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MALIGNANT ATROPHIC PAPULOSIS (DEGOS' DISEASE) : REPORT OF A RAPIDLY FATAL CASE

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A 21 year old Iranian man was seen at our department in November 1993 because of the eruption of multiple skin lesions on his trunk and upper arms. The lesions had first appeared six months before and were asymptomatic. His past medical history was unremarkable and there was no family history of a similar disease. On physical examination he had several erythematous papules and plaques on his chest, abdomen, back and upper arms (figures 1,2). Most of the lesions had central white porcelain-like zone of atrophy with a peripheral rim of erythema and telangiectasia (figure 3). No lesions were found on his scalp, face, palms, soles and mucous membranes. The patient was otherwise in complete health.

A biopsy taken from one of his lesions showed spongiosis and exocytosis in lower epidermis. Fibrinoid and mucin deposition ad lymphocytic infiltration were seen around blood vessels and eccrine sweat glands in middle and lower dermis. No wedge-shaped necrosis was present. Based on the pathognomonic skin lesions and histopathologic findings compatible with early lesions, the diagnosis of malignant atrophic papulosis (Degos' disease) was made.

Laboratory investigations including complete blood count, erythrocyte sedimentation rate, routine blood chemistry analysis, platelet function, fibrinolytic activity, brain CT-Scan and gastrointestinal radiology were normal at that time.

In May 1994, a year after begining of his disease, ataxia as the first neurological symptom appeared. In a few weeks paraparesis, diplopia, ptosis, fixed mydriasis on left side and clonus also developed. At that time brain CT-Scan and MRI were normal, HIV serology was negative, cerebrospinal fluid showed pleocytosis and increase in protein content.

The patient was treated with aspirin (325 mg daily) and dipyridamole (50 mg three times a day) but it was ineffective and the patient died in full paralysis in August 1994 (16 months after begining of his disease).

Discussion :

Malignant atrophic papulosis (MAP) or Dego's disease is a very rare vasculopathy of an unknown origin. Less than 150 cases of this disease have been reported so far. MAP usually begins with pathognomonic skin lesions but several other organs can also be involved, among them the gastrointestinal tract and central nervous system are the most frequently affected (61 and 20 percent, respectively)[1]. Skin lesions without systemic involvement and systemic involvement without skin lesions have rarely been reported[2,3]. Dego's disease is fatal in about half of the cases primarily due to intestinal perforations and central nervous system infarcts[1].

The pathogenesis of malignant atrophic papulosis is unknown. Decreased fibrinolytic activity and platelet dysfunction have been reported in some patients[4,5]. Also a few cases of MAP have been associated with anticardiolipin antibodies and the lupus anticoagulant [6] and MAP has been considered as one of cutaneous manifestations of antiphospholipid syndrome[7]. But this association has not been confirmed in a recent study on a larger group of patients[8].

Although wedge-shaped necrosis has been considered as a characteristic histopathological finding in MAP[1], it was present in only three of the 27 biopsy specimens in nine patients[2]. This feature is usually found only in old, well-developped lesions and is not present in most early lesions (such as our case).

There is not any effective treatment for MAP with

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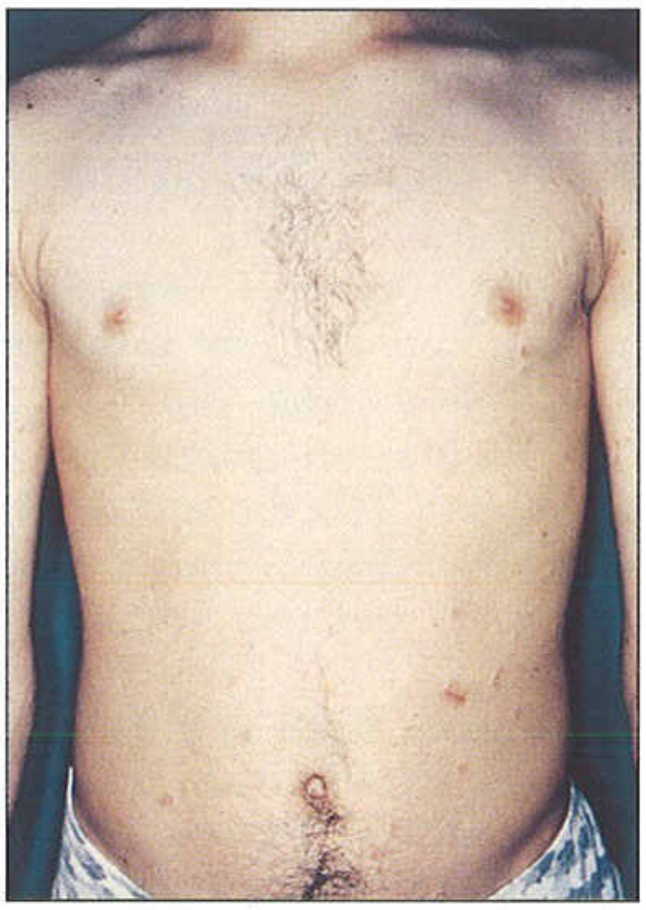

