

DOUBLE - POSITIVE VASCULITIS: A REPORT OF FIVE CASES

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Small vessel vasculitis with the presence of anti-neutrophil cytoplasmic antibodies (ANCA) and anti glomerular basement membrane (anti-GBM) antibodies are relatively rare and severe disorders with clinical courses different to when appearing separately. The clinical manifestation is characterized by a pulmonary renal syndrome with a rapidly progressive glomerulonephritis (RPGN) and diffuse alveolar hemorrhage (DAH). AntiGBM disease is known to be a non-relapsing disease, whereas ANCA vasculitis can relapse and supportive treatment is required.

The purpose of this study is to present the clinical evaluation and treatment of double-positive vasculitis.

We present five cases of double-positive vasculitis diagnosed in the nephrology clinic in 2013 - 2014. All patients were female, with an average age of 66 years (min. 48 max. 83). All of them presented RPGN, two of them were dialysis dependent, while another two needed temporary hemodialysis treatment. Hemoptysis was observed in 2 patients, whilst ground glass opacities in computer tomography were observed in 4. One of them needed mechanical ventilation. Circulating anti-GBM antibodies were detected in all patients (average level 126 U/ml, min. 20 U/ml, max. 281 U/ml). Anti-myeloperoxidase cytoplasmic antibodies (p-ANCA) were observed in 3 cases (average level 37,8IU/ml, min. 5,6 IU/ml, max. 70 IU/ml) and anti-proteinase-3 (c-ANCA) in 2 cases (average level 115 IU/ml, min. 38IU/ml, max. 192 IU/ml). Renal biopsies revealed necrotizing glomerulonephritis with crescents (> 50%) in all cases, and a linear deposition of IgG along glomerular basement membrane in cANCA cases or pauci-immune in pANCA cases.

Induction therapy including therapeutic plasma exchange (TPE) with an average number of 5 sessions (min. 3, max. 12), methylprednisolone followed by oral prednisolone and cyclophosphamide (CTX) were introduced in 4 patients. One patient, due to their advanced age and advanced stage of their illness, received TPE, glucocorticoids (GCS) and mycophenolate mophetil (MMF). GCS were used as a supportive treatment with either MMF or azathioprine (AZA).

Results

In all cases immunological remission was observed (ANCA and antiGBM antibodies were negative).

In all cases chest radiological abnormalities retracted.

Only two patients do not require dialysis treatment.

One patients continues dialysis treatment.

Two patients have died. One because of infection complication, another due to the advanced stage of the disease at the time of diagnosis.

Conclusion

Prompt diagnosis and treatment of dual-positive vasculitis is crucial and can improve long-term renal outcome and reduce mortality.

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WOJSKOWY INSTYTUT MEDYCZNY

