

# Systemic Lupus Erythematosus Presenting as Splenic Infarcts – A Rare Case Report

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**Abstract:** *Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease. In patients with SLE, the prevalence of antiphospholipid antibodies is considerably higher, and is largely responsible for thrombosis. Splenic infarction is a rare complication of arterial thrombosis in patients with SLE. We hereby present a rare case report of SLE with AIHA with splenic infarcts in a negative antiphospholipid antibody patient who responded well to steroid therapy.*

**Keywords:** Systemic Lupus Erythematosus ( SLE ), Autoimmune hemolytic Anaemia ( AIHA ), Splenic infarct (SE), anti-cardiolipin antibody ( ACA ), anti-phospholipid antibody ( APA ).

## 1. Introduction

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease. In patients with SLE, the prevalence of antiphospholipid antibodies is considerably higher, and is largely responsible for thrombosis. Splenic infarction is a rare complication of arterial thrombosis in patients with SLE. It is important to consider splenic infarction in a patient with SLE complaining of left upper quadrant (LUQ) pain because of the possibility of severe infarction-related complications, such as subcapsular hemorrhage and splenic rupture. Splenic infarction can occur in various diseases; however, the incidence is very low (49 cases recorded worldwide with associated 10% mortality) [1]. Thromboembolic events associated with atrial fibrillation, cardiac surgery, or infective endocarditis are well-known causes of splenic infarction. Additionally, several hematologic diseases including sickle cell anemia, lymphoma, and leukemia can cause splenic infarction. Chronic myeloproliferative disease can also lead to splenic infarction due to massive splenomegaly and a hypercoagulable state [2]. Massive splenomegaly may lead to splenic infarction and subsequent splenic rupture.

We report a case of multiple splenic infarction in a patient with SLE. The only symptom was LUQ pain of 3-day duration. Lupus anticoagulant activity was positive and abdominal-pelvic computed tomography (CT) was consistent with splenic infarction. She did not show any other evidence of thrombotic events. The patient was diagnosed with autoimmune hemolytic anaemia that presented as a splenic infarction in a SLE patient.

## 2. Case Report

19 years old married female came to the casualty with the chief complaints of breathlessness at rest and easy fatigability since 15 days, abdominal pain more on the left hypochondriac area, non-radiating with no relieving and aggregation factors and fever since 8 days. No history of

upper respiratory tract infection or urinary tract infection was noted. No history of headache, rash over the body, vomiting, nausea, loose motions. No history of joint pains and blood transfusions in past. On examination, she was febrile, conscious, oriented, tachycardia (pulse of 130 beats per minute) blood pressure of 120/90 mm Hg, and tachycardia (respiratory rate of 34 cycles per minute) was present. Pallor present moderate to severe, found to be icteric. Jugular venous pressure was raised with no presence of pedal oedema. No evidence of cyanosis, clubbing and lymphadenopathy. No evidence of any rash over the body. Per abdomen examination revealed presence of tender spleen and hepatomegaly. All other systems were normal.

Patient had received two units of packed cell volume in view of anaemia 2 days back. On investigation, hemoglobin was 4.5 mg/dl, TLC- 6400 /cumm, platelets- 168000 /cumm, MCV- 116 IU/lit. Peripheral smear was suggestive of moderate anisopoikilocytosis, macrocytic, hypochromic Total bilirubin was 3.01 mg/dl, (D.bil- 2.0mg/dl, I.bil- 1.01 mg/dl)SGOT- 82, SGPT- 37, ALP - 84, suggestive of hemolytic anaemia versus megaloblastic. Sickling test was found to be negative. Retic count was 2.8 %, Direct coombs test- positive. Bone marrow was suggestive of a normocellular marrow. Urine had evidence of albuminuria 1 plus ultrasonography of the abdomen revealed presence of multiple splenic infarcts with splenomegaly. Chest Xray was within normal limits. Patient had positive titers for serum ANA and DsDNA, whereas negative tests for hemoglobinuria which gave a diagnosis of Systemic Lupus Erythematosus. Hemoglobin electrophoresis was normal study. Serum anti-phospholipid antibodies were negative. Patient was given no blood transfusions and started on intravenous Methylprednisolone therapy for 5 days followed by oral steroids.



