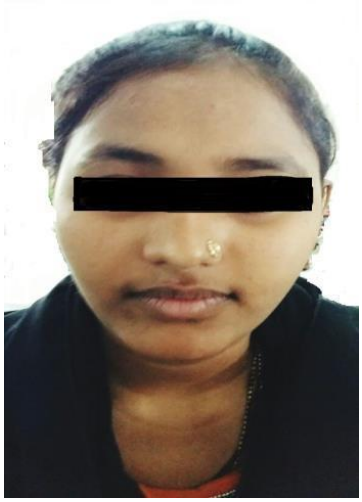

Figure 1

3. Discussion

Systemic lupus erythematosus (SLE) is the most common multisystem connective tissue disease. It ischaracterised 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 occular 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 erythopoiesis 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).

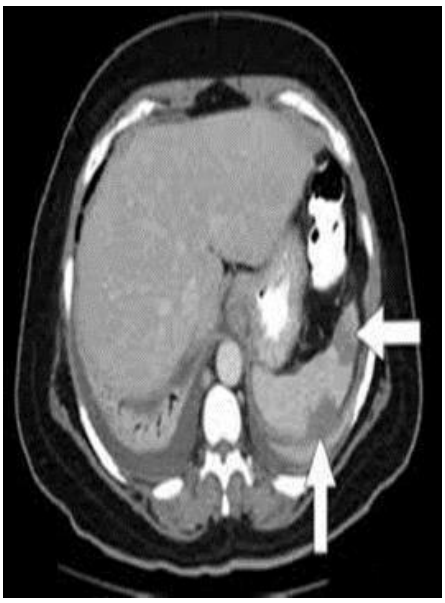

Figure 2: Showing multiple splenic infarcts.

References

[1] Nores M, Phillips EH, Morgenstern L, Hiatt JR (February 1998). "The clinical spectrum of splenic infarction". Am Surg 64 (2): 182–8. PMID 9486895.
[2] Antopolsky M, Hiller N, Salameh S, Goldshtein B, Stalnikowicz R. Splenic infarction: 10 years of experience. Am J Emerg Med. 2009;27:262–265. [PubMed] Naithani R, Agrawal N, Mahapatra M, Pati H, Kumar R, Choudhary VP. Autoimmune hemolytic anemia in India: clinico-hematological spectrum of 79 cases. Hematology. 2006;11:73–76. [PubMed]
[3] Eichner ER. Splenic function: normal, too much and too little. Am J Med. 1979;66:311–320. [PubMed]

[4] Horeau J, Robin C, Guenel J, Nicolas G. An unusual complication of acquired hemolytic anemia, splenic infarction. Concours Med. 1963;85:663–666. [PubMed]
[5] Tzanck A, Andre R, Dreyfus B. Acquired hemolytic anemia with infarct of the spleen; splenectomy; recovery. Bull Mem Soc Med Hop Paris. 1951;67:286–290. [PubMed]
[6] Khor B, Van Cott EM. Laboratory tests for protein C deficiency. Am J Hematol. 2010;85:440–442. [PubMed]
[7] Farah RA, Jalkh KS, Farhat HZ, Sayad PE, Kadri AM. Acquired protein C deficiency in a child with acute myelogenous leukemia, splenic, renal, and intestinal infarction. Blood Coagul Fibrinolysis. 2011;22:140–143. [PubMed]
[8] Olson JF, Steuber CP, Hawkins E, Mahoney DH., Jr Functional deficiency of protein C associated with mesenteric venous thrombosis and splenic infarction. Am J Pediatr Hematol Oncol. 1991;13:168–171. [PubMed]
[9] Erslev AJ. Pure red cell aplasia. In:Beutler E, Lichtman MA, Coller BS, Kipps TJ, Seligsohn U, eds. Williams Hematology. 6 th ed. New York: McGraw-Hill, 2001; 391-8.
[10] Tan EM, Cohen AS, Fries JF, et al. The 1982 revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheuma 1982; 25: 1271-7.
[11] Sun CF, Tsao KC. Positive predictive value of fluorescent antinuclear antibody test, anti-n-DNA test and LE cell preparation test in SLE. Chang Gung Med J 1986; 9: 17-25.
[12] Notman DD, Kurata N, Tan EM, California LJ. Profiles of antinuclear antibodies in systemic rheumatic diseases. Ann Intern Med 1975; 83: 464-9.
[13] Reichlin M. Undifferentiated connective tissue disease, overlap syndromes and mixed connective tissue disease. In: Koopman WJ, ed. Arthritis and Allied Conditions. 13 th ed. Maryland : William & Wilkins, 1997; 1309-18.
[14] Meyer RJ, Hoffman R, Zanjani ED. Autoimmune hemolytic anemia and periodic pure red cell aplasia in systemic lupus erythematosus. Am J Med 1978; 65: 342-5.
[15] Tepperman AD, Curtis JE, McCulloch EA. Erythropoietic colonies in cultures of human marrow. Blood 1974; 44: 659-69.
[16] Lang B, Straub RH, Weber S, Rother E, Fleck M, Peter HH. Elevated anticadiolipin antibodies in autoimmune haemolytic anaemia irrespective of underlying systemic lupus erythematosus. Lupus 1997;6:652-5.
[17] Cheng HM. IgG antiphospholipid autoantibody in normal human sera is reactive against bromelain treated human erythrocytes. J Rheumatol 1993;20:400-1