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.

Hemolytic Anemia

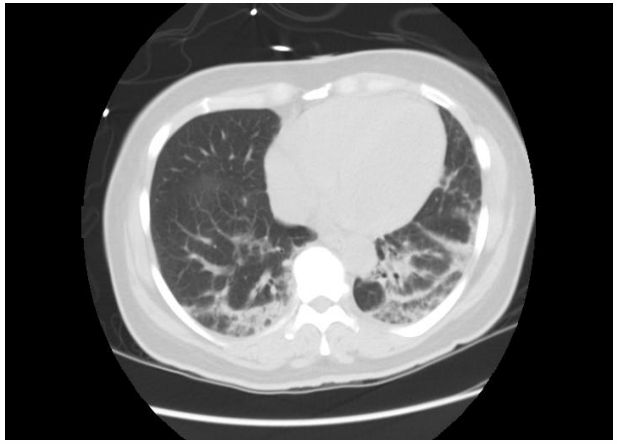
Extrinsic Causes
- Disseminated Intravascular Coagulation
- Thrombotic Thrombocytopenic Purpura
- Idiopathic and secondary antibody mediated
- Drug induced hemolytic anemia

Intrinsic Causes
- G-6-PD Deficiency
- Hereditary spherocytosis
- Hemoglobinopathies

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.

RADIOLOGY



CT chest showing coarse interstitial infiltrates and traction bronchiectasis

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.

Auto-antibody	Autoimmune Disease Associations
Anti-SSA	Systemic Lupus Erythematosus (SLE), Sjogren's syndrome
Anti-SSB	Sjogren's syndrome
Anti-dsDNA	SLE
Anti-centromere	CREST syndrome
Anti-Smith	SLE
Anti-histone	SLE and Drug Induced SLE
Anti-topoisomerase	Systemic Sclerosis
Rheumatoid Factor	SLE, Sjogren's, Rheumatoid Arthritis
Anti-RNP	Mixed Connective Disease, SLE, Sjogren's

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