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ANCA (anti-PR3) and Anti-GBM Double-Positive Vasculitis: Case Report

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Abstract **Case Report**

This case highlights systemic vasculitis, an inflammatory condition that affects small blood vessels, presenting in two distinct forms: one linked to anti-neutrophil cytoplasmic antibodies (ANCA) and the other to anti-glomerular basement membrane antibodies (GBM). The study focuses on a 36-year-old patient with double positive vasculitis. Laboratory tests revealed the presence of both anti-glomerular basement membrane antibodies (anti-GBM) and anti-neutrophil cytoplasmic antibodies (anti-PR3). The renal biopsy confirmed extracapillary glomerulonephritis on proliferative glomerulonephritis, with C3 deposits and mild indications of chronicity. The patient's treatment included hemodialysis sessions, plasma exchanges, corticosteroid boluses, and cyclophosphamide after showing improvement with antibiotics. Considering the rarity and severity of this association, comprehensive clinical studies are crucial to improve therapeutic approaches and gain a deeper understanding of this intricate pathological condition.

Keywords: ANCA-associated vasculitis, anti-GBM vasculitis, double-positive vasculitis.

INTRODUCTION

Systemic vasculitis cover a heterogeneous group of diseases characterized by inflammatory cellular reactions involving vessels of various sizes. Among the primary vasculitis affecting small and/or medium-sized vessels, two distinct diseases stand out: those associated with anti-neutrophil cytoplasmic antibodies (ANCAs) and those associated with immune complex deposits composed of antibodies directed against the glomerular basement membrane (GBM). These conditions frequently involve the kidneys and/or the lungs, leading to the development of the feared clinical entity known as pulmonary-renal syndrome [1].

The combination of pulmonary hemorrhage and glomerulonephritis (GN) was first described by Goodpasture in 1919 [2], the term pulmonary renal syndrome (PRS) is used to describe a combination of diffuse pulmonary hemorrhage and GN occurring as the presenting manifestation of multisystem autoimmune disease [2].

The concomitant research of both antibodies along with the performance of renal biopsy has allowed, since the 1990s, the identification of double-positive patients (DPP), namely, patients who exhibit the coexistence of ANCAs and anti-GBM antibodies in small vessel primitive vasculitis [3, 4].

Among the primary systemic vasculitis types affecting small and/or medium size vessels, those associated with anti-neutrophil cytoplasmic antibodies (ANCAs) and anti-glomerular basement membrane (GBM) antibodies are two distinct diseases that share frequent involvement of the kidneys and/or the lungs, which can be associated in pulmonary-renal syndrome [5].

CASE PRESENTATION

We present the case of a 36-year-old female patient with a history of untreated angina for 2 weeks before admission, accompanied by inflammatory arthralgias. Her paternal grandfather and aunt passed away due to an unidentified nephropathy. The patient was admitted to our facility with a pulmonary-renal syndrome, presenting respiratory distress with two episodes of hemoptysis and anuria lasting for more than 3 days. She also exhibited general deterioration and feverish sensations.

On examination, the patient was conscious but dyspneic, experiencing orthopnea, with oxygen saturation (SaO2) of 94% on 6 liters per minute oxygen therapy via a high-concentration mask, dropping to 84% in ambient air. Bilateral crackling rales were heard at the lung bases. Her blood pressure was 120/70 mmHg, and her temperature was 38.2°C. Urinalysis showed

proteinuria (0+) and hematuria (3+), but no other significant abnormalities. Her urine output was negligible, indicating anuria.

Biological examinations revealed elevated creatinine levels (87.7 mg/l), high potassium levels (6.1 mmol/l), low hemoglobin levels (6.9 g/dl), significantly elevated D-dimers (>10,000), and a high C-reactive protein level (210 mg/l). The urine analysis showed a red blood cell count of 8,074,000 elements/mm3.

Both anti-glomerular basement membrane (anti-GBM) antibodies and anti-neutrophil cytoplasmic antibodies (ANCA) were positive, with values of 27 U/ML and 79 U/ML, respectively. Pulmonary scintigraphy revealed diffuse bilateral alveolar involvement, ruling out pulmonary embolism. Renal histology showed extra capillary glomerulonephritis on a background of proliferative glomerulonephritis with C3 deposits and mild signs of chronicity.

