TRANSVERSE MYELOPATHY AND HYPERPHAGIA IN SYSTEMIC LUPUS ERYTHEMATOSUS: A CASE REPORT

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SUMMARY

We report a case of systemic lupus erythematosus complicated by transverse myelopathy and hyperphagia. To our knowledge the latter has not been reported before.

INTRODUCTION

Neurological manifestation of systemic lupus erythematosus (SLE) have been observed as early as 1875.[1] These included subtle personality changes to frank psychosis, fits, peripheral neuropathy and strokes. Until 1978 only 18 authors have reported a total number of 26 cases of transverse myelopathy in SLE.[2] To our knowledge hyperphagia complicating SLE has never been reported before. We present such a patient who was seen and managed at the Department of Medicine, Universiti Kebangsaan Malaysia (National University of Malaysia).

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CASE REPORT

TMY, a 10-year-old Chinese female was well till May 1983 when she developed severe vomiting, epistaxis, fever, facial rash, alopecia, mucosal ulceration and arthritis of both knees. On admission to a private hospital, laboratory studies confirmed a diagnosis of systemic lupus erythematosus (SLE). The patient was commenced on dexamethasone 6 mg and azathioprine 50 mg daily. She remitted with the treatment until February 1984, when she was referred to our clinic with exacerbation of the disease involving mainly the neurological system.

She presented with progressive weakness of both her lower limbs, slowing of speech together with bowel and urinary incontinence. Occasionally she had myoclonic jerks. Physical examination then showed a well nourished child with Cushingoid facies. She had alopecia and diffused vasculitic lesions in the palms and soles. Fundoscopy revealed crops of cytoid bodies. Neurological examination showed a withdrawn, inattentive, irritable and mentally slow child with obvious pyramidal tract signs in the lower limbs as evidenced by hypertonia, hyperreflexia, clonus and positive Babinski sign bilaterally. The sensory level was probably mid-thoracic. The cranial nerves were intact.

Investigation showed a normal leucocyte count and mild anemia. Cerebrospinal fluid (CSF)

examination was normal. Antinuclear antibody and lupus erythematosus (LE) cells were positive. Both complements C3 and C4 were slightly decreased and circulating immune-complex test was positive. An electroencephalogram (EEG) done four weeks later was reported as within normal limits. She was given intravenous methylprednisolone 500 mg and oral prednisolone 40 mg daily. Cyclophosphamide 50 mg/day and clonaxepam 1.5 mg/day were subsequently added to the treatment regime. During the two months stay in the ward she made considerable improvement and on discharge was able to talk normally and walk unaided although there were still obvious neurological signs in the lower limbs. Her bowel and urinary incontinence improved.

Five months later, following an upper respiratory tract infection, she again became withdrawn and was unable to walk without support. She developed urinary and bowel incontinence again. In addition, the patient's mother reported that she had excessive appetite resulting in marked increase in weight over a two-week period. Similar behaviour was observed in the ward. Vasculitis was again noted in the palms and soles. Neurological examination was essentially the same as the previous admission with grade 3 – 4 power in the lower limbs. A repeat lumbar puncture was unremarkable. She again responded well to intravenous methylprednisolone. The hyperphagia disappeared after one week and she was discharged walking unaided. She has since remained well on follow ups.

DISCUSSION

Andrianakos *et. al.*,[3] reported three cases of transverse myelopathy in systemic lupus erythematosus and reviewed the literature of 23 previously reported cases. The most common presenting symptoms of myelopathy were numbness and weakness of the legs. Other initial neurological manifestations included urinary retention, faecal incontinence, low backache, mid scapular and abdominal pain and paraesthesia in the legs. The interval between the presenting neurological symptom and the maximum deficit ranged from seven hours to four months but was usually of short duration, being less than 24 hours in ten patients and less than two days in three patients.

Paraplegia, the most common motor deficit, occurred in 19 of 26 patients. Quadraplegia manifested initially in one patient and six patients had paraparesis. There was loss of urinary bladder and rectal sphincter tone in all patients. The sensory losses were at a cervical level in four patients, a thoracic level in eleven and lumbar level in three.

The prognosis appeared to be significantly affected by the development of transverse myelopathy. Thirteen patients died two days to 34 months after onset of spinal cord involvement. Nine succumbed within 45 days. Four patients died of septicaemia, two of pneumonia, one of pulmonary embolism, one of pulmonary oedema and two of widespread vasculitis associated with cerebral haemorrhage. The cause of death remained unknown in three patients. Autopsies done on twelve patients showed that ischaemic necrosis was the cause of myelopathy in most instances.

Hyperphagia was a striking feature during the second admission. We believe this has never been reported before. In humans, appetite for food depends primarily upon the interaction of the "feeding centre" and "satiety centre" located in the hypothalamus. Stimulation of the former evokes eating behaviour whereas its destruction causes severe fatal anorexia. Conversely stimulation of the latter causes cessation of eating whereas its destruction causes hyperphagia. We postulate that there was a lesion at the "satiety centre" which led to the hyperphagic behaviour.

REFERENCES

1 Dubois E L. *Lupus Erythematosus* (2nd ed). Los Angeles : University of Southern California Press, 1974.

2 Hachen H J, Chantraine A. Spinal cord involvement in systemic lupus erythematosus. *Paraplegia* 1979; 3 : 337 – 346.

3 Andrianakos A A, Duffy J, Suzuki M. Transverse nyelopathy in systemic lupus erythematosus. *Ann Intern Med* 1975; 83 : 616–624.