SLE-An Atypical Presentation

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Systemic Lupus Erythematosus is a systemic autoimmune disorder characterised by widespread inflammation affecting many systems of the body. Arthralgias, fever and rash are the commonest presenting features amongst Indian Lupus patients. Though varying hematologic manifestations evolve over the course of illness, sole presentation in the form of a bleeding disorder is rare. We are reporting a case of SLE in a postmenopausal lady who presented with epistaxis and cutaneous bleeding manifestations.

A 55-year-old housewife presented with spontaneous appearance of purpurae and ecchymotic patches over the body of 6 months duration. She gave history of occasional episodes of epistaxis of 4 years duration for which she was evaluated at a local hospital. She also gives history of exertional breathlessness and exertional palpitation over past 2 years. There was no history of any other systemic, musculoskeletal or cutaneous symptoms. No history of diabetes mellitus, hypertension or any other chronic illness. She attained menopause 8 years ago and there was no history of any bleeding manifestations before.

On admission, she was pale with purpuric and ecchymotic patches over the abdomen and thigh. Her vitals were stable and there was no evidence of active bleeding. Her systemic examination was within normal limits. There was no lymphnode enlargement, bone tenderness or hepatosplenomegaly. Laboratory investigations revealed Hemoglobin of 8 g%. Total count of 4,800 cells/cmm, Differential count of Polymorphs-40%, Lymphocytes-52% and Eosinophils-8%. ESR was 145 mm/1st hour. Blood group- O +ve. Platelet count - 80,000/cmm Bleeding time - 8 min 50 sec Clotting time - 9 min 30 sec Peripheral smear showed normocytic normochromic anemia with moderate degree of thrombocytopenia. Urine Routine Examination revealed albuminuria +++ with tubular casts. RBS-150 mg% Blood urea -27 mg% S.Creatinine- 0.88mg% S.bilirubin-0.8 mg% Total protein 6.6mg% SGOT-18IU/L, SGPT -22IU/L, Serum alkaline phosphatase-156 IU/L USG of abdomen was normal.

After about 4 days of admission, the purpurae and ecchymoses increased in number with gum bleeding. Repeat blood analysis revealed Hemoglobin of 6.4g% and platelet count of 40,000/cmm. Peripheral smear showed marked thrombocytopenia. In the background of anemia, high ESR albuminuria and thrombocytopenia, SLE was suspected and the patient was further investigated. Anti nuclear antibody was positive. Anti double stranded DNA was positive. APLA was excluded by negative anticardiolipin antibodies and lupus anticoagulant. Diagnosis of SLE was made as per ACR criteria. [Thrombocytopenia, albuminuria with casts, positive ANA and Anti Ds DNA.]

She was started on steroids (1mg/Kg/day) and as platelet count was low with increasing number of purpurae and ecchymoses, platelet concentrate was given. Repeat analysis after one week showed a platelet count of 20,000/cmm, and she was started on Azathioprine 50 mg OD. Condition was better at discharge after two weeks, with a platelet count of 56,000/cmm and there were no fresh ecchymoses. She was on regular follow up with platelet count remaining in a range of 50,000 - 1 lakh/cmm and was hospitalised after 2 months with increasing fatigue, gum bleeding, epistaxis and fresh ecchymotic patches all over the body. Investigations revealed Hb of 8.8g%, TC-4,400/cmm,DCP56 L40 E4 ESR-110mm/1st hour. Platelet count-40,000/cmm. Pulse therapy with Methyl
Prednisolone 1 gm, diluted in 200 ml of Normal saline for 3 days given along with blood transfusion. Azathioprine dose increased to 50 mg BD and steroids in high dose continued. Her condition has improved and there was no further cutaneous bleeding, platelet count raised to 80,000/cmm after two weeks.

SLE is one of the most common autoimmune disorder in women during their child bearing years. It is an autoimmune disease in which organs, tissues and cells undergo damage, mediated by tissue-binding auto-antibodies and immune complexes (1) Most common clinical manifestations are systemic (in 95%), musculoskeletal (in 95%), cutaneous (80%) and hematological (85%).

Diagnosis of SLE is by ACR criteria. Haematologic manifestations can be anemia, leucopenia and thrombocytopenia. Anemia can be due to the chronic inflammatory disease, renal insufficiency, blood loss, drugs or autoimmune hemolytic anemia. Leucopenia is also common and almost always consists of lymphopenia.(1)

Thrombocytopenia may be a recurring problem in SLE and can be present as a part of severely active disease and correlates with the severity of SLE (2,3). It is present in less than 15% of patients throughout the course of illness and as initial and sole presentation is still rare (less than 5%) (3)

In most instances, the thrombocytopenia in SLE appears to result from immunologic platelet injury and is identical to idiopathic thrombocytopenic purpura in most respects. (4)

Thrombocytopenia in SLE can be due to either specific platelet autoantibodies or immune complex deposition on platelets. Only a few studies have been reported in which newest platelet antigens capture autoantibody tests have been used, and in these patients, platelet autoantibodies to platelet membrane glycoproteins have been detected (4,5). These antibodies, which may be present in SLE or otherwise healthy patients also bind to platelet membrane gplIb / IIIa or Ib / IX. (5)

Treatment of haematological manifestations: Patients with SLE and Immune thrombocytopenia should be treated the same as patients with Idiopathic thrombocytopenic purpura even though there are conflicting reports on the success rate of splenectomy in this population (6,7).

When low counts become clinically significant, steroids are started and if unresponsive or if steroids are to be tapered, cytotoxic drugs like azathioprine can be started. If no response, intravenous gamma globulin (IVIG) can be given to tide over the crisis or splenectomy has to be considered.

References