Autoimmune Hemolytic Anemia and Interstitial Pneumonitis as the Initial Presentation of Systemic Lupus Erythematosus

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OBJECTIVE
To emphasize the importance of considering hemolysis as a cause for anemia and the potentially catastrophic results that can occur if treatment is delayed.

BACKGROUND
• Autoimmune hemolytic anemia (AIHA) involves immunologic destruction of erythrocytes and can be life-threatening.
• AIHA may be idiopathic, however, it should prompt a work-up for secondary causes.
• Evaluation consists of a reticulocyte count, LDH, haptoglobin, indirect bilirubin, direct and indirect Coombs tests, auto-antibody testing, and a peripheral smear for erythrocyte morphology, including microspherocytes, polychromasia, and schistocytes.
• AIHA is not associated with a deficiency state; it is characterized by an elevated LDH and reticulocyte count, indirect hyperbilirubinemia, and a low haptoglobin.

CASE REPORT
• A 48-year-old female from Hong Kong with no significant past medical history presented to the emergency department with a one-month history of progressive dry cough and fatigue.
• On exam, she was noticeably icteric and had bibasilar coarse crackles.
• Labs on presentation: hemoglobin 4.9 g/dL, indirect bilirubin 7.1 mg/dL, reticulocyte count 35%, LDH 468 U/L, haptoglobin 4 mg/dL. Direct Coombs test positive secondary to warm agglutinins.
• Chest radiograph and CT scan revealed interstitial infiltrates.
• An immunologic work-up revealed positive anti-nuclear antibody, rheumatoid factor, anti-SSA, anti-Smith, anti-RNP, and a low complement level.
• High-dose intravenous corticosteroids were initiated, with a slow improvement in hemoglobin level as well as in the pulmonary infiltrates.
• Treatment has now been tapered to low dose oral corticosteroids, and the patient’s hemoglobin remains within normal limits.

DISCUSSION
• The final diagnosis in this patient was systemic lupus erythematosus (SLE), presenting with autoimmune hemolytic anemia and interstitial pneumonitis.
• In warm agglutinin mediated AIHA, IgG antibodies react at body temperature with protein antigens on the erythrocyte surface.
• This results in hemolysis by two mechanisms: (1) phagocytosis within the reticuloendothelial system, i.e. extravascular hemolysis, and (2) complement-mediated erythrocyte destruction.
• Systemic corticosteroids are the mainstay of treatment, while rarely, other immunosuppressants such as rituximab are used.
• Splenectomy may be necessary in refractory cases.

RADIOLOGY
CT chest showing coarse interstitial infiltrates and traction bronchiectasis

Hemolytic Anemia
Extrinsic Causes
• Disseminated Intravascular Coagulation
• Thrombotic Thrombocytopenic Purpura
• Idiopathic and secondary antibody mediated
• Drug induced hemolytic anemia
Intrinsic Causes
• G-6-PD Deficiency
• Hereditary spherocytosis
• Hemoglobinopathies

Autoimmune Disease Associations

<table>
<thead>
<tr>
<th>Auto-antibody</th>
<th>Autoimmune Disease Associations</th>
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<tbody>
<tr>
<td>Anti-SSA</td>
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REFERENCES