Figure 1

### 3. Discussion

Systemic lupus erythematosus (SLE) is the most common multisystem connective tissue disease. It is characterized by a wide variety of clinical features and presence of numerous auto-antibodies, circulating immune complexes and widespread immunologically determined tissue damage [3].

SLE can present in many ways out of which musculoskeletal accounts for 95 %, 80 % being cutaneous manifestation, haematological 85 %, neurological 60 %, cardiopulmonary 60 %, renal 50 %, gastroenterological 40 %, arterio-venous thrombosis 15 % and ocular manifestation 15 %[4].

Hematological abnormalities are very common in systemic lupus erythematosus. Impaired erythropoietin response and presence of antibodies against erythropoietin may contribute to the pathogenesis of this type of anemia.[5] Patients with autoimmune hemolytic anemia usually belong to a distinct category, which is associated with anticardiolipin antibodies, thrombosis, thrombocytopenia and renal involvement, often in the context of secondary antiphospholipid syndrome. Finally, as recently suggested, autoantibodies, T lymphocytes and deregulation of the cytokines network can affect bone marrow erythropoiesis leading to anemia.[6] Anemia is found in about 50% of SLE patients, many mechanisms contribute to the development of anemia, including inflammation, renal insufficiency, blood loss, dietary insufficiency, medications, haemolysis, infection, hypersplenism, myelofibrosis, myelodysplasia, and aplastic anemia that is suspected to have an autoimmune pathogenesis [7,8]. Hematological abnormalities are very common in systemic lupus erythematosus. Autoimmune hemolytic anemia (AIHA), caused by autoantibodies binding to the surface of RBCs, is an uncommon disease with an incidence of approximately 1-3 cases/100,000 per year in the general population [10]. In AIHA, massive hemolysis causes activation of the immune response, destruction of RBCs, and splenomegaly [11]. Splenic infarction can occur in AIHA, although rarely, and only 2 cases have been reported worldwide. There have been many reports of splenic infarction in patients with protein C deficiency [12,13], and a few of them were accompanied by hematologic diseases, such as hereditary spherocytosis or acute myeloid leukemia

[14].

A frequent cause of anemia in SLE is suppressed erythropoiesis from chronic inflammation (anemia of chronic disease or anemia of chronic inflammation), being the most common form (60 to 80 %) [15]. This type of anemia is normocytic and normochromic with a relatively low reticulocyte count. Although serum iron levels may be reduced, bone marrow iron stores are adequate and the serum ferritin concentration is elevated. In the absence of either symptoms attributable to anemia (eg: dyspnea on exertion, easy fatigability) or renal insufficiency, anemia of chronic inflammation does not require specific treatment.

Among SLE patients, the prevalence of antiphospholipid antibodies is high, ranging from 12% to 30% for anticardiolipin (ACL), and 15% to 34% for lupus anticoagulant antibodies. Several studies of patients with SLE demonstrated a significant correlation between ACL or lupus anticoagulant and Coombs' positive hemolytic anemia

(20-25). There is increasing evidence that ACL autoantibodies are not just a secondary phenomenon caused by haemolysis. They could also contribute to the pathogenesis of AHA by acting as anti-erythrocyte autoantibodies (16,17).

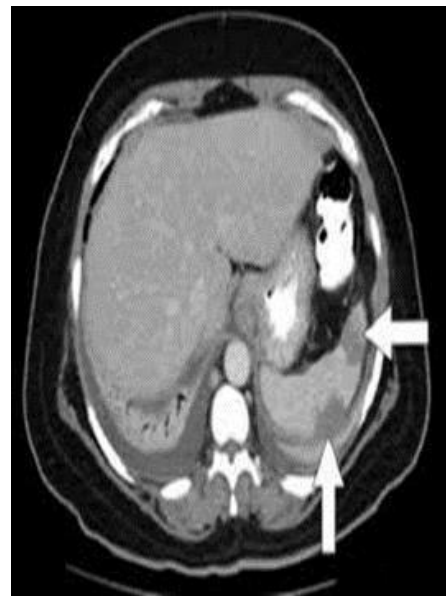


Figure 2: Showing multiple splenic infarcts.

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