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Case Report

Degos' disease: a rare disorder

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Abstract Malignant atrophic papulosis (Degos' disease) is a rare multisystem lymphocytic vasculitis characterized by widespread thrombosis of small vessels not only in the skin, but also in the gastrointestinal, ocular and central nervous systems. We report a middle aged lady who has Degos' disease. We diagnosed her on clinical and histological ground.

Key words
Degos' disease, malignant atrophic papulosis.

Introduction

Malignant atrophic papulosis is a progressive vasculopathy causing occlusion of small and medium sized arteries.[1,2] It is characterized by skin and gastrointestinal lesions, but neurological features are also frequent. The skin lesions are usually the first feature, and may be the only manifestation over many years.[3]

The etiology of Degos' disease is not clear. It is a vasculitis, and a thrombotic disorder. In some cases, antiphospholipid antibodies are identified. In histopathology the early lesions show a superficial and deep perivascular, perineural and peri-appendageal chronic inflammatory cell infiltrate.[4] Later, lesions show a classical 'wedge-shaped' pattern of sclerotic change in dermis which is usually only sparsely cellular. In skin, Degos' disease manifests as erythematous, pink or red papules. These papules heal to leave central, porcelain white atrophic scars. These papules usually have a peripheral telangiectatic rim.[5] In the systemic variant of Degos' disease gastrointestinal tract with intestinal perforations is the most sever complication. Other systems e.g. CNS, ocular, cardiovascular and pulmonary can also be involved.[1] Skin lesions can be confused with guttate lichen sclerosus but the histopathology is diagnostic. No successful medical therapy for Degos' disease is known. Antiplatelet drugs e.g. aspirin, dipyridamol may reduce the number of lesions in some patients with only skin involvement.[6] Surgery may help in cases of intestinal perforation.

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Case report

A 50-year-old lady presented with history of papular lesions on her legs for the last 1 year. Initially these lesions started as small pink-coloured papules on thighs. Within a few days these lesion increased in number involving lower legs and abdomen. These lesions healed after some days and new lesions appeared. On examination small pink-coloured papular lesions were seen on legs and abdomen. Some of the lesions had central atrophy with white scars. No other systems were involved. Skin histopathology showed epidermal atrophy, a wedge shaped

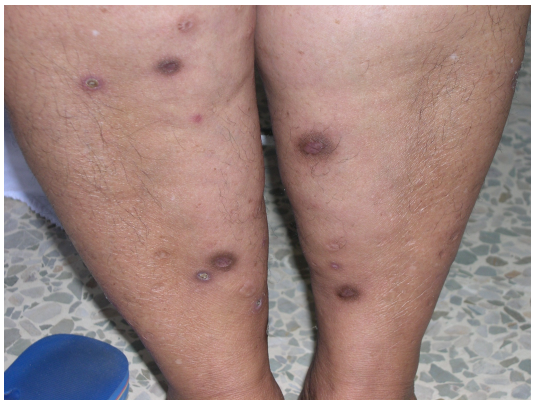

Figure 1 Multiple papular lesions over legs. Few of them show central atrophy.

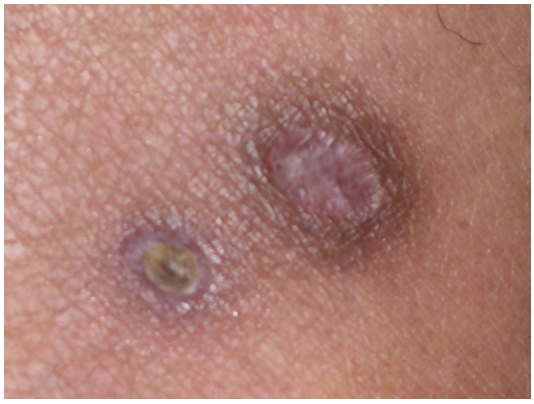

Figure 2 A close-up showing central atrophy and porcelain-white scales.

area of necrosis in dermis and vessels walls were thick. Other laboratory investigations like blood complete, ESR and coagulation profile were normal.

Discussion

Malignant atrophic papulosis (Degos' disease) was first described by Kohlmeier in 1941[17] and recognized as a specific entity by Degos in 1942.[18] It is a rare disease. About 150 cases have been reported in the world literature. Broadly speaking Degos' disease is a vasculopathy or an endovasculitis.[7] It is a progressive, small and medium-size arterial occluding disease, leading to tissue infarction.[8] Degos' disease occurs both in a limited benign cutaneous forms and in a lethal multiorgan, systemic variant.[9] In systemic variant of Degos' disease the gastrointestinal tract is affected in 50% of cases.[10] 20% of cases of systemic Degos' disease involve the CNS. Systemic manifestation usually develop from weeks to year after the onset of the skin lesion, or in rare instances, they may precede the skin lesions. Because of the broad overlap in clinical and histological findings, High *et al*. contended in 2004 that Degos' disease may not be a specific entity but, rather may represent a common end point to a variety of vascular insults, many of which have not been fully elucidated.[11] In 2003, Bull *et al*. proposed that Degos' disease is just a variant of lupus.[8] Some authorities suggest that Degos' disease involves a primary endothelial cell defect with secondary thrombosis, leading to infarctive changes. No evidence exists for antibodies to the component of endothelial cells. Although, in some cases antiphospholipid antibodies are isolated.[12] Three possible mechanisms have been suggested: disturbance in immunity, viral infection and abnormality in clotting system of blood. This disorder usually occurs in adults and male to female ratio is approximately 3:1.The cutaneous eruption is constant and pathognomonic. These lesions are pink or yellow papule and heal with typical central porcelain-white zone of atrophy. In our patient there were pink papular lesions as well as healed lesion with central porcelain-like zone of atrophy.

Gastrointestinal lesions usually occur a few months after the onset of skin lesions. GI tract involvement may remain asymptomatic. Sometimes, patient suffer only from dyspepsia, but usually it is an abdominal emergency that reveals the

intestinal involvement and leads to perforation.[1] The gastrointestinal tract is involved in about 50% of patients. In our patient the skin lesions were present for the last one year but there was no evidence of gastrointestinal complaint on history and examination. Neurologic manifestations of Degos' disease are observed in 20% of patients. They include hemiparesis, aphasia, multiple cranial nerve involvement, monoplegia and seizures.[13] In our patient there were no neurologic abnormalities. A variety of ocular findings occur in Degos' disease. In 1986, Sibillat *et al*.[14] reported that ophthalmologic symptoms were present in 35 of 105 observations published. They are posterior subcapsular cataracts, visual field defects, third cranial nerve palsies, papilledema and scleral plaques.[15] In our patient there was no eye involvement on ophthalmoscopic examination. In 1997, Katz *et al*.[16] described a familial variant of Degos disease. In our patient there was no family history of such disease. No successful medical therapy is known. Antiplatelet drugs may reduce the number of new lesions in some patients with only skin involvement. Other treatments include: topical corticosteroids pheniormin, iodohydroxyquinoline, sulphonamides, heparin, azathioprine, methotrexate, cyclosporine, tacrolimus and pentoxifylline.

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Authors Declaration

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The material or similar material has not been and will not be submitted to or published in any other publication before its appearance in the *Journal of Pakistan Association of Dermatologists.*