Systemic lupus erythematosus presenting as chorea

Systemic lupus erythematosus (SLE) is uncommon in childhood and is predominantly a female disease (80–90%)(McLean et al., 1975). Multisystem involvement most commonly includes skin, central nervous system (CNS), and kidney. CNS disease is present in 20 to 75% of patients (Johnson and Richardson, 1968; Singsen et al., 1976) and is the second most common cause of death. Although there may be numerous presentations of CNS disease, chorea as the initial presentation is rare; there are only 31 reported cases (Lusins and Szilagyi, 1975) and few of these were children. We report a boy who presented with chorea and had a dramatic response to haloperidol.

Case report

An 11-year-old Caucasian boy was admitted to our hospital on 11 August 1976. Approximately one month before admission he developed an erythematous maculopapular rash on the face, arms, and palms. The lesions on the arms were scaly and located more on the extensor surfaces. He had been treated symptomatically with diphenhydramine (Benadryl).

Three days before admission he noticed weakness, and was unable to sit well, eat, or dress himself because of awkward movements. His mother also noted emotional lability and slurred speech as well as facial grimacing. His history was negative and the family history was negative for any collagen-type disease.

He weighed 51.7 kg (6.8 kg weight loss in the previous 4 weeks). Blood pressure was 120/70 mmHg. An erythematous maculopapular rash was present on the face in the malar areas, and on the extremities and trunk, particularly the buttocks. On neurological examination he was alert, cooperative and oriented. His speech was slurred, and he showed facial grimacing, choreoathetoid movements, and evidence of dyskinesia. Initial blood count showed Hb 11.4 g/dl, haematocrit 33%, WBC 4.2 × 10^9/l with 69% polymorphonuclears, 29% lymphocytes, and 2% eosinophils; erythrocyte sedimentation rate 42 mm/h; blood urea nitrogen 26 mg/100 ml (9.28 mmol/l). Repeat blood urea nitrogen examinations were normal. Initial LE cell preparation was negative, rheumatoid factor weakly positive, fluorescent antinuclear antibody weakly positive, urinalysis negative, x-ray studies within normal limits. Initial electroencephalogram (EEG) showed nonspecific slowing. Sydenham's chorea was ruled
out on the basis of negative antistreptolysin O titres, normal electrocardiogram, normal chest x-ray, and a normal cardiac evaluation.

He was started on haloperidol 0·5 mg twice daily on admission. Within 48 hours his symptoms had dramatically cleared. Repeat EEG after 2 days of haloperidol was normal. He was maintained on haloperidol for another 7 days. After hospitalisation, he developed fever, particularly in the afternoon, to 38·3–38·9°C, as well as intermittent arthralgia in the knees and shoulders. One week after admission the WBC count fell to 3·2 × 10⁹/l and the platelet count, initially normal, dropped to 100 × 10⁹/l. Haematocrit had dropped to 30 % and Hb to 10·6 g/dl. Serum electrophoresis showed a reverse albumin/globulin ratio—globulin 37 g/l, albumin 29 g/l, and raised γ-globulin 303 g/l (normal 120–210 g/l). Direct and indirect Coombs’s tests were positive. Repeat examinations of urine were negative and renal function studies normal. One week after admission his fluorescent antinuclear antibody changed from weakly reactive to strongly positive. An anti-DNA titre was also positive at this time (1:1200). C3 was 0·1 g/l (normal 0·55–1·2 g/l) and C4 was 0·04 g/l (normal 0·2–0·4 g/l). A skin biopsy on 25 August showed vasculitis compatible with SLE (Fig.). He was started on prednisone, 60 mg/day, and followed as an outpatient.

In October 1976 he presented with high fever (40°C) and arthralgia. He was hospitalised and treated with increased doses of steroid. Antinuclear antibody was 1:1280 and C3 0·33 g/l. He had pyuria and WBC casts at this time also, but repeat urine cultures were negative. Six months after the initial diagnosis his urine began to show protein, cells, and casts. The urine sediment continues to be positive but renal function studies are within normal limits. He continues to manifest active disease as determined by raised antinuclear antibody levels and depressed complement (C3, C4).

