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Uncommon initial presentations of SLE – A Case series

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Introduction

Systemic lupus erythematosus (SLE) is a systemic autoimmune disease that is characterized by a highly variable multisystem involvement with relapsing-remitting nature. The clinical heterogeneity of SLE can give rise to diagnostic challenge and can delay treatment initiation. We report a series of three female patients with unusual initial presentations who were subsequently diagnosed with SLE and treated successfully with immunosuppressants.

Case Presentations

A 29 year old patient presented with progressive dyspnea (mMRC grade 3) over 4 months. Her 2D Echocardiogram revealed moderate pulmonary hypertension with right ventricular dilatation. She had a history of inflammatory polyarthritis and constitutional symptoms. Her ANA was 1: 80 (Homogenous pattern). HRCT-Chest, CTPA, bubble contrast study and serology tests including APLS screening were all negative. A diagnosis of pulmonary arterial hypertension (PAH) was made. She also had an incidental detection of diffuse splenic calcification which is a rare association of SLE. She was treated with immunosuppressants and vasodilators for which she had a symptomatic improvement.

A 31 year old patient presented with progressive abdominal pain, distention and diarrhoea for 2 weeks duration. She underwent an exploratory laparotomy which revealed gross ascites and dilated small intestinal loops. Subsequent Abdominal CT showed small intestinal wall ischemia involving multiple arterial territories. She also reported on alopecia and gave a history of ITP during her pregnancy in 2016. With positive ANA (1:1280 titer), low complement levels, negative APLS screening and exclusion of other causes, SLE with mesenteric vasculitis was confirmed.

A 40 year old patient presented with a 2 week history of fever, nonproductive cough, inflammatory type polyarthritis and bilateral axillary lymphadenopathy. She later developed a pancytopenia and alopecia. Her ANA was 1:10000; Diagnosis of SLE was made. Axillary lymph node biopsy showed necrotizing lymphadenopathy and immunohistochemistry was strongly positive for CD68 favoring Kikuchi disease.

Conclusion

Initial clinical presentations of SLE can be highly variable; can mimic infections and malignancies and may lead to delays in diagnosis. Therefore the clinicians should be vigilant on rare

presentations of SLE such as PAH, mesenteric vasculitis and necrotizing lymphadenopathy to initiate timely treatments. Timely management in mesenteric vasculitis is crucial to prevent the possible catastrophic complications such as necrotic bowel, perforation, and sepsis.