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 followed by glucocorticoids with rituximab.

Patients with Evan syndrome should be on venous thromboembolism prophylaxis, because hemolysis from AIHA increases thromboembolic complications.

REFERENCES