Clinical Image

A Case of Minimal Change Nephropathy Overlapping the Köhlmeier-Degos Disease

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Abstract

The Malignant Atrophic Papulosis or Köhlmeier-Degos disease is a rare occlusive vasculopathy characterized by pathognomonic skin lesions and systemic involvement. The exact nature of this disease remains unclear. There are not reports on renal involvement in Köhlmeier-Degos disease.

We report a case of Minimal change Nephropathy overlapping the Köhlmeier-Degos disease.

INTRODUCTION

Malignant Atrophic Papulosis or Köhlmeier-Degos Disease (DD) is an occlusive vasculopathy with pathognomonic skin lesions and systemic involvement. Although most cases are sporadic, familial variant with an autosomal dominant pattern were also described. Until 2013 less than 200 cases have been reported. DD is a chronic and potentially life-threatening disease because of inner organs involvement in a severe and frequently progressive angiopathy syndrome. In addition to the skin, gastrointestinal tract, central nervous system, the lung and heart can be involved, there are no reports on renal involvement in DD.

CASE PRESENTATION

A 74-year-old previously healthy male was admitted to hospital because of nephrotic syndrome. Renal biopsy showed a minimal change glomerulopathy, without vascular lesions. Immunological and microbiological tests were unremarkable. Thorax-abdomen CT, colonoscopy and malignancy markers were negative for pathologic findings. Abdominal examination disclosed a DD pathognomonic skin lesion. We observed a 1cm large papule with atrophic porcelain-white centre and a surrounding erythematous rim. No lesions on the chest, back and extremities. Histopathological examination disclosed dermal necrosis without inflammatory infiltration, calcification and sclerosis in papillary dermis, blood vessel wall hyalinosis in deep layer of dermis. The patient didn’t respond to steroids and cyclophosphamide therapy. After 10 years of follow-up the skin lesion is unchanged. The patient has continuously been receiving support therapy for nephrotic syndrome without vasculitis signs or new skin lesions appearance.

DISCUSSION

Although DD has been known since 1941 its aetiology is still unknown [1]. The diagnosis of DD is based on the pathognomonic skin lesions (papular skin lesions with central porcelain-white atrophy and surrounding teleangiectatic rim). First manifestation is skin rash and often DD remains skin-limited (benign form), whereas others cases progress to systemic involvement (malignant form). DD-like skin lesions have occasionally been reported in autoimmune disorders but no specific laboratory alterations have been reported in DD [2-3]. Minimal inflammatory lupus-like changes have been described. Although many theories have been proposed (genetic factors, viral infection, vasculitis, associations with collagen diseases, primary endothelial cell disorder), the pathogenesis remains unclear [4-6].

Recently Magro and Coll. highlighted the thrombotic microangiopathy as a fundamental pathogenetic component to the vascular disease in DD [7]. In renal pathology, the thrombotic microangiopathy is the pathogenetic basis of haemolytic-uremic syndrome.

Two variants of DD have been described: one benign, confined to the skin, and one malignant, with Systemic involvement (bowel perforation, peritonitis, cerebral arteries thrombosis or haemorrhage, encephalitis, meningitis). Extracutaneous, systemic involvement includes multiple limited infarcts of the gastrointestinal system, central nervous system and other organs [8]. Approximately 50-60% of patients with systemic symptoms die within 2-3 years, mostly due to gastrointestinal perforation. Following Theodoridis et al., DD should be classified into a malignant, systemic form and a benign, cutaneous one and the latter is more common [9]. We believe that lack of vessel occlusion is a prognostic indicator of benign course of DD.
In the present case, we think that it exists an immunopathogenetic association between renal and skin lesions. Cutaneous antigens could have produced the characteristic changes of minimal change nephropathy.

CONCLUSIONS

DD is a chronic occlusive vasculopathy. The prognosis was determined mainly by the presence of systemic involvement. This evolution can develop suddenly or years after the occurrence of skin lesions. Therefore it is necessary to pursue a regular medical follow-up of the patients with DD.

REFERENCES