Discussion

This child demonstrated an acute onset of chorea as a presentation for SLE. His symptoms dramatically resolved with haloperidol within 48 hours and the EEG returned to normal. Haloperidol has been given to adults with SLE for chorea with dramatic improvement (Fermaglich et al., 1973). Our patient, at the time of diagnosis, did not manifest renal disease. It has been suggested that renal and CNS involvement occur together infrequently (Singsen et al., 1976). The depressed complement levels, we feel, reflect CNS and skin involvement. Our patient also had a low C4 which may correlate with CNS lupus as shown by Halder et al. (1973).

Summary

A child presenting with chorea developed the full clinical and laboratory findings of systemic lupus erythematosus after hospitalisation. He responded dramatically to haloperidol. 6 months after diagnosis he began to manifest renal involvement.
Serum 25-hydroxyvitamin D levels in thalassaemia

In patients with hepatic dysfunction low levels of serum 25-hydroxyvitamin D (25-OHD) have been reported (Hepner et al., 1975; Long et al., 1976; Olson et al., 1976; Wagonfeld et al., 1976). These authors stress the importance of further studies in order to correlate the low levels of serum 25-OHD in cirrhotic patients with their bone lesions. It is well known that thalassaemic children have bone lesions and hepatosplenomegaly with evidence of hepatic dysfunction in some cases. This led us to investigate serum 25-OHD in thalassaemia.

Material and methods

Our patients were 36 children, 20 boys and 16 girls, with homozygous β-thalassaemia aged from 5 to 15 years, and 27 controls (19 boys, 8 girls) with the same age range. The controls were chosen from children coming to the hospital for tonsillectomy or adenoidectomy. In the thalassaemic children blood specimens were taken at least 10 days after the last blood transfusion and haemoglobin was above 8 g/dl.

25-OHD was estimated in the serum of all children by the competitive protein-binding assay as described by Edelstein et al. (1973). Venous blood (7 ml) was taken in both groups for routine haematological examination and part of the specimen was used for this study. Serum was separated about half an hour after collection, kept at −20°C, and estimations were performed not later than 7 days after collection.

Blood specimens were collected between the beginning of April and the end of October 1976. We considered as the winter period the first 3 months of observation (1 April–30 June) and the summer period the last 4 months (1 July–31 October).

Results

Table 1 gives the mean values and standard deviations of serum 25-OHD in thalassaemic and control children. As the table shows, thalassaemic children had lower levels of serum 25-OHD than controls (t=2.4, P<0.01).

Table 2 gives the seasonal variation of serum 25-OHD in both groups. Serum 25-OHD is lower in thalassaemic patients than in the control group at both winter and summer periods. (Winter t=3.7, P<0.0025; summer t=1.7, P<0.05.) Serum levels of 25-OHD in both groups were higher during summer. In the thalassaemic group this difference was more marked (t=2.9, P<0.005).

Table 1 Serum 25-OHD in control and thalassaemic children

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<th>No. of cases</th>
<th>Serum 25-OHD (ng/ml) (mean ± SD)</th>
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<tr>
<td>Controls</td>
<td>27</td>
<td>18.6±8.6</td>
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<tr>
<td>Thalassaemic children</td>
<td>36</td>
<td>12.8±9.9</td>
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Table 2 Seasonal variation of serum 25-OHD in the two groups

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<th>Serum 25-OHD (ng/ml) (mean ± SD)</th>
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<tbody>
<tr>
<td></td>
<td>Winter period</td>
</tr>
<tr>
<td>Controls</td>
<td>14.0±5.4 (9)</td>
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<tr>
<td>Thalassaemic children</td>
<td>5.6±4.4 (10)</td>
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Number of patients in parentheses.