Northeast Georgia Medical Center
GRADUATE MEDICAL EDUCATION

Naproxen-Induced Evans Syndrome

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Background/Introduction

Evans syndrome: autoimmune disorder with AIHA and ITP occurring simultaneously.

May be primary when it occurs by itself or secondary when it happens with other autoimmune and lymphoproliferative disorders.

Corticosteroids and immunoglobulins are first line treatments.

Subsequent options include other immunosuppressive medications.

Case Presentation

68-year-old female with medical history of chronic eosinophilia, sinusitis and allergies presents with complaints of shortness of breath and fatigue following Naproxen therapy.

Anemia (6.1 g/dL), thrombocytopenia 62 k/ul, neutropenia, elevated reticulocyte counts. Pulmonary embolism on imaging.

DAT positive for IgG and C3. Eluate notable for pan agglutinin and warm autoantibodies.

Negative tests include lead screening, HIV, leukemia and lymphoma panel, hepatitis, FISH analysis, flow cytometry, antibodies to extract nuclear antigen, normal complement C3 and C4 levels, lupus anticoagulant, rheumatoid factor, ANA, beta-2 glycoproteins, and Epstein-Barr virus.

Positive tests included mycoplasma pneumonia IgG and parvovirus IgG.

Discontinuation of Naproxen, heparin infusion, improved with packed red blood cells and steroids therapy.

LITERATURE REVIEW

According to Sanford-Driscoll et al., nonsteroidal anti-inflammatory drugs have been reported for their ability to cause Evans Syndrome [1].

Per Barbaryan et al., drug-induced immune hemolytic anemia can be further classified depending on whether antibodies to the drug are present or absent [2].

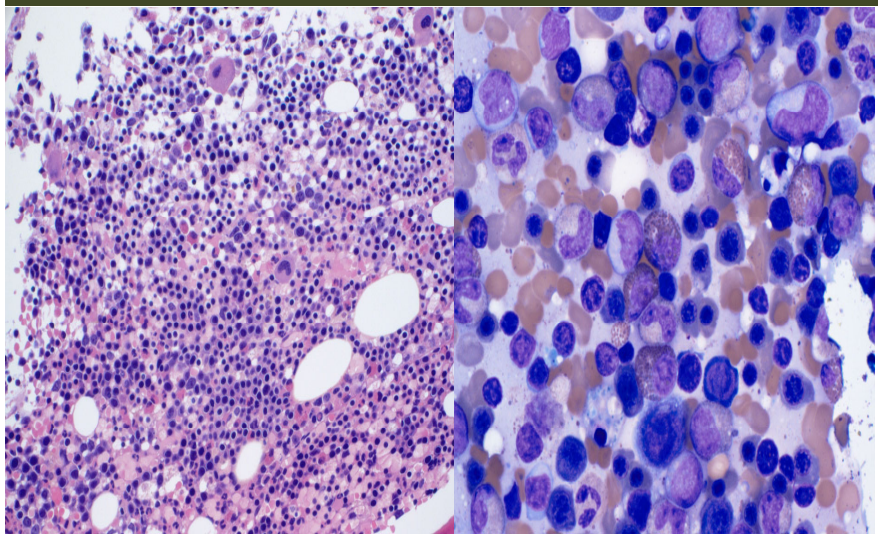
Dhingra et al. reported that bone marrow aspiration is necessary in the workup of Evans syndrome as it allows the clinician to rule out aplastic anemia or infiltrative disorders [3].

According to Norton et al., the first line treatment for primary Evans syndrome involves corticosteroids and/or intravenous immunoglobulins [4].

Rituximab, Mycophenolate, cyclosporine and splenectomy, cyclophosphamide and Alemtuzumab are subsequent treatments [5].

Per Michel et al. the prognosis of Evans syndrome is guarded, variable, and is characterized by relapses [6].

MICROSCOPIC IMAGING



Images 1 and 2: Core biopsy and peripheral smear showing hyper eosinophilia, erythroid and megakaryocytic hyperplasia.

Discussion

Nonsteroidal anti-inflammatory drugs that have been implicated in Evans syndrome include Mefenamic acid, ibuprofen, sulindac, naproxen, tolmetin, feprazone and aspirin.

Positive DAT and positive elution test for pan agglutinin and warm autoantibodies.

In the presence of DVT or pulmonary embolism, consider starting anticoagulants.

Recommendations

Consider Evans syndrome in the differential of patients presenting with autoimmune hemolytic anemia.

NSAIDS are a common cause, review patient's list of medications.

Close follow up with patient after discharge from the hospital.

References

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