Central Nervous System Lupus: Concomitant Occurrence of Myelopathy and Cognitive Dysfunction


Summary

Reported are two cases of patients with central nervous system systemic lupus erythematosus (SLE) with clinical features of a myelopathy confirmed by magnetic resonance imaging (MRI). In one case, further evaluation including Single Photon Emission Computerized Tomography (SPECT) and neuropsychological testing, indicated higher cortical deficits which improved after treatment with monthly pulse intravenous cyclophosphamide. Subsequent cessation of cyclophosphamide was associated with further progression of the neurological disease in this patient.

Key words


INTRODUCTION

The manifestations of cerebral involvement in systemic lupus are protean, ranging from focal abnormalities to global cerebral dysfunction (1). One of the most serious complications is lupus myelopathy which can be more easily demonstrated since the advent of MRI-scanning (2-4). There are anecdotal reports on the beneficial effect of steroids and cyclophosphamide (5,6) in lupus myelopathy but controlled trials are lacking.

Cerebral involvement including cognitive impairment in SLE patients has recently received increased attention (7,8); however, the diagnosis remains difficult since no specific diagnostic tests are available. Neuropsychological testing has been proposed to be a sensitive investigation to detect cognitive dysfunction in cerebral lupus (7). More recently, SPECT-scans have been used to demonstrate regional cerebral blood flow abnormalities in the evaluation of cerebral lupus (9-11).

We report here two patients with SLE who had lupus myelopathy and concomitant cerebral involvement. In one patient treatment with monthly IVI pulse cyclophosphamide resulted in improvement of the clinical status as well as SPECT- and neuropsychological testing. These cases reinforce the need for a full systematic evaluation when assessing SLE patients with central nervous system symptoms or signs. It also shows the potential value of using different diagnostic techniques not only to diagnose cerebral lupus, but also to help assess the efficacy of treatment.

CASE REPORTS

Case 1

A 26-year-old white female presented in May 1984 with an acute onset of polyarthritis and butterfly facial rash. A positive antinuclear antibody (ANA, 1:640, speckled pattern) and raised anti-double stranded DNA-antibodies (dsDNA) led to the diagnosis of SLE. She was treated with NSAID and remained well until June 1986 when she suddenly developed a paresis of the left leg accompanied by paraesthesias. There was also transient mild weakness of the right leg. Myelography was normal and computerized tomography of the brain demonstrated multiple ischaemic zones in the corona radiata on the right and the left. The diagnosis of cerebral lupus was made and treatment with IVI pulse steroid resulted in almost complete resolution of all neurological deficits. The patient was subsequently commenced on hydroxychloroquine 200 mg/day and aspirin and remained well for 6 years. In September 1992 of her own volition she stopped all medication. One week later she...
noted low back pains which radiated into both groins and a feeling of heaviness in both legs without sensorimotor deficit. One day later she developed acute urinary retention with preserved bladder sensation. Otherwise, the physical examination was unchanged.

Catheterization of the bladder showed a resting volume of 1400 mls of urine in the bladder. Spinal fluid examination was normal and there was no evidence of intrathecal immunoglobulin production by IgG index determination. MRI of the spinal cord revealed a pencil-shaped signal in the conus medullaris with swelling (Fig. 1). MRI of the brain demonstrated three hyperintense structures in the parietal lobes bilaterally resembling old infarcts (Fig. 2) and a single gadolinium-enhancing lesion in the left parietal lobe (Fig. 3). Serological investigations were as follows: ANA 1:640, speckled pattern, anti-dsDNA-antibodies 29 (normal up to 7 IE/ml), C-reactive-protein (CRP) 0.92 mg/dl (normal up to 0.5 mg/dl), normal complement (C3, C4 and CH 50) and weakly positive anti-cardiolipin-antibodies. The diagnosis of acute lupus myelopathy was made and the patient was treated with pulse corticosteroids 500 mg intravenously, daily for 3 days. She experienced a complete resolution of the bladder atony without a resting urinary volume. She was subsequently recommenced on hydroxychloroquin and has remained well.

**Case 2**

A 26-year-old female had suffered from migraine since 1980 for which she saw a neurologist in 1987 who found no neurological abnormality. In November 1989 she developed lethargy, malaise, polyarthritis of the right shoulder, the small joints of both hands and both wrists as well as morning stiffness and Raynaud's phenomenon. Serological investigations showed a positive ANA 1:640 with speckled pattern, positive rheumatoid factors (Rose-Waaler 1:256) positive ribonuclear-protein-antibodies but negative anti-cardiolipin-antibodies and dsDNA-binding. The diagnosis of undifferentiated connective tissue disease was made and she was intermittently treated with low dose steroids with good response.