Haematological Presentation of Systemic Lupus Erythematosus


Abstract
Haematological manifestations are quite common in systemic lupus erythematosus (SLE) but bone marrow aplasia and secondary myelofibrosis are rare manifestations. We report a case of 45 years old male patient who presented with fever, malaena and anaemia without any clinical features of SLE. He had patches of vitiligo for 25 years for which he used to take psoralen with sunlight exposure. This probably precipitated SLE in this patient. The patient presented with pancytopenia which was due to a combination of Comb’s positive haemolytic anaemia and myelofibrosis.

Introduction
Systemic lupus erythematosus (SLE) is a chronic autoimmune inflammatory disease of unknown aetiology characterized by multiorgan involvement. Amongst the haematological manifestations bone marrow hypoplasia is rare and myelofibrosis extremely rare. Ultraviolet light (or sunlight) is a well documented environmental trigger. The present case report highlights the development of lupus in a patient undergoing prolonged exposure to sunlight following psoralen administration and manifesting as aplastic anaemia with myelofibrosis.

Case Report
A 45 year old male presented in March’08 with fever and malaena of 2 weeks duration. The fever was low grade, without chills. There were no other constitutional symptoms. He noted malaena about 100 ml each, 3-4 times a day after the fever. He also had a few episodes of epistaxis over the last two weeks. He gave no history of significant past illness or bleeding diathesis. There was no history of oral ulcers or skin rash. He has had patches of vitiligo for the last 25 years for which he used to take herbal medicines along with exposure to sunlight. Family history was insignificant except that his father was suffering from bone cancer. However details were not available.

Examination revealed gross pallor, pulse rate of 100/min and supine BP of 90/50 mm of Hg. Gastrointestinal examination revealed no organomegaly and there was no tenderness. Intestinal peristaltic sounds were hurried. Examination of other systems was normal. There were patches of vitiligo scattered discreetly over forearm, back and face including lips. He didn’t have psoriasis.

Laboratory investigations showed Hb 5.3g/dl, TLC 2500/mm$^3$ (P50 L40 E10), platelets 25000/mm$^3$ with increased reticulocyte count (corrected reticulocyte production index = 3). Liver function tests, blood sugar, urea, creatinine were normal. Urine routine examination showed no albumin or casts. LDH was 337U/L (mildly elevated). Chest X ray and USG abdomen were normal. Upper GI endoscopy revealed gastric erosions with remnants of altered blood. Patient received 4 units of blood along with antibiotics and proton pump inhibitors.

Bone marrow aspiration showed normal erythropoiesis, myelopoiesis and megakaryopoiesis with adequate iron stores.
Bone marrow trephine biopsy showed hypercellular marrow with focal lymphocytic infiltration (Fig. 1) and moderate degree of fibrosis (Fig. 2).

Direct Coomb’s test was positive on two occasions. ANA by indirect immunofluorescence was positive (>1 in 640), with homogeneous pattern. Anti dsDNA antibody was positive in 1 in 80 dilution. Though the vital parameters of the patient improved he continued to get low grade fever.

A repeat chest X ray after 2 weeks showed blunting of both costophrenic angles (Rt>Lt). Aspiration of pleural fluid showed exudative features, ADA was less than 20, PAP smear was negative for malignant cells.

The patient was started on 60 mg prednisolone daily with remarkable improvement. Haemoglobin level after 1 week was 10.4 g/dl, the leucocyte count was 5000 cells/mm³ and platelets 90,000/mm³. This dose was continued for 4 weeks with gradual step down to 5 mg/day and the patient is now maintaining good health.

Discussion

Though haematological abnormalities are frequently encountered, haematological presentation per se without other manifestations are rare in SLE. Pancytopenia is extremely rare, and that too as the sole presentation makes this case very unusual. In this case the haematological manifestations predated pulmonary manifestations by several months. Pathogenesis of pancytopenia in SLE is due to multiple mechanisms such as T cell dysregulation, inhibition of haematopoietic function. Peripheral autoantibody production may contribute to the final clinical picture.

Failure of multiple cell lines as a presenting feature of SLE has been rarely described.

Another characteristic feature of this case is the role of UV rays in the aetiopathogenesis of SLE. This patient had vitiligo for a long time and was taking psoralen along with exposure to UV rays of sunlight. UV rays induce increased apoptosis in the cells and in the process the nucleotides are frequently expressed on the surface of dying cells as blebs, thereby increasing the chance of being recognized as autoantigens. Hence UV rays frequently unmask SLE.

References