MICROSCOPIC POLYANGIITIS BEYOND PULMONARY-RENAL SYNDROME

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BACKGROUND

Microscopic polyangiitis is a rare systemic necrotizing autoimmune vasculitis, related to antineutrophil cytoplasmic antibodies (ANCA), with a perinuclear pattern, compromising small vessels. It usually presents itself with constitutional manifestations as well as skin vasculitis and pulmonary-renal syndrome. It is characterized by a rapid and progressive course when it is not diagnosed and treated quickly.

CASE REPORT

Male, 45 years old, brown-skinned, former smoker, referred to hospital in mechanical ventilation, with history of productive cough and progressive dyspnea which got worse a day prior. At admission he had renal dysfunction and a need for renal replacement therapy. The first hypothesis was pulmonary sepsis with secondary renal impairment due to the infectious process. A chest computed tomography (CT) was done, and it showed diffuse parenchymal infiltrates suggestive of fibrosis and cardiomegaly, with no alveolar hemorrhage (Figure 1). We proceeded with the investigation: echocardiogram, reduced ejection fraction (0.14), with severe compromise of the left ventricle, and dilated cardiomyopathy; positive ANCA with a perinuclear pattern (1:80); antimyeloperoxidase antibodies in high levels (203 UI). Due to the severity of the presentation and the patient's clinical instability at that moment it was chosen not to do a renal biopsy. As the diagnosis of microscopic polyangiitis was confirmed, we treated the patient at first with broad spectrum antibiotics due to multiple nosocomial infections ensuing, and then pulse therapy with methylprednisolone, followed by endovenous cyclophosphamide. He got clinically better, getting discharged from the hospital, but in need of long-term oxygen and regular renal replacement therapies. The patient was not followed by Rheumatology for 6 months. Coming back, due to the severity of his lung, renal and cardiovascular impairment, it was opted to start rituximab. After the second treatment cycle he presented with satisfactory response, with corticotherapy weaning, decrease in inflammatory markers and lesser need of long-term oxygen. He keeps follow-up with Cardiology, waiting for the possibility of a heart transplant.

Figure 1. Chest CT: diffuse parenchymal infiltrates and cardiomegaly.

CONCLUSION

We sought to present a case report of a patient with microscopic polyangiitis with pulmonary fibrosis, need of long-term oxygen therapy, renal replacement therapy and an atypical presentation of myocarditis and refractory heart failure, substantially compromising the patient's quality of life. We highlight the importance of a thorough clinic view on the diagnosis, broadening the spectrum of hypothesis facing less frequent clinical manifestations, and to quickly start treating to minimize target organs repercussions.

KEYWORDS

Microscopic polyangiitis, Pulmonary-renal syndrome, Vasculitis.