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Antiglomerular basement membrane disease (Goodpasture syndrome) associated with ANCA-negative central nervous system vasculitis

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ABSTRACT

Anti-glomerular basement membrane disease is a rare pediatric autoimmune disease characterized by anti-glomerular basement membrane antibodies that cause rapidly progressive glomerulonephritis and alveolar hemorrhage. Central nervous system (CNS) involvement is sporadic. We present the case of a 13-year-old female patient with cerebral hemorrhage, septic *shock*, and multiorgan involvement, in whom the diagnosis of anti-glomerular basement membrane disease and CNS vasculitis in the absence of antineutrophil cytoplasmic antibodies (ANCA-negative) was confirmed. Combined treatment with glucocorticoids, cyclophosphamide, and plasmapheresis was performed, resulting in complete neurological and renal recovery.

The rare coexistence of these two entities is analyzed. The case highlights the importance of maintaining a high index of diagnostic suspicion in the face of atypical clinical presentations.

Keywords: *Goodpasture syndrome; central nervous system vasculitis; anti-glomerular basement membrane disease; anti-neutrophil cytoplasmic antibodies; pediatrics.*

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INTRODUCTION

Anti-glomerular basement membrane disease (anti-GBMD), also known as Goodpasture syndrome, is a rare autoimmune disease characterized by autoantibodies directed against the glomerular basement membrane and the alveolar membrane of the lung, leading to rapidly progressive glomerulonephritis and alveolar hemorrhage.[1] Although cases have been documented in which it has been associated with CNS vasculitis, such involvement is rare and extremely uncommon in the absence of antineutrophil cytoplasmic antibodies (ANCA).[2] The following is a case report of a patient with Goodpasture syndrome and CNS vasculitis with negative ANCA serology.

CLINICAL CASE

A previously healthy 13-year-old female patient was referred to the Hospital de Niños Ricardo Gutiérrez (HNRG) from another institution for cerebral intraparenchymal hemorrhage and septic shock secondary to cellulitis in the left thigh.

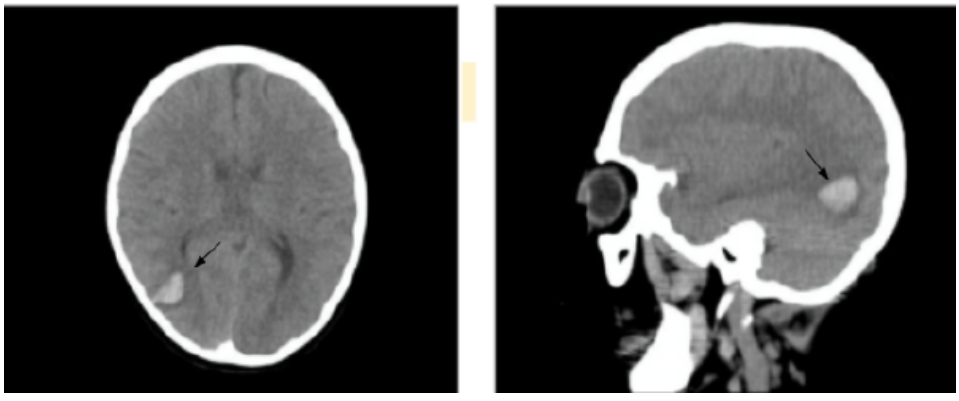
Initially, the patient presented to a health center near her home after a seizure. Upon admission, she was febrile and presented with cellulitis in her left lower limb; the condition was interpreted as septic shock of cutaneous origin. Blood cultures were taken, and empirical treatment with vancomycin, clindamycin, and amikacin was initiated. Subsequently, she developed acute sensory deterioration (Glasgow 8/15), requiring orotracheal intubation and mechanical ventilation (MV). A computed tomography (CT) scan of the brain was performed (*Figure 1*), revealing intraparenchymal hemorrhage in the right parieto-occipital region, and a chest CT scan showed bilateral interstitial involvement (*Figure 2*). Given these findings, it was decided to refer her to a more specialized center for neurosurgical evaluation.

Upon admission to the HNRG, she was admitted to the pediatric intensive care unit under MV, hemodynamically decompensated with a requirement for inotropics and under the effects of sedation and analgesia. The neurosurgical evaluation ruled out the need for surgical treatment.

The patient remained hemodynamically unstable with septic shock and a cutaneous focus of infection. Blood cultures were repeated and were negative; a culture from healthy skin in the affected area isolated methicillin-sensitive *Staphylococcus aureus* (MSSA). Multiple organ failure was observed with cardiac involvement (requiring inotropics, troponin 238 ng/L, and proBNP 36,664 pg/mL), renal failure (glomerular filtration rate estimated by Schwartz formula: 37 ml/min/1.73 m^2), abnormal liver function tests (ALT 245 U/L, AST 360 U/L), and coagulopathy (APTT 51%, PT 25 s). The condition was interpreted as septic shock with multiple organ failure secondary to MSSA infection. She initially received vancomycin, which was later switched to clindamycin, with a good clinical response, progressive reduction in inotropic agents, and improvement in cardiac enzyme levels, liver function tests, and coagulation tests.

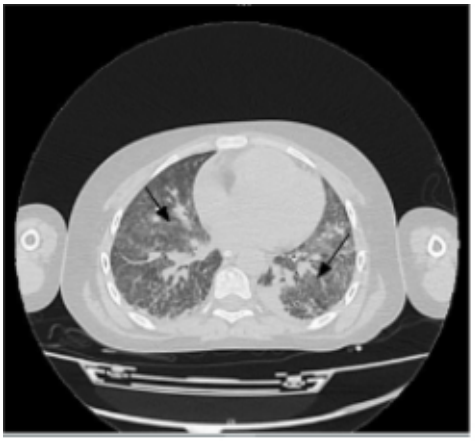
Given the need for inotropic agents, an

Figure 1. Computed tomography of the central nervous system



Right temporo-occipital corticosubcortical intraparenchymal hematoma.