Figure 1 :Erythematous papules and plaques on the chest and abdomen of the patient

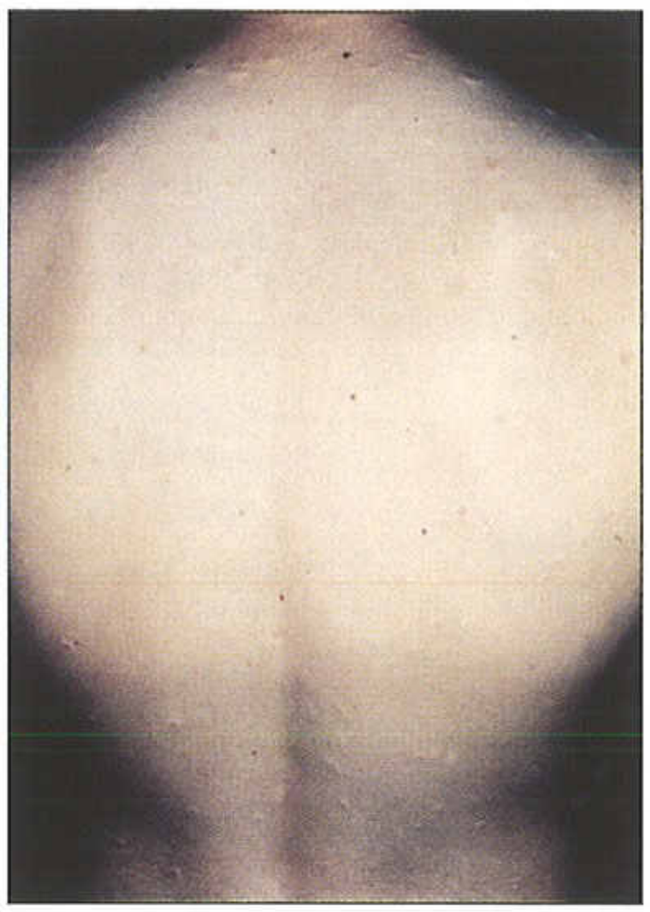

Figure 2 :Erythematous papules and plaques on the back of the patient

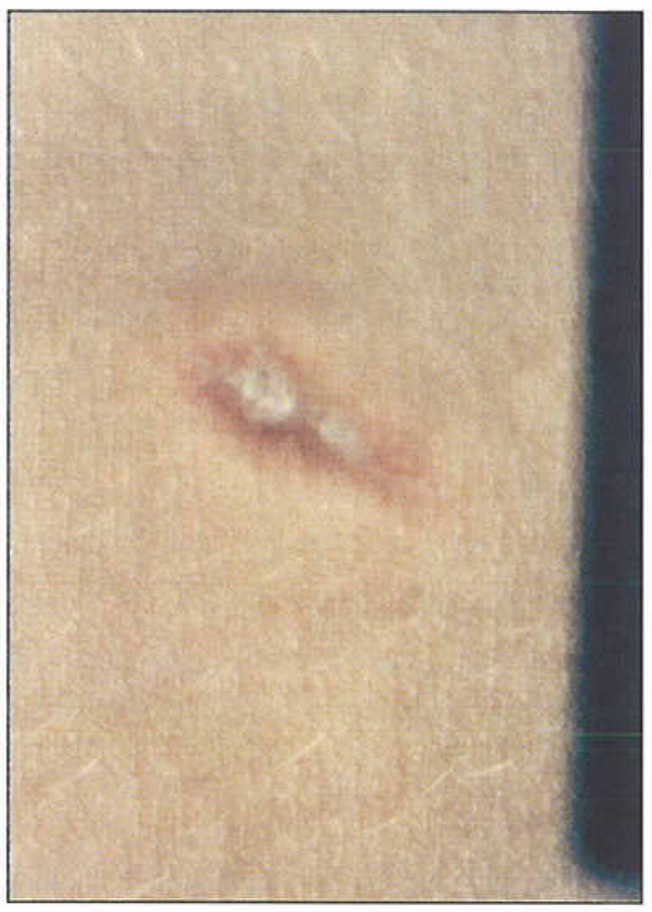

Figure 3 :Close up of a lesion with central white porcelain-like center and peripheral rim of erythema and telangiectasis

systemic involvement. Although anticoagulants, anti-platelets and fibrinolytics have been found useful in a few cases [9,10] but were ineffective in most cases (including our case) [11,12].

This case is the first case of MAP reported from Iran Who did not show any abnormalities in fibrinolysis and platelet treatment functionm did not respond to anti-platelet treatment and became repidly fatal due to the involvement of the central nervous system.

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