Aseptic Leptomeningitis in Systemic Lupus erythematosus
A Case Report

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Abstract
Objective: Systemic lupus erythematosus (SLE) with leptomeningeal involvement is a rare condition. We report a case in a 17-year-old woman. Clinical Presentation and Intervention: The patient was admitted to hospital with the complaints of vertigo, nausea, vomiting, headache, diplopia, ptosis on the left and weakness of the left leg. A diagnosis of SLE was established, with diffuse leptomeningeal involvement demonstrated by cranial magnetic resonance imaging. When treated with steroids, the clinical problems resolved almost completely. Conclusions: Although rare, leptomeningeal involvement can complicate SLE. Cranial magnetic imaging can demonstrate tissue involvement. The condition responds to steroid therapy.

Introduction
Systemic lupus erythematosus (SLE) is a chronic inflammatory disease that is thought to be mediated by an autoimmune process. It is seen most frequently in women and is usually diagnosed in the 3rd decade of life. Clinically detectable central nervous system (CNS) involvement has been reported for 25–75% of patients with SLE [1]. Patients with SLE often suffer from headaches. Other manifestations of CNS involvement include epilepsy, psychosis, focal infarcts, extrapyramidal symptoms, cerebellar dysfunction, pseudotumor cerebri, papilledema, subarachnoid hemorrhage, aseptic meningitis, transverse myelitis, optic neuritis and cranial nerve palsies or sensory neuropathy [2]. There are few reported cases of SLE with diffuse leptomeningeal involvement [3, 4].

Case Report
A 17-year-old woman was admitted to hospital with a 40-day history of vertigo, nausea, vomiting, headache and a 25-day history of diplopia, ptosis on the left and weakness of the left leg. Personal and family histories were unremarkable. Physical examination showed diffuse thyroid gland enlargement. Neurologically, the patient was confused and had signs of meningeal irritation (neck stiffness and Kernig’s sign). Cranial nerve examination showed ptosis of the left leg.
eyelid, restriction of adduction (3rd cranial nerve palsy), abduction (6th cranial nerve palsy) and vertical gaze upward in the left eye movements and bilateral papilledema. Motor examination revealed that the left leg (proximal and distal muscles) was only able to resist gravity (2/5). Other extremities were found to have normal motor properties (5/5). Cerebellar and sensory examination results were found to be normal. Tendon reflexes were bilaterally absent in the lower extremities. Babinski response was positive in the left leg and negative in the right leg.

Laboratory examination gave the following results: platelets 34,000/µl, Hb 9.6 g/dl, direct and indirect Coombs tests (+) and activated partial thromboplastin time was prolonged. An unenhanced brain CT scan showed diffuse edema. Electroencephalographic (EEG) evaluation on admission showed a baseline activity of 2–4 cyc/s delta waves and periodic 2 cyc/s slow waves. Cranial MRI showed contrast enhancement at the superior sagittal, right transverse and sigmoid sinuses, leptomeninges and dura (diffuse) (fig. 1, 2).

The patient was started on dexamethasone 16 mg/day. One day after initiation of treatment the weakness in the left leg recovered completely.

Immunological studies showed: anti-ds-DNA 12 IU/ml (0–7 IU/ml), antinuclear antibodies (+), LE cells (-), cardiolipin IgM antibody 34 mplU/ml (normal range: below 10), cardiolipin IgG antibody 15 mplU/ml (normal range: below 14). With respect to syphilis serology, a venereal disease research laboratory (VDRL) test was positive at first examination but during the second it was negative and Treponema pallidum immobilization test was negative, suggesting that the first test was false positive. Twenty-four hours urinary protein was 1,056 mg.

Cerebrospinal fluid (CSF) studies, performed after CT and MRI, showed that opening pressure was 51 cm H2O, erythrocyte count was 120/mm3, white blood cell count was negative, glucose was 39 mg/dl, simultaneous blood glucose was 89 mg/dl, chloride was 115 mg/dl and protein was 56 mg/dl (moderately elevated). CSF cultures were negative for routine microorganisms and tuberculosis; PCR for tuberculosis was also negative. PPD was negative. CSF serological examination was negative for HSV-1 and HSV-2.