Figure 2. Computed tomography of the chest



Interstitial involvement in both lung fields.

echocardiogram was performed, which showed moderate aortic insufficiency and dilation of the aortic root and descending aorta. The transesophageal echocardiogram revealed thickening of the right coronary artery and coronary refringence, findings suggestive of vascular inflammation. Meanwhile, renal failure initially attributed to shock showed partial improvement, raising the possibility of previously undiagnosed nephropathy. Additionally, upon reevaluation of the chest CT scan from the hospital of origin, bilateral interstitial involvement was noted (*Figure 2*). The combination of these findings, together with the cerebral hemorrhage, led to suspicion of an underlying systemic disease. A complete immunological profile was requested, including immunoglobulins, complement, antineutrophil cytoplasmic antibodies (ANCA), anti-DNA antibodies, perinuclear ANCA (p-ANCA), cytoplasmic ANCA (c-ANCA), antiproteinase 3 antibodies, anti-GBM antibodies, and antimyeloperoxidase (MPO) antibodies.

The patient subsequently developed pulmonary hemorrhage, and the clinical picture was interpreted as pulmonary-renal syndrome due to acute renal injury associated with pulmonary involvement. In conjunction with the Rheumatology and Nephrology Departments, it was decided to initiate immunosuppressive treatment with methylprednisolone pulses at 30 mg/kg/day for three days. Immunological studies yielded no positive results, except for the presence of anti-GBM antibodies, thereby confirming the diagnosis of anti-glomerular basement membrane disease. Plasmapheresis and cyclophosphamide were initiated.

Concerning neurological involvement, a brain magnetic resonance angiography with a black-blood protocol revealed scattered microhemorrhages in both cerebral hemispheres and the cerebellum, an ischemic lesion in the right putamen, and segmental narrowing with "beaded" appearance of the cerebral arteries, findings consistent with central nervous system vasculitis. Given these findings and the absence of other positive antibodies, ANCA-negative CNS vasculitis was diagnosed, suggesting an association between anti-glomerular basement membrane disease and ANCA-negative CNS vasculitis.

The patient underwent seven plasmapheresis sessions until anti-glomerular basement membrane antibodies became negative and six pulses of cyclophosphamide at 500 mg/dose, with a favorable clinical outcome. Subsequent laboratory tests were negative for anti-GBM antibodies, and renal function gradually improved, with a glomerular filtration rate of 94 ml/min/1.73 m^2. The patient continued outpatient follow-up at the Nephrology Department, with a progressive reduction in oral corticosteroids.

During the course of the disease, a percutaneous renal biopsy was performed, which revealed a pattern consistent with extracapillary glomerulonephritis with linear IgG deposits on immunofluorescence, thereby confirming the diagnosis of anti-glomerular basement membrane disease.

DISCUSSION

Goodpasture syndrome, also known as anti-GBM disease, is a rare autoimmune disease that primarily affects the glomerular and alveolar capillaries and is mediated by antibodies directed against the NC1 domain of the α3 chain of type IV collagen.[1] In pediatrics, it represents a rare cause of rapidly progressive glomerulonephritis, with a potentially fatal outcome if not treated early.[3] The classic presentation includes pulmonary hemorrhage and renal involvement, although the severity and progression can vary widely.[4]

In this case, the patient presented a characteristic triad of glomerulonephritis, pulmonary hemorrhage, and the presence of anti-GBM antibodies, which confirmed the diagnosis. The favorable response to immunosuppressive treatment and plasmapheresis underscores the importance of early intervention, especially in pediatric patients, who may achieve complete renal function recovery if therapy is initiated before irreversible damage occurs.[5,6]

What makes this presentation particularly interesting is the presence of CNS vasculitis in a patient with ANCA-negative Goodpasture syndrome. Although up to 30% of cases may present with some form of extrarenal vasculitis,[2] CNS involvement is rare and has been described in pumonary-renal syndrome, where circulating anti-GBM and ANCA antibodies coexist.[7] In these cases, CNS vasculitis is associated with ANCA, although CNS manifestations can occur even when these antibodies are negative.[8]

In the case presented, the patient had findings on magnetic resonance angiography consistent with cerebral vasculitis (segmental "beaded" narrowing and microhemorrhages). Despite the absence of ANCA, the diagnosis was supported by the clinical presentation, imaging findings, and the exclusion of other etiologies.

Although rare, the literature describes similar cases of association between Goodpasture syndrome and ANCA-negative CNS vasculitis. To date, five comparable cases have been reported in the medical literature.[2,9] Combined treatment with glucocorticoids, cyclophosphamide, and plasmapheresis has been shown to be effective in controlling both Goodpasture syndrome and associated cerebral vasculitis.[1,2] In this case, treatment resulted in complete neurological and renal recovery, with anti-GBM antibodies becoming negative and the glomerular filtration rate normalizing.

This case illustrates the importance of maintaining a high index of diagnostic suspicion in patients with of atypical clinical presentations. It highlights the need to consider autoimmune diseases in the context of multiorgan involvement. The association between Goodpasture syndrome and CNS vasculitis, even in the absence of ANCA, although rare, should be considered as a potentially serious and treatable systemic manifestation. Documenting such rare associations contributes to collective knowledge and may facilitate early identification in future cases. ■

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