

Degos' disease in association with rheumatoid arthritis in a young boy

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Abstract Degos' disease (malignant atrophic papulosis) is an occlusive arteriopathy involving small-caliber vessels. Specifically, it is a progressive, small- and medium-size arterial occluding disease, leading to tissue infarction and initially involving the skin. The disease occurs both in a limited benign, cutaneous form and in a lethal multiorgan, systemic variant.

We report an 11-year-old boy who has Degos' disease. We diagnosed him on the clinical and histological ground.

Key words

Degos' disease

Introduction

Degos' disease (DD), also known as 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 cause of DD is unknown. Suggested causes include a virus, an immune defect, or a clotting defect. In some cases, antiphospholipid antibodies are identified.⁴

In the skin, DD initially manifests with erythematous, pink or red papules. These papules heal to leave scars with pathognomonic, central, porcelain white atrophic centers. These papules usually have a peripheral telangiectatic rim.⁵ In the systemic variant of DD, the gastrointestinal tract is affected in 50% of cases. Intestinal

perforation is the most severe complication and the most common cause of death in systemic DD. Other systems can also be involved; approximately 20% of cases of systemic DD involve the CNS. Systemic manifestations usually develop from weeks to years after the onset of skin lesions, or, in rare instances, they may precede the skin lesions. 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. In histopathology the early lesions show a superficial and deep perivasular, perineural and periappendageal chronic inflammatory cell infiltrate.⁴ Later lesions show a classical 'wedge-shaped' pattern of sclerotic change in dermis which is usually only sparsely cellular. No successful medical therapy for Degos' disease is known. Antiplatelet drugs e.g. aspirin, dipyridamol etc. 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

An 11-year-old boy presented with history of papular lesions on legs for the last 3 months. These lesions started as small pink colour papules on thighs. Within a few days these lesions increased in number involving lower legs and abdomen. These lesions healed after some days and new lesions appeared. On examination small pink colour papular lesions were present on legs and abdomen. Some of the lesions had central atrophy with white scars (**Figure 1**).

Patient also gave history of joint pains along with some degree of swelling. Arthritis initially involved the knee joints symmetrically, later ankle joints got involved with some degree of improvement in knee joint symptoms. Patient also gave history of morning stiffness lasting for more than an hour. Patient also gave history of off and on abdominal pain with no associated history of diarrhea, vomiting, anorexia or weight loss. There was no other complaint pertaining to any other system. Skin histopathology showed epidermal atrophy, a wedge shaped area of necrosis in dermis. Vessels walls were thick, with some vessels showing thrombosis which is the characteristic feature of disease (**Figure 2**). Rheumatoid factor was positive. Other laboratory investigations like blood complete picture, ESR and coagulation profile were normal.

Discussion

Malignant atrophic papulosis (Degos' disease) was first described by Kohlmeier in 1941¹⁷ 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



Figure 1 Papular lesions with central atrophy.

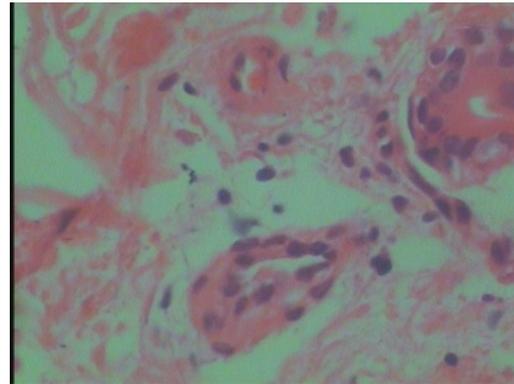


Figure 2 Vascular endothelial swelling and thrombosis.

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 form 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. The disease has occurred in patients with rheumatoid arthritis,¹¹ HIV infection, and antiphospholipid antibodies and antiphospholipid syndrome. Systemic manifestation usually develop from weeks to year after the onset of the skin lesions, or in rare instances, they may precede the skin lesions. Because of the broad overlap in

