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CASE REPORT

Degos' disease: a rare condition simulating rheumatic diseases

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Abstract Dego's disease is an uncommon thrombo-occlusive vasculopathy that presented with skin rash and thrombotic complications affecting internal organs that may simulate rheumatic diseases and may be brought to the attention of rheumatologists. We present here a case of a middle-aged woman who presented with acute bowel infarction, persistent fever, elevated inflammatory markers and reversed albumin/globulin ratio suspicious of systemic vasculitis clinically. The diagnosis of Dego's disease was made from the classical skin lesions which were pink to brown papules with central depression and surrounding violaceous rim that were distributed over the trunk and extremities. Histology showed typical wedge-shaped infarction in the affected organs with endothelial proliferation and occlusion by thrombus. Our patient was put on aspirin but suffered from recurrent bowel infarction 1.5 years later and eventually succumbed to septic complications.

Keywords Anti-phospholipid antibodies · Malignant atrophic papulosis · Systemic vasculitis · Thrombo-occlusive vasculopathy

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Case report

A 43-year-old Chinese woman was admitted to the surgical unit because of acute lower abdominal pain. She presented with high fever of 40°C and was in shock with a blood pressure of 90/70 mmHg. There were diffuse guarding and rebound tenderness over the abdomen. Fluid resuscitation, intravenous ceftriaxone (2 gm daily) and metronidazole (1.5 gm daily) were given. Computed tomography (CT) scan of the abdomen revealed free peritoneal gas and fluid at the pelvis and a small bowel perforation of 2.7 cm in diameter was found. Emergency laparotomy revealed perforation of the jejunum. Peritoneal lavage, partial small bowel resection and end-to-end anastomosis of the jejunum were performed. Subsequent peritoneal fluid and blood culture grew *Escherichia coli*. Despite a prolonged course of antimicrobial therapy, the patient suffered from persistent fever after the operation. Further, blood tests revealed normochromic normocytic anaemia (Hb 9.5g/dl), elevated white blood cell (WBC; 21.4×10^9/l) and platelet (726×10^9/l) counts, reversed albumin to globulin (A/G) ratio of 29/47 and elevated C-reactive protein (CRP) (9.8 mg/dl). Microscopic examination of the resected jejunum showed transmural inflammation with focal ulceration and haemorrhage. Vascular changes were seen in the medium-sized mesenteric veins and arteries including marked intimal thickening with severe diminution in the lumen diameter, accumulation of foamy histocytes and thrombosis in various stages of organisation and recanalisation (Fig. 1).

Rheumatologists were consulted for clinical suspicion of systemic vasculitis. CT scan of the abdomen showed patent superior and inferior mysenteric arteries with no feature to suggest vasculitis. Immune profile showed negative antinuclear, anti-double-stranded DNA and anti-neutrophil cytoplasmic antibodies, normal levels of C3 (155 mg/dl),

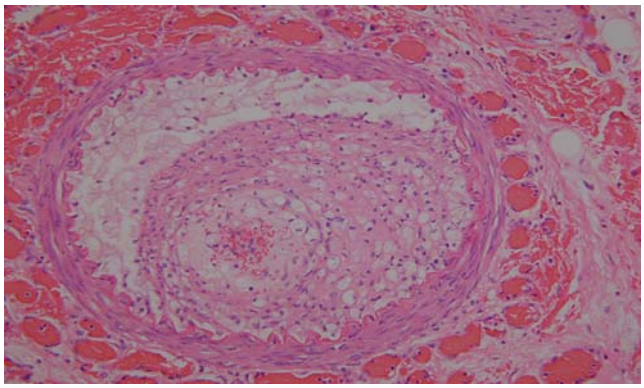

Fig. 1 Photomicrograph of the mesentery of the jejunum showing florid intimal hyperplasia of the medium-sized vessels. Accumulation of foamy histocytes is noted in the media of the medium-sized arteries. Several medium-sized veins show evidence of thrombosis with extensive recanalisation. Medium magnification (H&E)

C4 (15 mg/dl) and negative lupus anti-coagulant. The level of IgG anti-cardiolipin antibody was marginally raised (15 GPL/ml) but returned to normal on follow-up. On clinical examination, around 30 non-tender papular lesions, some with a central porcelain-white depressed zone surrounded by a telangiectatic rim, were noted over the face, trunk and limbs (Fig. 2). The cutaneous lesions and the recent history of intestinal perforation were highly suggestive of Degos' disease. An incisional biopsy of a skin lesion on the back showed a wedge-shaped zone of dermal infarction, the base of which was parallel to the epidermis. This zone is pale and relatively acellular and is associated with focal melanin pigmentary incontinence and deposition of mucin which showed positive alcian blue staining. The intact epidermis showed a vacuolar type interface dermatitis resembling the appearance of 'lichen sclerosus et atrophicus'. There were fibrin-platelet thrombi in some of the dermal vessels associated with a mild perivascular lymphocytic infiltrate. Immunofluorescence studies revealed no deposition of IgG, IgM, IgA, C3, C1q or fibrinogen.

