11, 12].

The cause of DD has yet to be defined. There is no agreement among authors whether the main disturbance

in DD is vasculitis or coagulation abnormalities. According to Hight *et al.* Degos disease is not a separate entity but the end-point reaction for, in many cases, a not fully elucidated pathological process in vessels, found in different diseases with vascular infarctions. Currently, three major concepts of DD pathogenesis are taken into consideration: immunological (autoimmunological), infectious and connected with haemostasis disturbances. The association of DD with immunological disorders remains unclear. Lymphocytic infiltrations and lymphocytic vasculitis necrotisans present in the histological picture of the early phase of Degos disease may give evidence of the immunological background. Skin lesions characteristic for DD were described in patients with lupus anticoagulant and antiphospholipid antibodies [19, 20]. The presence of these antibodies may constitute pathogenetic linkage between Degos disease and lupus erythematosus (LE) [21]. Ball and Ackermann assumed Degos disease to be a variant of LE. They pointed out the similarity between the histopathological picture of DD and LE, especially some forms of LE, such as lupus tumidus and lupus profundus. In the microscopic picture of these two variants of lupus lymphocyte and plasmacyte infiltrate, hyaline degeneration and mucin deposits may be present [17, 21]. Other authors believe that the main cause of the thrombotic process in vessels in DD is coagulation disturbances [22-24]. Elevated fibrinogen level, increased platelet aggregation, presence of fibrin degradation products, changes in tissue plasminogen activator or its inhibitor concentration, and decreased blood fibrinolytic activity were observed in patients with DD. Another pathogenic theory – viral – was described in a publication on the association of Degos disease with HIV infection [25]. Moreover, a male patient with DD in the course of parvovirus B19 viraemia was reported. RNA viral transcripts were found in the endothelium of the described patient [26]. Genetic predisposition has also been discussed because familial cases of DD with autosomal dominant mode of inheritance were observed [5-7, 27].

The cutaneous findings in DD are pathognomonic. Well-separated, erythematous, pink or red, painless papules, 3-5 mm in diameter with umbilicated central atrophy are the first manifestation of the disease. The eruption occurs more often on the trunk and proximal parts of extremities, although any body area may be involved. Face, palms and soles are usually spared. Papules may be rarely present on oral and genital mucosa. As a result of progressive necrosis papules evolve into typical porcelain-white scars with a cyanotic-reddish rim of delicate telangiectasia (*halo*). The number of lesions ranges from a few to several hundred [12, 28, 29].

The diagnosis of DD rests on clinical appearance and histological examination, which is the basic diagnostic tool in this disease. Routine histology demonstrates thickening of vessel walls due to proliferation of endothelium with thrombosis. Oedema and fibrinoid

necrosis of vessel walls are also seen. As a consequence of vascular changes, formation of a characteristic ischaemic wedge-shaped zone of necrosis with slight lymphocytic infiltrate is present. Slight or absence of inflammatory infiltration is the main feature differentiating Degos disease from other types of allergic and thrombotic vasculitis [10-12]. A similar histopathological picture is observed in the intestine, with the greatest intensity in the submucosa, and other organs involved by the process [12, 19]. Laboratory tests in cutaneous Degos disease are not contributory and it is difficult to estimate their value.

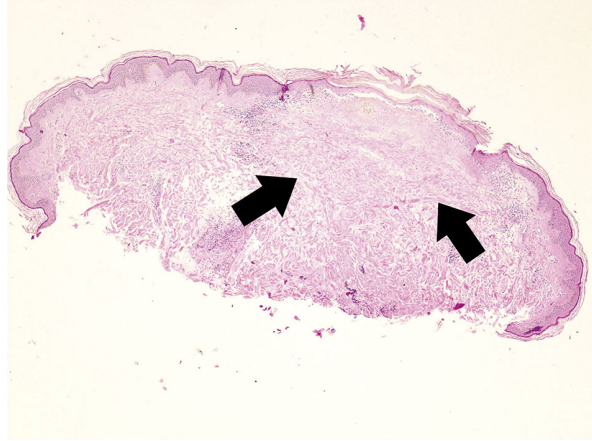
Because of the enigmatic pathogenesis of DD only empirical therapies have been tested. Anticoagulants (heparin, fibrinolytics) and anti-platelet drugs (aspirin, dipyridamole) may have a role in patients with abnormalities in coagulation tests [21]. Oral corticosteroids combined with cyclophosphamide, methotrexate, azathioprine or intravenous immunoglobulins have been tried without success [14, 22, 30].

