Double the Trouble: A Rare Case of Pediatric Pulmonary Renal Syndrome

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Introduction: Pulmonary renal syndrome (PRS) is a rare autoimmune disease characterized by glomerulonephritis and pulmonary vasculitis. One cause of PRS is anti-glomerular basement membrane (anti-GBM) disease, estimated to occur in 0.5-1 per million adult patients, and is rarer still in children. A subset of patients with anti-GBM disease also have anti-neutrophil cytoplasmic antibodies (ANCA). We present a child who developed severe pulmonary hemorrhage due to anti-GBM disease with positive ANCA. Clinical presentation: An 11-year-old female presented to her pediatrician with two weeks of fatigue, dyspnea, and cough. She was hypoxemic to 68% on room air, placed on 20 LPM supplemental oxygen, and hospitalized. Chest x-ray revealed patchy opacities concerning for infection. Venous blood gas was normal and a complete blood count showed leukocytosis and hemoglobin of 9.4 g/dL. Urinalysis was notable for hematuria (117 cells/HPF). She was admitted to the pediatric intensive care unit, subsequently developed respiratory failure and was intubated. Due to her anemia with pulmonary infiltrates, an evaluation for diffuse alveolar hemorrhage was performed, revealing positive anti-GBM antibodies (>8 U), and positive anti-myeloperoxidase antibodies (43.1 U/mL) in a cytoplasmic pattern (c-ANCA). Creatinine, normal on admission, rose to 1.67 mg/dL. Her respiratory symptoms worsened requiring placement on extracorporeal membrane oxygenation (ECMO). She received six sessions of plasma exchange, intravenous (IV) pulse steroids (1 gram/day for 5 days) followed by daily oral steroids (60 mg) and one dose of IV cyclophosphamide (500 mg/m²). Due to her positive ANCA, she was also treated with two doses of IV rituximab (750 mg/m²), two weeks apart. She significantly improved with treatment and was discharged off supplemental oxygen with a normal creatinine. Discussion: Anti-GBM disease is a rare entity with few reported pediatric cases. A subset of adult patients (21-38%) with anti-GBM antibodies are also seropositive for ANCA, and are more likely to relapse. PRS with both anti-GBM and ANCA positivity has only been reported in nine children in the literature, two of which died and three developed end-stage renal disease (ESRD), highlighting the importance of early recognition and aggressive treatment. Due to these factors, we felt treatment with rituximab was indicated, and our patient responded well without evidence of persistent renal disease or pulmonary relapse two months after presentation. A patient presenting with fatigue, hypoxemia, and dyspnea with anemia and hematuria should raise concern for PRS. It is important to evaluate patients for both antibodies, as the presence of both changes long-term management.

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