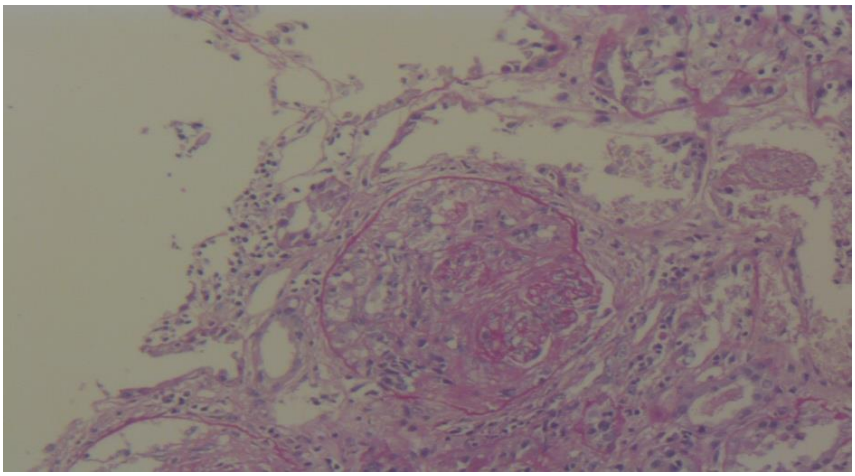

Figure 1: Optical microscopy: extra-capillary glomerulonephritis

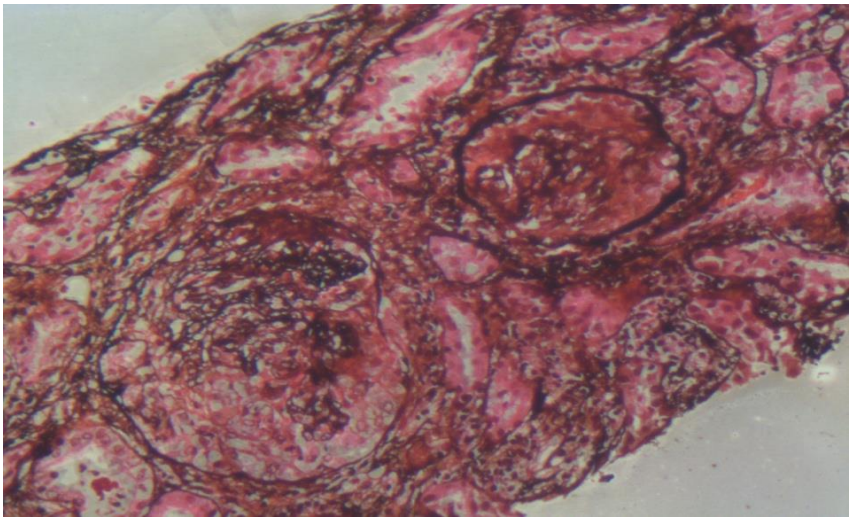

Figure 2: Optical microscopy: extra-capillary glomerulonephritis

TREATMENT AND EVOLUTION

Plasma Exchange with 3 sessions
Antibiotic Therapy
3 boluses of Solumedrol followed by an oral dose of 40mg/day
Intravenous bolus of Cyclophosphamide
Additional Plasma Exchange sessions

Respiratory condition showed significant improvement after the initiation of treatment, however, the patient remains anuric and dependent on hemodialysis for renal support.

The patient was initiated on intermittent hemodialysis and underwent three sessions of plasma exchange. She received antibiotic therapy and was administered three intravenous boluses of solumedrol, followed by an oral dose of 60 mg/day. Additionally, she received intravenous bolus of Cyclophosphamide and underwent further plasma exchange sessions. The respiratory condition improved significantly after the initiation of treatment, but the patient remained anuric and dependent on hemodialysis for renal support.

DISCUSSION

According to various studies, this clinical entity represents 30 to 50% of cases of anti-GBM vasculitis and 5 to 10% of cases of ANCA-associated vasculitis. Among double-positive patients, ANCA directed against myeloperoxidase (MPO) is found in 60 to 82% of cases, while rare cases of triple positivity (anti-GBM, anti-MPO, and anti-proteinase 3) have been reported [6, 7].

Stephen P. and colleagues observed comparable disease severity at presentation between single-positive anti-GBM and double-positive cases, with around 60% of patients requiring renal replacement therapy and one-third experiencing lung hemorrhage in both groups [7].

It is noteworthy that these double-positive patients tend to have a higher relapse rate than those with anti-GBM vasculitis but a lower rate compared to ANCA-associated vasculitis, with relapse rates of 22%, 0%, and 37% at 1 year, respectively [7].

CONCLUSION

Among the primary systemic vasculitis types affecting small and/or medium size vessels, those associated with anti-neutrophil cytoplasmic antibodies (ANCAs) and anti-glomerular basement membrane (GBM) antibodies are two distinct diseases that share frequent involvement of the kidneys and/or the lungs, which can be associated in pulmonary-renal syndrome [5].

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