It is difficult to predict the course of Degos disease. In most cases DD progresses into systemic disease. The period between onset of disease and occurrence of systemic symptoms is variable and ranges from several months to over a dozen years; mean duration is 30 months [7, 15]. Most often, in 50-61% of patients, the gastrointestinal tract is affected. Due to this fact Degos disease is termed by some authors disseminated intestinal and cutaneous thromboangiitis [1]. The eyes, cardiopulmonary system and nervous system are rarely involved. In cases with systemic symptoms the prognosis is grim. The cause of death is intestinal perforation (61%), neurological infarction (18%) and pleuritis [1, 15, 29].

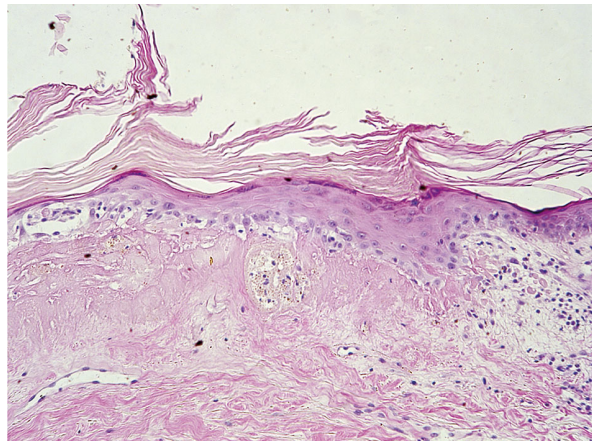
The first report on a benign variant of Degos disease was published over 20 years ago. Recently, more reports have appeared on Degos disease with benign, purely cutaneous course [3-8]. The authors are not consistent concerning the frequency of benign DD. Plantin *et al.* in a retrospective study of 120 patients with DD diagnosed the benign variant in 4% of patients [7]. Others estimate that this variant affects 15% of patients, being somewhat more frequent in females [31, 32]. The true figure may be even higher, because it is likely that the variant limited to the skin is underdiagnosed and underreported [4].

Heymann in 2009 suggested reclassification of Degos disease. He distinguished two variants of DD – classical disease with systemic manifestations and benign cutaneous form. The systemic variant was subclassified as autoimmune, coagulopathy-associated and provoked by viral infection. According to the author the diagnosis of benign variant is justified in patients with no abnormalities in laboratory tests and no systemic symptoms for three years of the disease [33].

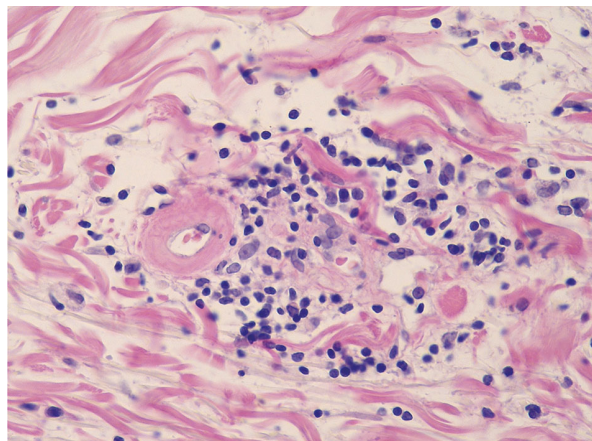
The present patient with a benign nine-year course of the disease appears to confirm that there is a strictly cutaneous, life-long form of DD. Only a few cases of



**Fig. 2. A.** Histopathological picture of papule. A wedge-shaped area of sclerosis of superficial dermis (arrows) resembling a reverse cone with slight lymphocyte infiltrate around. H&E × 20



**Fig. 2. B.** Flattened epidermis with loss of basal cell layer abutting hypocellular and sclerotic dermis resembles the appearance of lichen sclerosis, H&E, × 200



**Fig. 2. C.** Small vessel with thickened wall presenting hyaline degeneration

benign disease with such a long course have been reported in the literature [3, 7, 10].

Nevertheless, it should be emphasized that the absence of systemic symptoms for three years should be regarded as a positive prognostic factor with caution as, according to the literature, there is a risk of systemic progression even after many years of the disease being confined to the skin [2, 10, 12, 33].

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