11. Idiopathic Warm Autoimmune Hemolytic Anemia in a 5-Month-Old Infant: Navigating a Refractory Course

Shreya Gupta [1] Piali Mandal [2] Vandana Puri [3]

[1] Final-year Medical Student, Lady Hardinge Medical College, New Delhi, India [2] MD Pediatrics, DM Pediatric Oncology. Professor, Lady Hardinge Medical College, New Delhi, India [3] MD Pathology, DM Hematopathology. Professor, Lady Hardinge Medical College, New Delhi, India

BACKGROUND: Autoimmune hemolytic anemia (AIHA) in early infancy is an exceptionally rare entity, with idiopathic disease in infants under six months representing a distinct and poorly characterized subset. The overall annual incidence of AIHA in children under 18 years is estimated at 0.8 per 100,000; however, robust population-level incidence data are lacking for the idiopathic infantile subgroup. Evidence-based management guidelines for this age group are absent, with therapeutic decisions largely extrapolated from older pediatric and adult populations.
CASE PRESENTATION: A 5-month-old male infant presented with a 1-month history of progressively worsening pallor, jaundice, and dark-colored urine, with respiratory distress developing over the preceding 15 days. He had previously received multiple packed red cell transfusions at an outside facility without sustained improvement, and developed congestive heart failure secondary to refractory severe anemia prior to referral. On examination, the infant weighed 8kg, was markedly pale and deeply icteric, with tachycardia (194 beats per minute), tachypnea (68 breaths per minute) and hepatosplenomegaly. Admission hemoglobin was 2.4 g/dL, indirect bilirubin 7.33 mg/dL, LDH 1,228 U/L, and reticulocyte count 7%. Systematic evaluation excluded intracorpuscular causes of hemolysis. Immunohematological workup confirmed warm AIHA on the basis of a strongly positive DAT (3+) with IgG warm autoantibodies and a negative cold agglutinin titer. Exhaustive evaluation for secondary etiology, infectious, immunological, genomic, and marrow-based — was entirely negative, establishing primary idiopathic warm AIHA. Despite first-line treatment with methylprednisolone and IVIG, hemolysis persisted with only transient responses. Rituximab at 375 mg/m^2 weekly was introduced as second-line therapy, yet hemoglobin declined to a nadir of 2.3 g/dL with respiratory compromise necessitating mechanical ventilation and ICU-level management. In light of continued hemolysis, MMF at 20 mg/kg/day was added to the ongoing rituximab regimen. Gradual and sustained hemoglobin improvement followed, rising to 9.7 g/dL, reflecting the cumulative effect of stepwise multimodal immunosuppression. The patient was discharged on antibiotic prophylaxis, oral prednisolone, folic acid, and MMF. **CONCLUSION:** This case highlights two clinically important observations. First, that idiopathic warm AIHA in early infancy can remain without identifiable etiology despite exhaustive evaluation. Second, that it can pursue a profoundly refractory course necessitating stepwise escalation to multimodal immunosuppression to achieve hematological stabilization. These findings underscore a critical gap in evidence-based guidance for this age group and reinforce the imperative for systematic documentation of infantile AIHA to inform future therapeutic decision-making in this rare and vulnerable population.

Key Words: Idiopathic Autoimmune Hemolytic Anemia, Refractory Anemia, Mycophenolate Mofetil, Rituximab