CHOREOATHETOSIS AND SEIZURES ASSOCIATED WITH SYSTEMIC LUPUS ERYTHEMATOSUS — A CASE REPORT

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SYNOPSIS

A 15-year old girl with systemic lupus erythematosus (SLE) presenting with convulsive seizures and choreoathetosis is reported. Treatment with prednisolone resulted in a prompt and complete subsidence of symptoms. Movement disorders are uncommon manifestations of SLE, and a combination of seizures and choreoathetosis in SLE is, we believe, the first reported case in the local literature.

INTRODUCTION

Neurological complications of systemic lupus erythematosus (SLE) are common (1-3), but chorea is rare. Johnson and Richardson (1), in 1968, reviewed 26 reported cases of SLE with choreoathetosis. Lusins and Szilagyi (4), in 1975, reported three cases of chorea associated with SLE and presented their clinical features as well. They also reviewed 31 cases from world literature, of which 21 cases had been reviewed earlier by Johnson and Richardson (1). Tay and Khoo (5) studied 75 cases of SLE with neurological involvement in 1971 and mentioned one case of hemichorea without specific clinical information. Feng (6) in a review of 183 patients over a ten year period found that neuro-psychiatric manifestation although rare at diagnosis were observed in 56 patients. Of these, commoniest were organic brain syndrome (47%), seizures(24%)and movement disorders(11%).

There has not been any previous report in the literature, to the authors' knowledge, of seizures occurring in association with chorea in SLE at the time of presentation. This paper reports a case of grand mal seizures and choreoathetosis in a young female with SLE. The patient fulfilled the American Rheumatism Association criteria for SLE (7).

CASE REPORT

A 15-year old Chinese girl presented on May 6, 1982 with a one-week history of fever and an episode of grand mal seizure on the day of admission. The patient was well prior to this admission and there was no past history of epilepsy. Nor was there previously any change in behaviour or presence of involuntary movements observed by her family. She had no antecedent respiratory infection or pharyngitis.

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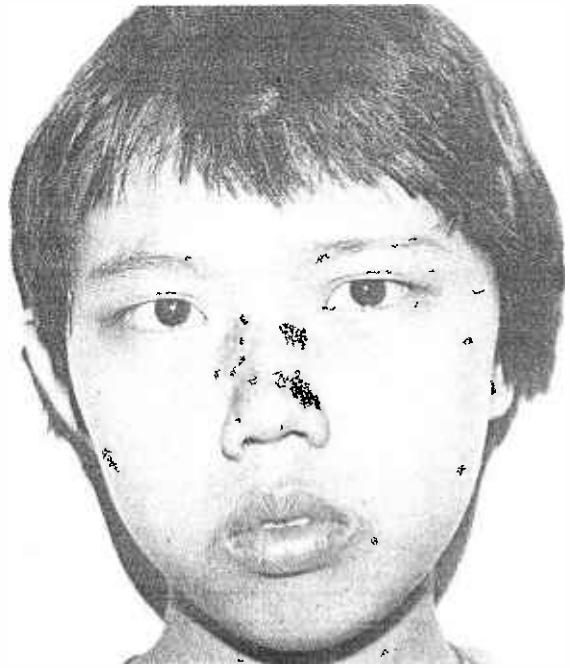

Fig. 1: The 15-year old patient with the facial erythema of SLE

On admission, she was conscious and alert, with a pyrexia of 39°C. Facial erythema was present (Fig. 1). Bilateral choreoathetoid movements involving the extremities, face, trunk and tongue were noted. The pupils were equal and reactive to light. The deep tendon reflexes were present and equal, and the plantar responses were normal. All modalities of sensation were intact. There were no signs of meningeal irritation.

Laboratory data revealed a hemoglobin level of 11.2 g%, thrombocytopenia of 35,000/ul and leucopenia of 3000/ul. The sedimentation rate was 14 mm/Hour. The antinuclear antibody test was positive on four occasions and antibody to extractable nuclear antigen (ENA) positive on two occasions. The serum complement CH50 level was less than 10 units. The anti-doublestranded DNA antibody test was negative. Lupus erythematosus (LE) cell preparations were negative on four occasions. The Rose Waaler test was negative. The spinal fluid was clear and colourless. There were 6 cells. The protein was 30 mg/dl and the glucose 50 mg/dl. Spinal fluid isolation for neurotropic viruses did not yield significant titres. The antistreptolysin 0 titres were negative in 100 Todd units on four separate occasions. Electroencephalogram showed mild bilateral nonspecific disturbances. The electrocardiogram and roentgenograms of the chest and skull were normal. Urinalysis was normal and the blood urea level was 9 mg/dl.

