RECURRENT PURPURA AS AN UNCOMMON PRESENTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS

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A middle-aged female with frequent attacks of non-inflammatory purpura was confirmed to have systemic lupus erythematosus. The coagulation profile was normal.

Key words : Purpura, Systemic lupus erythematosus.

Systemic lupus erythematosus (SLE) is a multisystem disease with protean forms of clinical presentations. The following report describes an unusual occurrence in a middle-aged woman with predominantly frequent episodes of purpura.

Case Report

A 50-year-old lady was referred for asymptomatic, erythematous eruptions over the lower limbs of 10 days duration. History disclosed that her lesions commenced 8 years ago and were of a recurrent nature. The eruptions generally subsided within a fortnight. A few months following the first appearance of the eruptions, she had excessive bleeding from a minor trauma which necessitated blood transfusion. Subsequent investigations revealed the coagulation profile, bone marrow biopsy and liver scan to be normal. The eruptions continued but the patient stopped seeking any advice as these were asymptomatic. Careful interrogation revealed that 12 years ago she had moderate pain and swelling of the small joints of the hands along with pale blue discoloration of the fingers which was more marked in winter. Since then, the swellings had subsided but arthralgia persisted. Two years ago she was treated for an attack of psychosis with chlorpromazine 100 mg/day and she became normal after 4 months of therapy. Six months back she had three episodes of bleeding following tooth extraction, the last two requiring blood transfusions. A few weeks later, this was followed by the skin eruptions, when she sought our opinion. The patient is a known case of bronchial asthma controlled on regular treatment with salbutamol. She had no significant lymphadenopathy. Cutaneous lesions were discrete, multiple, non-tender, erythematous macules and papules over the legs and lower part of the thighs. Diascopy was negative. Hyperpigmented spots in various stages of disappearance were seen over the sites of previous lesions. Nail folds of fingers showed telangiectasia. Mucous membranes, palms and soles appeared normal. Systemic examination revealed scattered rhonchi over the chest and a normal heart. The spleen was palpable 3 cm below the costal margin. Liver was not enlarged. Fundus examination was normal.

Her haemoglobin was 9.8 gm, TLC 6,800, DLC, P40, L 56, M1, E3, ESR 108, platelets 1,50,000, bleeding time 3' 20". clotting time 8' 20", prothrombin time 17" (control 15"), thromboplastin time 20" (control 20"), partial thromboplastin time with kaolin 50" (control 42") and clot retraction 70%. LE cell test was positive. Blood VDRL was negative. Blood sugar, urea and liver function tests, routine urinalysis, X-rays of the chest, knees, small joints of hands and barium swallow examination were normal. Histopathology from a fresh papular lesion over the thigh showed areas of extravasation of erythrocytes in the dermis

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without any vasculitic changes. No other significant findings were seen. Immunoglobulin levels were IgG 3407 mg% (normal value 712-1550), IgA 670 mg% (normal 120-220) and IgM 352.68 mg% (normal 65-170). Serum complement levels were C3 70 mg% (normal 80-120) and C4 10 mg% (normal 22-40). Direct immunofluorescence from both involved and un-involved sun-exposed skin was negative. Indirect immunofluorescence showed anti-nuclear antibodies (ANA) in a titre of 1 : 640 with a speckled pattern. Serum electrophoresis revealed no abnormal proteins.

Comments

The patient had an insidious onset of the disease which began as arthritis without deformity. Inclusion of psychosis, positive LE cell and fluorescent ANA tests fulfils 4 of the 11 criteria required for the diagnosis of SLE.[1,2] This was further substantiated by raised immunoglobulins, decreased serum complement levels, anemia, raised ESR and presence of Raynaud's phenomenon.

Purpura is not included in the American Rheumatism Association's criteria for the diagnosis of SLE though it has been documented that SLE should be suspected in cases of purpura and easy bruising.[3] A case presenting predominantly as purpura is rather uncommon. Purpura and ecchymoses have been recorded in 19.8% of patients with SLE.[4] Three major causes of purpura in SLE include corticosteroid therapy, thrombocytopenia and cutaneous vasculitis.[5] A circulatory anticoagulant is also implicated which probably retards the conversion of prothrombin to thrombin.[6] None of these factors seems to be operative in the present case.

Our patient is unique, showing non-inflammatory purpura without thrombocytopenia and a normal coagulation profile. Circulating immunoglobulins were markedly raised but the lupus band test was negative from both involved and un-involved sun-exposed skin indicating that the skin was not primarily affected. These features can be compared to the case of pulmonary hemorrhage in SLE where no immunoglobulins or complement deposits could be demonstrated in the vessel walls or lung alveoli.[7] Purpura in our patient may be related to increased immunoglobulin levels as in hyperglobulinemic purpura[8] or the lupus anticoagulant may be acting in some way, other than altering the coagulation pathway.

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