

ATYPICAL VENOUS THROMBOSIS IN YOUNG PATIENT WITH SYSTEMIC LUPUS ERYTHEMATOSUS (SLE) AND HIGH- RISK APL PROFILE: PROVOKED OR NOT?.

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Background: Antiphospholipid syndrome (APS) is a systemic autoimmune disorder characterized by venous and arterial thrombosis, recurrent abortion, and thrombocytopenia and the presence of antiphospholipid (aPL) antibodies. APS may exist in its primary form when it occurs in isolation or may be associated with other autoimmune diseases, particularly systemic lupus erythematosus (SLE). We describe a case of secondary APS in young patients with SLE.

Case report: In August 2019, a 14-year-old female patient presented with a 1-week history of bruising on her lower extremities. Her laboratory test showed severe thrombocytopenia (platelet count was 2000/ml), anemia, negative Coombs's test, normal LDH and normal peripheral smear. The results of hepatitis B surface antigen, anti-hepatitis C virus, cytomegalovirus, and human immunodeficiency virus serology were negative. Anti-nuclear antibodies (ANA), anti-extractable nuclear antigen (ENA) and anti-double stranded deoxyribonuclease (dsDNA) were positive (ANA 1: 320, ENA ++, anti dsDNA+++). C3 and C4 complement factors were low. Activated partial thromboplastin time (aPTT) Ratio was prolonged (1.93 n.v 0.7-1.3) with a positive test for lupus anticoagulant antibodies and negative anticardiolipin antibodies. Total body CT scan with contrast media was negative for thrombosis and for active malignancy. According to the EULAR/ACR-2019 she was diagnosed with SLE without APS but with high- risk aPL profile. She started hydroxychloroquine with a low dose of corticosteroids and belimumab, as biologi-

cal agent. In March 2025, the patient was admitted to our hospital due to edema and pain in the left arm. Venous Doppler ultrasound revealed thrombosis of the left axillary vein, left subclavian vein and left jugular vein. Total body CT scan with contrast media also showed pulmonary embolism of the left pulmonary artery. It was negative for active malignancy. She had never smoked, and did not take oral contraceptive (COC). She was not pregnant but was overweight and hypertensive. She reported having suffered accidental trauma to her left arm. Fondaparinux at a dosage of 7.5 mg/die started. Anticoagulant protein S, protein C, and Antithrombin were normal. Factor V Leiden and prothrombin G20210A mutation were negative. Her laboratory test confirmed a prolonged of the activated partial thromboplastin time (aPTT) Ratio of 1.95 (0.7-1.3) with a positive test for lupus anticoagulant antibodies and negative anticardiolipin antibodies suggested for secondary post traumatic APS in patient with SLE and high-risk aPL profile. She was discharged with prolonged oral anticoagulant therapy, maintaining an international normalized ratio (INR) between 2 and 3.

Conclusion: aPL antibodies occur in 20 % to 30 % of patients with SLE, but their presence does not confirm the diagnosis of APS. They are associated with an increased risk of thrombosis or pregnancy morbidity, depending on aPL characteristics and coexistence of other risk factors. Therefore, it is important to report on the clinical features of patients with SLE associated with risk aPL profile to improve management and therapeutic strategies.

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