Seizures were initially treated with phenytoin sodium and phenobarbitone. One week after admission, when the diagnosis of SLE was confirmed, therapy with prednisolone, 45 mg/day, was started. A week later, there was a decrease in the abnormal involuntary movements and seizures. The patient was discharged on June 7, 1982, on a regimen of prednisolone together with phenytoin sodium and phenobarbitone.

On June 16, 1982, the patient was readmitted with oral mucosal ulcerations and increased facial erythema. No seizures or choreoathetosis was present. She had stopped the treatment prescribed and had taken Chinese herbal medicine instead. She had normal leucocyte and platelet counts and the sedimentation rate was 15 mm/Hour. Prednisolone was restarted and the patient improved.

Follow up one month later showed that the patient was well with no recurrence of chorea and seizures. She was maintained on prednisolone on a reduction regimen, and is now on a maintenance dose of 10 mg b.d.

DISCUSSION

Although seizures are commonly observed in the course of SLE, chorea is relatively rare. The reported frequency of convulsive seizures ranges from 8 per cent (8) to 54 per cent (1). The incidence of chorea associated with SLE has not been documented but Lusins and Szilagyi (4) have reviewed 31 cases from world literature and further reported three cases. Tay and Khoo (5) studied 75 patients with SLE and reported only one case of chorea. Feng (6) found 24 per cent with seizures and 11 per cent with movement disorders.

Seizures (9, 10) and chorea (11, 12)have been reported as initial signs and symptoms of SLE, but can also occur late in the disease. Our patient had both seizures and choreoathetosis when first presented. The presence of choreoathetosis in this patient had prompted the exclusion of Sydenham's chorea in rheumatic fever in view of the not infrequent occurrence at this age. Antistreptolysin 0 titres were elevated in three cases reviewed at the time of chorea (11, 13, 14) and the possibility that these subjects had two concomitant or related disease processes cannot be excluded. The antistreptolysin 0 titres of our patient were not elevated.

It is of interest to note that the LE cell preparation was negative in our patient. Fries and Holman (8) commented on the unusually high frequency of positive LE cells of 92 per cent seen in the preliminary criteria for the classification of SLE of the American Rheumatism Association, and reported a 40 per cent positivity in their studies but noted that only half of the patients had a positive test at the onset. Tay and Khoo (5) reported positive LE cells in 71 per cent of patients. Dubois (15) also showed positive LE cells in 71 per cent of proven SLE patients.

Fries and Holman (8) gave a frequency of anti-ENA and anti-DNA antibodies of 100 per cent and 52 per cent, respectively, in patients with SLE, and emphasized that antinuclear antibodies are the rule in SLE. Both the antinuclear antibody and antibody to ENA were present in our patient.

Autopsy investigations to correlate the clinical picture of chorea in SLE failed to localise any specific lesions of the brain. Johnson and Richardson (1) found that the gross and microscopic examinations of the brain could not explain the neurological manifestations. Findings included proliferative vascular lesions and thrombotic occlusions of leptomeningeal and parenchymal vessels, encephalomalacia, and petechial hemorrhages throughout the brain. Lesions in the basal ganglia were noted in only one case (16). These consisted of intimal proliferation of arterioles with occulsion of vessels by thrombi and concomitant infarction in the lenticular nuclei.

Several mechanisms for central nervous system disease in SLE had been proposed. These included active small-vessel arteritis (8); strategically placed lesions throughout the brain in a pathway involving the cortex, basal ganglia and thalamus, resulting in dyskinesias (17); gamma globulin, especially IgM, deposition in the choroid plexus of the central nervous system (18); and alterations in the dopamine levels in the basal ganglia in cases of chorea (19).

Corticosteroid therapy has been used for central nervous system manifestations in SLE (2, 8, 15). Rapid and dramatic improvement in chorea after administration of prednisone, 30-60 mg/day, has been reported (4, 8, 11, 20). Our patient was treated with prednisolone, 45 mg/day, and improvement in abnormal involuntary movements and seizures was evident a week after institution of therapy. The use of haloperidol in the treatment of chorea

associated with SLE has also been reported with success (4, 19, 21). Haloperidol is thought to inhibit dopamine at the central receptor sites in the basal ganglia (19).

Although the pathophysiology of chorea is still not clear, Fermaglich, Steib and Auth (19), contend that it is a dopaminergic phenomenon. The efficacy of corticosteroids and/or haloperidol in the treatment of chorea associated with SLE favours the involvement of both immunological and biochemical processes in its pathogenesis.

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