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PULMONARY-RENAL SYNDROME AS MANIFESTATION OF GRANULOMATOSIS WITH POLYANGIITIS: CASE REPORT.

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BACKGROUND

Granulomatosis with Poliangiitis (GPA) is a multisystemic vasculitis characterized by the predominance of small vessels with necrotizing granulomatous inflammation and proteinase 3 (PR3-ANCA) specific anti-neutrophil antibody (ANCA) antibodies association. Occurs with involvement mainly of upper and lower airways and kidneys with varying severity, which may evolve with isolated respiratory manifestations, but may also affect several organs simultaneously leading to a significant increase in morbidity and mortality. We report a severe case of GPA progressing with pulmonary-renal syndrome.

CASE REPORT

Patient, male, 20 years old, presented with abdominal pain in left flank, after 2 weeks evolved with irradiation to testicles, associated with fever, hyporexia, fatigue, nausea and vomiting, besides the presence of violaceous plaques lesions in back of the hands, knees and dorsum of the feet, slightly painful, and weight loss (6 kg in 2 months). During hospitalization, developed worsening of renal function (Creatinine: 6.5), requiring hemodialysis, and an acute dyspnea, hemoptysis, low hemoglobin levels, pleuritic pain, crepitations in both hemithorax, x-ray with infiltrate and diffuse condensation, suggesting diffuse alveolar hemorrhage. Evolved with hemodynamic instability necessitating vasoactive drugs and mechanical ventilation. P-ANCA non-reagent, C-ANCA reagent 1:80, anti-proteinase 3 (PR3-ANCA) reagent and anti-myeloperoxidase (MPO-ANCA) weakly reactive, FAN and anti-basement membrane antibody: non-reagent. He was treated with methylprednisolone, plasmapheresis and cyclophosphamide. Patient evolved with complete improvement of respiratory and cutaneous conditions, persisting on dialysis.

CONCLUSION

The term pulmonary-renal syndrome syndrome was first described by Goodpasture in 1919. This term is currently used to describe the association of pulmonary hemorrhage and rapidly progressive glomerulonephritis occurring in the context of a systemic autoimmune disease. The vasculitis ANCA associated is its main etiology, accounting about 50% of the cases.

The diagnosis of GPA is based on the symptoms, presence of ANCAs, with PR3-ANCA showing 99% specificity for the diagnosis. Histopathology also helps in the diagnosis and necrotizing granulomas with an associated vasculitis is the dominant feature. Due to the severity of our case, the patient could not be submitted to a biopsy.

Patients with pulmonary-renal syndrome from GPA are generally treated with corticosteroids combined with another immunosuppressive medication such as cyclophosphamide or rituximab. Plasmapheresis is indicated for patients with rapid progression to renal insufficiency, with concomitant anti-basement membrane disease or with alveolar hemorrhage. Despite the treatment, these patients can have a fulminant course with high mortality ranging from 25 to 50%.