Warm Agglutinin Autoimmune Hemolytic Anemia (W-AIHA)

Warm agglutinin autoimmune hemolytic anemia is the most common type of autoimmune hemolytic anemia. This type of anemia occurs when patients form IgG autoantibodies to RBCs that react at body temperature. Splenic macrophages recognize the Fc portion of these IgG antibodies and induce extravascular hemolysis. Etiologies for warm agglutinin autoimmune hemolytic anemia include idiopathic, medications, CLL, and SLE. The direct Coombs test is used for diagnosis. Treatments for W-AIHA include corticosteroids, splenectomy, IVIG, and rituximab.



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Pathophysiology

Most Common Autoimmune Hemolytic Anemia

#1 Foam-finger of Auto-in-moon Anemone

Warm agglutinin autoimmune hemolytic anemia occurs when IgG autoantibodies react against the surface of RBCs at body temperature, 37 degrees celsius. It is the most common form of autoimmune hemolytic anemia.

IgG Coats RBCs at Warm Temperatures

Gold-goblin Coats RBC at Warm-thermometer

At warm temperatures equal to or greater than 37 degrees celsius, the average human body temperature IgGs coat RBCs. The Fc portion of the immunoglobulin sitting on the RBC is recognized by macrophages, both from the spleen and liver. Macrophages sequester these RBCs and hemolyze them extravascularly.

Fc Portion of IgG Recognized By Macrophages

Fc-crystal with Gold attracting Mac-man

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Extravascular Hemolysis

Extravascular hemolysis-RBCs

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Medications

Med-bottle

Various medications can precipitate the formation of warm agglutinin autoantibodies. These include alpha-methyldopa, cephalosporins, diclofenac, ibuprofen, and levodopa. Some drugs stimulate the production of IgG autoantibodies, while others attach the RBC surface membrane and form haptens that induce IgG production.

Alpha Methyldopa

Afro Metal-dough-boy

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Chronic Lymphocytic Leukemia (CLL)

CaLL me Crone

Some studies have shown that greater than 10% of patients with chronic lymphocytic leukemia also develop warm agglutinin autoimmune hemolytic anemia. CLL patients that are treated with purine analogs may be at an increased risk.

Systemic Lupus Erythematosus (SLE)

Loopy-butterfly

SLE is associated with warm agglutinin autoimmune hemolytic anemia. Patients may also have a positive Coombs test without any clinical evidence of hemolysis.

Diagnosis

Positive Coombs Test

Positive Comb

The Coombs test detects if a patient has formed autoantibodies to their RBCs. A Direct Coombs test involves mixing a patient's RBCs with test serum containing anti-IgG antibodies. If a patient has W-AIHA, their RBCs will be coated in IgG and anti-IgG in the test serum will agglutinate the RBCs; this reaction is referred to as a positive Coombs test.

Treatment

Corticosteroids

Quarter-on-steroids

Corticosteroids are used to treat many autoimmune conditions, including W-AIHA. Treatment helps to decrease antibody formation and increase hemoglobin. Approximately half of patients require long-term treatment with steroids.

Splenectomy

Chopped-off Spleen

Because much of the extravascular hemolysis in W-AIHA occurs in the spleen, splenectomy is the preferred second-line treatment for patients with W-AIHA refractory to corticosteroids.

Intravenous Immunoglobulin (IVIG)

Ivy-gold-goblin

IVIG, or intravenous immune globin, has limited efficacy for treatment and is employed in refractory cases resistant to corticosteroids.

Rituximab

Red-tux-mob

Rituximab is a monoclonal antibody that works against B-cells, which produce autoantibodies. This drug is also a second-line treatment for patients who fail corticosteroids.

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