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ANCA Vasculitis associated with IgA Nephropathy - A case report

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Abstract

Introduction : Neutrophil cytoplasmic antibody (NCA)-associated vasculitides are rare systemic disorders characterized by pauci-immune necrotizing glomerulonephritis with extracapillary proliferation. IgA nephropathy is characterized by the presence of mesangial IgA deposits, which can lead to rapidly progressive glomerulonephritis. The association of the two entities is rarely described.
Observation : We report the case of a 65-year-old patient admitted for generalized petechial purpura with macroscopic hematuria and renal failure, who underwent renal biopsy revealing IgA mesangial deposits nephropathy at an advanced stage of sclerosis. The patient was put on chronic hemodialysis. Six years later, he was hospitalized for the therapeutic management of pulmonary tuberculosis. After two months of antibacillary treatment, the patient presented with hemoptysis associated with extensive intra-alveolar hemorrhage. Immunological tests revealed highly positive p-ANCA with MPO specificity (>135 IU/mL). The patient was put on corticosteroids and Cyclophosphamide with complete remission, followed by Azathioprine as maintenance therapy.
Discussion: ANCA-positive patients with IgA nephropathy have a more severe clinical presentation and histological lesions, but a better response to treatment. The therapeutic management of this association has not been codified.
Conclusion: The association of IgA nephropathy and ANCA vasculitis is exceptional. One hypothesis suggests the presence of IgA-isotype ANCA, responsible for degranulation of polynuclear cells and interaction with mesangial cells. As the treatment of this association has not been codified, most of the protocols used are similar to those for ANCA vasculitis.

Keywords
IgA Nephropathy, ANCA Vasculitis, Association

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Main Article

Introduction:

Neutrophil cytoplasmic antibody (NCA)-associated vasculitides are rare systemic autoimmune disorders whose renal involvement is associated with pauci-immune necrotizing glomerulonephritis with extracapillary proliferation, without immunoglobulin deposition. IgA nephropathy is a glomerulonephritis characterized by deposition of IgA immune complexes associated with endo- and/or extracapillary proliferation, which can lead to rapidly progressive glomerulonephritis, and is rarely associated with ANCAs specific for myeloperoxidase (MPO) or proteinase 3 (PR3). We report the case of a patient with microscopic polyangiitis and IgA nephropathy.

Observation:

A 65-year-old patient, with no notable pathological history, was admitted following the appearance of generalized petechial purpura, predominantly in the lower limbs, associated with macroscopic hematuria. The biological work-up showed renal failure, and a renal biopsy was carried out, which indicated nephropathy with mesangial IgA deposits at an advanced stage of sclerosis. The patient was put on chronic hemodialysis. Six years later, the patient was hospitalized for the therapeutic management of pulmonary tuberculosis, bacteriologically proven by the presence of BK in the bronchoaspiration fluid. After two months of antibacillary treatment, the patient presented with haemoptysis, fever and general deterioration. Biological tests revealed normocytic normochromic anaemia at 7 g/dl and an inflammatory syndrome (CRP=80 mg/L, SV=60 mm). A chest CT scan showed a ground-glass appearance suggestive of intra-alveolar haemorrhage. Immunological tests revealed highly positive MPO-specific ANCA (>135 IU/mL). The patient was treated with corticosteroids and bolus cyclophosphamide, with azathioprine maintenance therapy. The course was favorable, with radiological clearance on follow-up chest CT and disappearance of ANCA.

Discussion:

The study by Rathman et al. of 270 patients with ANCA vasculitis following a respiratory infection showed that there was an association between this antecedent and ANCA vasculitides of MPO specificity. (1)

Another study by Yang et al. showed that the clinical and histological presentation of ANCA-positive IgA nephropathy patients was more severe than that of ANCA-negative patients.

However, a series of 20 patients treated with aggressive immunosuppressive therapy favored ANCA-positive patients with a better renal prognosis. (2).

In a retrospective study of 393 patients with IgA nephropathy, Bantis et al. found eight ANCA-positive patients. These were compared with 26 ANCA-negative patients (3). As in a series by Haas et al (4), those with positive ANCA had a more severe clinical presentation and histological lesions, but a better response to treatment.

Conclusion:

The association of IgA nephropathy and ANCA vasculitis is rarely described. One hypothesis is the appearance of ANCA vasculitis in a patient with an unknown pre-existing IgA nephropathy, given that the latter is common in the general population and only manifests clinically in a proportion of these patients. Another hypothesis suggests the presence of an IgA-isotype ANCA responsible for degranulation of polynuclear cells and interaction with mesangial cells. As the treatment of this association is not codified, the authors opt for a protocol similar to that of ANCA vasculitides.

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