clinical and histological findings, High *et al.*¹² contended in 2004 that DD 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. In 2003, Ball *et al.*⁸ proposed that DD is just a variant of lupus. 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 component of endothelial cells. Although, in some cases antiphospholipid antibodies are isolated.¹³ In 2005, the author adduced strong evidence that DD is a distinct condition because, unlike lupus, (1) it does not involve the face, (2) it does not respond to therapies such as corticosteroids that at least abate lupus, (3) it does not manifest with photosensitivity, (4) viral inclusions are present in some cells in patients with DD, and (5) systemic DD is universally fatal, usually within 1-2 years, whereas lupus (even if severe) takes years to be fatal (Scheinfeld, 2005). The etiology and the pathophysiology of DD are unknown. Some have classified DD as a vasculitis, a mucinosis, or a thrombotic disorder. In most cases, no circulating immune complexes, antiendothelial cell antibodies, or anticardiolipin antibodies are isolated. Although, in some cases, antiphospholipid antibodies of uncertain significance are identified, the actual physical damage to blood vessels involves, at least in part, impaired fibrinolytic activity and alterations in platelet function. Classifying DD as a vasculitis may not be appropriate because inflammation of the vessel walls is minimal and because immune complexes have not been found in the vessel walls. Three possible mechanisms have been suggested:

disturbance in immunity, viral infection and abnormality in clotting system of blood. In familial cases, an autosomal dominant mode of inheritance has been suggested, but this is uncertain. All ages are affected. The fatal systemic variant of DD can occur in children. This disorder usually occurs in adults and male to female ratio is approximately 3:1. In most cases, the first manifestation of DD is a skin rash. Some have stated that the disease involves the skin as the sole clinical manifestation in 37% of patients. The gastrointestinal tract is involved in about 50% of patients. The rash of DD develops slowly and is usually asymptomatic, but it may be accompanied by a slight burning sensation. The rash can arise anywhere except on the soles, the palms, and the face. The rash starts as pink or red papules that are 2-15 mm in diameter. The papules evolve into atrophic scars that are porcelain white. Sometimes, abdominal symptoms precede the skin rash, but this finding is uncommon. Gastrointestinal lesions usually occur a few months after the onset of fair skin lesion. GI tract involvement may remain asymptomatic. Sometimes, patients experience abdominal pain. Some patients suffer only from dyspepsia, but usually it is an abdominal emergency that reveals the intestinal involvement and leads to perforation.¹ 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. Neurological manifestations of Degos' disease are observed in 20% of patients. These include hemiparesis, aphasia, multiple cranial nerve involvement monoplegia and seizures.¹⁴ In our patient no neurological abnormalities were found. A variety of ocular findings occur in DD.

Posterior subcapsular cataracts, visual field defects, ptosis, third cranial nerve palsies, blepharoptosis, and optic atrophy may be associated with DD. Optical neuritis, papilledema, and scleral plaques can be present. In 1986, Sibillat *et al.*¹⁵ reported that ophthalmologic symptoms were present in 35 of 105 observations published. Eye findings include posterior subcapsular contracts, visual field defects, third cranial nerve palsies, papilledema and scleral plaques.¹⁶ In our patient there was no eye involvement on ophthalmoscopic examination. Constrictive pericarditis has been reported in DD. The lungs can be affected in DD. Pulmonary manifestations include pleuritis and bilateral pleural effusions. The liver and the kidneys may be involved and associated with a vasculitis. In 1997, Katz *et al.*¹⁷ described a familial variant of Degos disease. In our patient there was no family history of such disease. No specific laboratory test can be used to aid in diagnosing DD. In fact, most laboratory test results are normal, with the exception of the manifestation of anemia secondary to intestinal bleeding. Biopsy of the lesion is considered to be diagnostic. Early papules in DD are skin colored and can demonstrate a superficial and deep perivascular, periadnexal, and perineural chronic inflammatory cell infiltrate associated with interstitial mucin deposition. Biopsy results from the epidermis of the papules of DD can show a mild vacuolar interface reaction, and, at this early phase, the histologic appearances of DD can mimic tumid lupus erythematosus. Fully developed papules can be raised, with umbilicated porcelain-white centers and a surrounding erythematous rim. Histologically, these papules demonstrate wedge-shaped degeneration of collagen. An

interface dermatitis can be present but is often limited to the central portion of the tissue examined histologically. Additionally, squamatization of the dermoepidermal junction, melanin incontinence, and epidermal atrophy can manifest. No successful medical therapy is known. Antiplatelet drugs may reduce the number of new lesions in some patients with only skin involvement. Some believe intravenous immunoglobulin may have been tried without real effect. Other therapies include topical corticosteroids, pheniormin, iodohydroxyquinoline, sulphonamides, heparin, azathioprine, methotrexate, cyclosporine, tacrolimus and pentoxifylline. When gastrointestinal bleeding, intestinal perforation, bowel infarction, or intracranial bleeding occurs, proper surgical intervention is necessary.

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