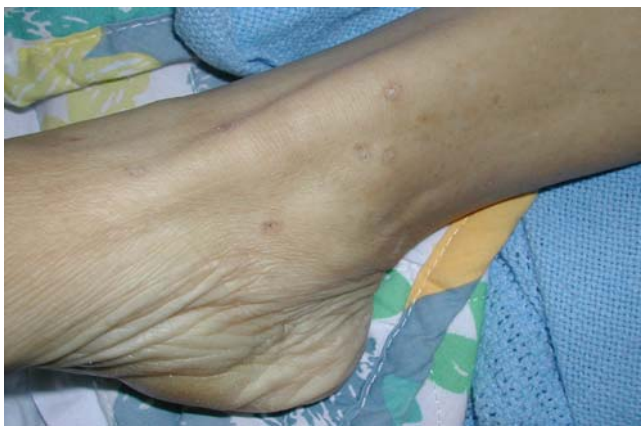

Fig. 2 Photograph of the skin lesions over the ankle showing discrete small patches composed of a depressed white central zone, fine scale, narrow red or violaceous rim with associated telangiectasia

Aspirin 100 mg daily was started. The use of an immunosuppressive agent was not considered in view of the post-operative sepsis and lack of proven efficacy. However, her compliance to aspirin was poor. The patient presented to the surgical unit again a year later. She complained of abdominal pain and vomiting with diffuse guarding on abdominal examination. A CT scan of the abdomen showed intra-peritoneal-free gas and oedematous small bowel wall. Emergency laparotomy was performed and another 2 mm perforation was found at the proximal jejunum. The perforated site was resected and the bowel was anastomosed end to end. Histology of the resected bowel showed similar ischaemic changes and perforation to those seen in the previously resected intestinal specimen. The patient had an anastomotic leakage requiring repeat laparotomy a week later. Further, bowel resection and side-to-side anastomosis were performed. On repeat, both IgG anti-cardiolipin (2 GPL/ml) and IgG anti-beta-2-glycoprotein 1 (8 G units) antibodies were negative. Despite the use of intravenous ceftriaxone 2 gm/day, the patient continued to suffer from persistent fever, chills and rigors after the operation. Subsequent abdominal CT scan showed an 8.0×2.1×6.0-cm collection in the left upper abdomen. A third laparotomy was performed with peritoneal lavage and drainage grew *E. coli*. The patient developed refractory shock and multi-organ failure and succumbed 1 week after the operation.

Discussion

We presented here an uncommon condition that may simulate rheumatic diseases and may be brought to the attention of rheumatologists. Degos' disease is a rare condition first described in 1942 by Degos and is characterised by thrombo-occlusive vasculopathy affecting skin and various internal organs [1]. Most of the reported cases are Caucasians in their third decade of life with a slightly higher prevalence in males [2]. Degos' disease has been described in two forms: the cutaneous and the systemic form. The systemic form has a poorer prognosis and is usually fatal within the first 2 years after diagnosis because of major organ involvement. Our patient had skin and gastrointestinal manifestations that are amongst the most commonly involved organs. Gastrointestinal involvement has been reported in 47% of patients and may manifest as dyspepsia, abdominal pain, gastrointestinal bleeding or bowel perforation [3]. Central nervous system involvement has been reported in around 20% of cases. Mortality occurs commonly as a result of intestinal perforation or cerebral infarction. Other sites of involve-

ment include the peripheral nervous system, heart, lung, eye, pancreas, adrenal gland and the kidneys [4, 5].

The diagnosis of Degos' disease is established on clinicopathological grounds. Skin lesions consist of numerous pink to brown papules up to 1 cm in diameter affecting the trunk and proximal extremities and they may appear in crops. With progression, they develop a characteristic appearance with central depression surrounded by violaceous rim. Histology of early skin lesions may be non-specific but mature lesions often show the typical wedge-shaped zone of infarction within the dermis associated with hyalinisation of collagen and increase in alcianophilic dermal mucopolysaccharide. There is usually minimal inflammation in established lesions although there were reported histology with lymphocytic infiltrate and necrotic microvascular alterations [6]. Histology of the bowel may show endothelial proliferation associated with partial or complete occlusion of the vessel lumen by thrombus. In most cases, the cutaneous manifestations precede the gastrointestinal and neurological involvement.

The pathogenesis of Degos' disease is not known. Immunological dysfunction, hyper-coagulation, fibrinolytic disturbances, genetic predisposition and viral infection have been proposed to be involved in the underlying pathogenesis [7]. Even though there was suggestion that Dego's disease is a vasculitic rash similar to systemic lupus erythematosus [8], it is generally believed that this condition is a vasculopathy rather than vasculitis. Presence of serum anti-phospholipid antibodies was reported in one case report [9]. Our patient had borderline elevated IgG anti-cardiolipin antibodies that was not present on repeat.

Rheumatologists were brought to the attention because of the clinical suspicion of systemic vasculitis in this case because of the presence of normochromic normocytic anaemia, elevated WBC and platelet counts, reversed A/G ratio, elevated CRP and focal bowel ischaemia. It is important for rheumatologists who encounter such patients to exclude other diseases with similar presentations, such as anti-phospholipid antibody syndrome and thromboangiitis obliterans, systemic lupus erythematosus, rheumatoid arthritis, systemic sclerosis, dermatomyositis and Crohn's disease [10]. The clinical appearance may also resemble other dermatological conditions including atrophie blanche and lichen sclerosus.

Our patient suffered from a relapsed thrombotic event after a period of 1.5 years from her first presentation. There has been no proven effective treatment for Degos' disease, at least in part due to the rarity of the condition. Antiplatelet agents including aspirin and dipyridamole have been inconsistently reported to reduce the formation of new skin lesions but the evidence in preventing systemic complications is lacking [11, 12]. Topical nicotine patches have been used with some success on cutaneous lesions only [13]. Immunosuppressants, anti-coagulants and plasma exchange have been shown to be ineffective. Prompt surgical intervention is often needed for bowel infarction, perforation or intracranial haemorrhage.

In conclusion, Degos' disease is a multi-systemic vasculopathy associated with life-threatening complications that may be brought to the attention of rheumatologists. It is important to recognise the typical cutaneous lesions and promptly treat the complications associated with this thrombo-occlusive condition that could be associated high mortality.

Disclosures None

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