The diagnosis of SLE was made according to the ARA criteria [2] on the 7th day of admission on the basis of the following clinical and laboratory findings: thrombocytopenia, immunological abnormalities, anti-ds-DNA 12 IU/ml, false positive VDRL tests, positive antinuclear factor, hemolytic anemia and renal involvement. Treatment with methylprednisolone at a dosage of 60 mg/day was given and dexamethasone therapy was stopped.

Meningeal irritation signs (neck stiffness and Kernig’s sign) were absent on the 11th day of treatment. Repeat CSF examination after 3 weeks revealed an opening pressure of 54 cm H2O, glucose 38 mg/dl, simultaneous blood glucose 52 mg/dl, protein 10 mg/dl and no cells were apparent on cytological examination. A 9–10 cyc/s alpha parieto-occipital background activity was seen on the repeat EEG. Steroid treatment at a dosage of 60 mg/day led to the complete recovery of neurological functions on the 21st day of treatment. MRI was not repeated because of financial constraints.

Discussion

SLE is diagnosed according to the SLE Classification Criteria published in 1982 [2], including: malar rash, discoid rash, photosensitivity, oral ulcers, arthritis, renal involvement, neurological involvement, hematological

![Fig. 1. An axial T1-weighted image of the brain. The scan image shows leptomeningeal involvement.](image1)

![Fig. 2. T1-weighted MR image showing dural involvement.](image2)
Leptomeningeal Involvement in SLE

involvement, immunological involvement and presence of antinuclear antibodies. Any combination of four of these criteria has a specificity of 98% and sensitivity of 97% [2]. The findings in our case fulfilled four of the eleven items for the classification of SLE. The SLE antiphospholipid syndrome [2, 5] was diagnosed because of the presence of a prolonged activated partial thromboplastin time, thrombocytopenia and antcardiolipin antibodies.

Clinically, two types of CNS involvement are seen in SLE: focal neurological symptoms and mental symptoms [3]. The predominant lesions are multifocal microinfarcts and increases in microglial cells around the small arteries and capillaries [6]. In 1991, investigators reported on 12 SLE cases with neurological involvement: 8 of them had infarction, 2 had intracranial hematomas and 2 had thromboembolic processes [7]. CSF pressure may be elevated in SLE. Elevated levels of CSF protein are thought to be due to plasma infiltration through an impaired blood-brain barrier resulting from increased vascular permeability because of vasculitis [8]. On computed tomography, edema and small infarctions are seen [8]. Our case showed moderately increased protein in CFS and elevated CSF pressure.

Leptomeningeal involvement has not been reported frequently. Okano et al. [3] reported 1 case in which a 41-year-old patient was admitted with mental symptoms which improved with steroid therapy. Bertrand et al. [9] reported histopathological changes in leptomeningeal and cerebral blood vessels in a 46-year-old woman who died of a stroke after a 19-year history of subacute cutaneous lupus erythematosus with CNS involvement. In this case, general autopsy showed changes in the kidneys, myocardium, spleen and pancreas typical of SLE [9]. Clinical features of our case were similar to the cases reported by Okano et al. [3]. Steroid treatment has been shown to be effective in restoration of the blood-brain barrier for vasculitis or lupus microangiopathy [8].

MRI provides a sensitive method for studying cerebral lesions. In CNS involvement of SLE, MRI reveals high signal intensities in the subcortical white matter [10, 11]. In our case, cranial MRI showed contrast enhancement at the superior sagittal, right transverse and sigmoid sinuses, leptomeninges and dura (diffuse).

Conclusion

Although rare, leptomeningeal involvement can complicate SLE. MRI provides a sensitive method for studying cerebral lesions. This case of SLE with diffuse leptomeningeal involvement showed complete response to steroids.

References


