

Systemic Lupus Erythematosus with Chorea

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Nervous system involvement in systemic lupus erythematosus (SLE) varies from 20 %-75 % in reported series.¹ Manifestations of neurological involvement due to SLE vary from seizures, psychoses, and headaches to peripheral neuritis.² There has been only few reported cases of chorea associated with SLE.³⁻⁵ The present study describes a case of chorea that occurred after the diagnosis of SLE was made.

Case Report

A 13 year old girl was admitted to Hacettepe Children's Hospital in June 1976 with the complaints of erythematosus, macular facial rash, arthralgia, arthritis, and edema of both eyelids for one and a half months duration.

The initial clinical examination revealed erythematosus eruption, cervical, axillar, and inguinal microlephadenopathy, hepatomegaly and splenomegaly, 2 and 4 cm below the costal margins respectively. Blood pressure was 120/80 mmHg.

Laboratory Findings: Hb 5 g/dl, leukocyte count 7800/mm³, ESR 152 mm/h, reticulocyte count 1.2 %, BUN 43-80 mg/dl, creatinine 1.4-2.4 mg/dl. Both Coombs test and FANA were positive, numerous L. E. cells were seen in the peripheral blood, complement component (C₃) was 22 mg/dl, latex agglutination test for rheumatic factor was negative. An IVP showed bilateral kidney enlargement. Liver function tests were normal except for the prothrombin time and PTT which were 46" (normal 12") and 210 "(normal 60-120") respectively.

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The patient was treated with prednisolone (15 mg/day) and azathioprine (2 mg/kg/day) and was discharged under this treatment.

In September of 1976 she was rehospitalized with the complaints of involuntary movements involving all four extremities, head, face, and difficulty in speaking and walking. After admission, she showed continuous uncoordinated movements of all extremities, trunk, head, and face. Physical examination was otherwise as previously.

Laboratory Findings: Hb 8.70 g/dl, leukocyte count 5000/mm³ with normal peripheral smear. No hemolytic Streptococci was found in the throat culture. ASO was 166 Todd U/ml, ESR 125 mm/h. Both Coombs test and FANA were positive and numerous L. E. cells were again seen in the peripheral blood. Serum complement (C₃) was 48 mg/dl. Endogenous creatinine clearance was 58.08 ml/min/m². Urine analysis showed moderate proteinuria and there was red blood cells, leukocyte and granular casts in the urinary sediment. Percutaneous renal biopsy showed changes compatible with active proliferative SLE nephritis.

On the day of admission, phenobarbital (3 mg/kg) and lithium (15 mg/day) were started. Within 7 days the patient's involuntary movements were suppressed with this therapy.

Discussion

Although neurological complications of SLE are common, chorea is rare. We found 31 reported cases in the literature up to 1975 in which chorea was described in association with SLE.⁶ Commonly chorea preceded the diagnosis of SLE by weeks or months but rarely by years.^{3,7} When chorea followed the diagnosis of SLE it appeared a few weeks or as late as 8 years after the diagnosis of SLE. The time relation between chorea and SLE shows no characteristic pattern.⁶ In this case chorea appeared two months after the diagnosis of SLE. The case is very interesting since chorea occurred while the patient was under corticosteroid and azathioprine treatment. The duration of chorea varies from few days to three years.^{8,9} The motor dysfunction in our case was suppressed within a week.

Investigators have not previously shown specific lesions in the brain which correlate with the clinical picture of chorea associated with SLE. The distribution of lesions in the brain such as involvement of the smaller arteries with fibrinoid necrosis, thrombosis, perivascular hemorrhages, small infarcts and encephalomalacia, varies in reported cases.^{5, 10-12} The pathophysiology of chorea is unknown,^{6, 13} but some investigators

suggest that chorea may be related to an increase in the striatal dopamine level.¹⁴ Since Haloperidol inhibits the activity of dopamine at central receptor sites it has been used for the treatment of chorea in SLE.^{14,15} Others have treated their cases with prednisolone and/or mercaptopurine.¹⁶ Chorea was diagnosed in our case while under treatment with prednisolone 15 mg/day and azathioprine 2 mg/kg. After diagnosing chorea librium 15 mg/day and phenobarbital 3 mg/kg were added to the treatment and prednisolone was increased to 60 mg/m².

Central nervous system involvement is commonly a secondary cause of death to renal disease in reported SLE cases. Although chorea was seen while the patient was receiving prednisolone and azathioprine, it has been suggested that the level of drugs in the CNS was too low to be effective and that some immunosuppressive drugs might be given by direct intrathecal administration before the appearance of overt neurological complications as in leukemia.¹⁷

Summary

A case with systemic lupus erythematosus and chorea is presented. Chorea occurred two months after the diagnosis of SLE was made and while the patient was under treatment, using prednisolone and azathioprine. The involuntary movements of the patient were controlled with azathioprine 3 mg/kg, prednisolone 60 mg/m², librium (Chlordiazepoxide hydrochloride) 15 mg/day, and phenobarbital 3 mg/kg within a week.

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