SECONDARY EVAN SYNDROME WITH ELEVATED ANTIPHOSPHOLIPID ANTIBODY TITERS

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INTRODUCTION

- Evans syndrome is an autoimmune condition characterized by concurrent or sequential development of autoimmune hemolytic anemia (AIHA) and immune-mediated thrombocytopenia
- Primary Evans syndrome is a diagnosis of exclusion
- Secondary Evans syndrome is due to:
  - Autoimmune conditions such as lupus, antiphospholipid syndrome, Sjogren's syndrome
  - Immunodeficiency disorders such as common variable immunodeficiency (CVID), Autoimmune lymphoproliferative syndrome (ALPS), IgA deficiency
  - Malignancy such as Non-Hodgkin's lymphoma, chronic lymphocytic leukemia (CLL)

CASE

- A 29-year-old male with no medical history who initially presented with atypical headache for the past few weeks
- His blood pressure was 135/74 with tachycardia to the 116

Labs:

- HgB 3.9 g/dL (Ref Range 12.5-17.0)
- WBC 6.1 thou/cmm (4-10.5)

LABORATORIES

- Platelets 57 thou/cmm (140-350)
- Cr 1.38 mg/dL (0.53-1.30)
- TBil 2.3 mg/dL (0.2-1.0)
- Direct Bil 0.4 mg/dL (0.0-0.2)
- LDH 900 U/L (100-250)
- Peripheral smear: nucleated RBCs, spherocytes, polychromasia. No schistocytes
- Direct antiglobulin test (DAT): Positive for Bound IgG, Negative for bound complement

DIAGNOSIS

- Low HgB 3.9, High Indirect Bil 1.9, High LDH 900 -> **Hemolysis**
- Hemolysis + Positive DAT for Bound IgG -> **Warm AutoImmune Hemolytic Anemia (AIHA)**
- Platelets 57k -> **Thrombocytopenia**

Warm AIHA and Thrombocytopenia = Evan Syndrome

TREATMENT

- Patient received blood transfusions at initial presentation for severe anemia
- Subsequently, patient was started on prednisone and rituximab as initial treatment of warm AIHA
- Intravenous immune globulin (IVIG) was given as adjunctive therapy due to persistent anemia status-post blood transfusions
- Workup for headache revealed acute/subacute infarcts in bilateral cerebral hemispheres and right cerebellum on MRI brain. Due to patient's age and stroke findings, thrombophilia workup revealed:
  - elevated anti-beta2-glycoprotein antibodies
  - elevated anti-cardiolipin antibodies
  - negative lupus anticoagulant
- With elevated antiphospholipid titers, patient was started on warfarin with enoxaparin bridge
- Pt was later discharged in stable condition with hemoglobin of 8

DISCUSSION

- Given the elevated antiphospholipid antibody titers, patient is suspected to have secondary Evans syndrome due to antiphospholipid syndrome
- To confirm the diagnosis of antiphospholipid syndrome, patient will need to return in twelve weeks to repeat antibody titers based on the revised Sapporo classification criteria
- Regardless of etiology of Evan syndrome, first-line treatment involves blood transfusion for symptomatic anemia follow by glucocorticoids with rituximab
- Patients with Evan syndrome should be on venous thromboembolism prophylaxis, because hemolysis from AIHA increases thromboembolic complications

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