UCLA Health

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Abstract Form

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Project Title:	Visceral infarction as the initial manifestation of systemic lupus erythematosus overlap syndrome complicated by antiphospholipid syndrome.

Research Category (please check one):

☐	**Original Research**	☒	**Clinical Vignette**	☐	**Quality Improvement**	☐	**Medical Education Innovation**

Abstract

Introduction: Systemic lupus erythematosus (SLE) is an autoimmune disorder that causes multisystem inflammation that can be complicated by antiphospholipid syndrome (APS). Clinical manifestation of APS often affects young women ages 30-40, present as arterial and/or venous thrombosis, and pregnancy morbidity in the setting of detectable circulating antiphospholipid antibodies. Herein, we describe a rare case of multiple visceral infarctions as a presenting manifestation of SLE with overlap syndrome with Rheumatoid arthritis (RA) and Sjögren's syndrome (SS) complicated by late onset secondary APS.

Case Report: A 68-year-old woman with seropositive non-erosive RA and SS presented for worsening left upper quadrant and left flank pain for 8 days. In the Emergency room, vital signs were normal. Examination revealed left upper quadrant and left costovertebral angle tenderness along with left knee pain. Initial laboratory studies were significant for WBC 3.8 K/cumm, Hgb 7.9 g/dL, Hct 23.9%, Plt 109 K/cumm, PT 15.5 sec, and INR 1.26. CT imaging of the abdomen and pelvis revealed marked splenomegaly and multiple infarcts of the spleen, left upper and midpole regions of the kidneys. She was subsequently started on a heparin drip.

EKG and telemetry were negative for atrial fibrillation. Transthoracic echocardiography was negative for valvular pathologies and structural heart disease. Doppler ultrasound of the lower extremities was negative for deep vein thrombosis. The hypercoagulable work up including anti-cardiolipin, beta-2 glycoprotein, antithrombin III, and factor V Leiden mutation were normal. Whereas, lupus anticoagulant was indeterminate, protein C (59%) and protein S (67%) were mildly low. Given APS was highly likely given her significant rheumatologic history and acute arterial thromboses, rheumatology was consulted. Based on the European Alliance of Associations for Rheumatology (EULAR) and American College of Rheumatology (ACR) classification criteria, she met SLE criteria for diagnosis with leukopenia (+3), joint involvement (+6), anti-Smith Ab (+6) (15 points total). In consultation with rheumatology, prednisone and hydroxychloroquine was initiated and home immunosuppressive medications etanercept and leflunomide were held. On discharge, she was anticoagulated with warfarin given her multiple thrombotic events. Repeat hypercoagulable work up 3 months later was notable for a positive beta-2 glycoprotein.

Discussion: APS is a multisystemic autoimmune disorder which can be secondary to autoimmune processes like SLE. Among SLE patients, around 40% of patients who are antiphospholipid antibody-positive develop thrombotic events in comparison with 10-20% of those screening negative for antibodies. As part of the clinical and laboratory criteria to confirm an APS diagnosis, antibodies need to be positive on 2 occasions at least 12 weeks apart. Optimally, testing should not be performed during an acute thrombotic event as interpretations may be difficult. As in this case, protein C and S levels were decreased, lupus anticoagulant may be falsely negative, or anticardiolipin may be transiently elevated.

When evaluating SLE related abdominal pain, the differential should include medication-related, lupus enteritis, pancreatitis, vasculitis, pseudo-obstruction, acalculous cholecystitis and APS related infarctions. Only a few cases of splenic or renal infarction have been documented with the presence of antiphospholipid antibodies, but rarely seen simultaneously in SLE. Along the same lines, the incidence of thrombotic events in secondary APS with overlap syndromes is lacking in the literature. Given this patient's high burden of inflammatory disease from overlap syndrome, which is characterized by the clinical and biochemical features of at least two connective tissue diseases, in this case with SLE, RA and SS, we can infer she had a high burden of inflammatory disease from her autoimmune conditions which served as a provoking source for acute thrombosis.

Conclusion: This case emphasizes that visceral infarctions should be considered in patients with a history of autoimmune conditions who complain of unusual localized pain symptoms without typical SLE manifestations; thus providers should have a low threshold for pathogenic antiphospholipid antibody testing.