Case report

Transverse myelitis occurring during pregnancy in a patient with systemic lupus erythematosus

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SUMMARY Myelopathy is a well recognised but infrequent neurological manifestation of systemic lupus erythematosus (SLE). The case of a 27 year old woman with SLE of seven years' duration who developed a spastic paraparesis during her second pregnancy is reported. Magnetic resonance imaging did not show any intrinsic abnormality of the spinal cord. Anticardiolipin antibody was weakly positive and C4 was low. The patient responded dramatically to steroids.

Key word: myelopathy.

Transverse myelitis is a rare but well recognised neurological manifestation of systemic lupus erythematosus (SLE). Lupus related myelopathy has not been previously reported in pregnancy.

We describe a patient with known SLE who developed an acute spinal cord lesion during the second trimester of pregnancy. Magnetic resonance imaging, recently reported to show abnormalities in the spinal cord of a lupus patient with transverse myelitis, showed no abnormal signals in this case.

Case report

The patient, a 27 year old woman, was diagnosed as having SLE in 1979 on the basis of strongly positive antinuclear antibody, oral ulceration, Raynaud's phenomenon, photosensitivity, pyrexia, arthralgias, and a skin biopsy which showed a positive lupus band test. Her first pregnancy in 1983 was complicated by hypertensive disease necessitating delivery by caesarean section at 32 weeks of a live female infant.

Early in her second pregnancy in 1986 she was admitted to hospital with hyperemesis gravidarum. This settled spontaneously and the patient was well until 16 weeks of gestation; she then presented with

a two day history of paraesthesiae in the legs spreading to involve the abdomen and thorax in association with weakness of the legs, which was worse on the right side. Awareness of fetal movements had been lost. Sphincter disturbance was present. Prednisolone 7 mg daily was the only drug treatment on admission. Examination showed spastic paraparesis with bilateral extensor plantar responses and a sensory level at approximately T3/T4. No other abnormal physical findings were apparent.

Investigations were as follows: full blood count showed a mild leucocytosis of $15.6 \times 10^9/l$; normal urea and electrolytes; erythrocyte sedimentation rate 76 mm/h (Westergren); antinuclear antibody 1/1024 with normal DNA binding of 27.8%; rheumatoid factor 1/256 (Rose-Waaler); positive anti-Ro and anti-La with negative anti-Sm and anti-RNP antibodies; C4 low at 110 mg/l (normal range 200-550); other complement components normal; anticardiolipin antibody 6 units (IgG; negative <5, low positive 5-20, definite positive >20). Viral titres were negative. In view of her advancing pregnancy lumbar puncture was not performed.

Magnetic resonance imaging of the spinal cord and posterior fossa showed no obstructive lesion or intrinsic cord abnormality. The final diagnosis was transverse myelitis.

Prednisolone dosage was increased to 30 mg daily, and the patient gradually improved over the following six weeks. Some mild pyramidal signs persisted.

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The remainder of her pregnancy was complicated by hypertensive disease, and a live female infant was delivered by caesarean section at 29 weeks' gestation. After the initial postpartum period steroid dosage was gradually reduced over a period of six months; sensory disturbance reappeared in association with extensor plantar responses, however. Prednisolone dosage was immediately increased to 60 mg daily with rapid improvement of symptoms.

Discussion

The effect of pregnancy on the manifestations of SLE remains controversial. Certainly, patients with mild or moderate disease do conceive, even in the presence of renal disease or while taking immunosuppressive drugs.¹ Patients who become pregnant are less likely to have a history of neuropsychiatric or serosal involvement.²

Current opinion suggests that major, systemic, non-renal manifestations of SLE during pregnancy are not usual, though the experience of some centres differs from this.^{1 3} In a series of 52 pregnancies in 39 patients with SLE reported by the UCLA group only four pregnancies were complicated by major systemic illness. None of these had neuropsychiatric disease and all had a satisfactory maternal and fetal outcome.¹ Conversely, a Mexican study looked prospectively at 102 pregnancies in 75 lupus patients. In their patients exacerbations of lupus were seen in almost 60% of cases. Eight patients had neurological disturbances, five had headache, and none of the others had focal neurological signs.⁴

The best pregnancy outcome can be expected if the disease has been quiescent during the three months before conception. Such patients tend to remain in remission during the pregnancy, but all patients are at risk of postpartum exacerbations.²

Neuropsychiatric manifestations of SLE occur in up to 50–60% of cases.^{5–8} Many of these patients have seizures or psychosis. Myelopathy is rare and may occasionally antedate other manifestations of lupus. Such cases are often labelled as multiple sclerosis.^{5 9 10}

The myelopathy usually presents acutely with paraesthesiae developing in the legs and usually ascending to the thorax over 24–48 hours.¹¹ This is accompanied by paraplegia, which is usually flaccid but may present with spasticity in up to 22% of cases.¹² Sphincter problems are universal. The level of the lesion is usually in the low to mid-thoracic cord.^{13 14} Diagnosis is based on clinical features supported by serological tests, cerebrospinal fluid examination, and various imaging modalities. Most patients with central nervous system lupus have a positive antinuclear antibody, though the titre is

often low. The value of cerebrospinal fluid examination is controversial, but abnormalities reported include pleocytosis, low C4, low glucose, altered immunoglobulin concentrations, and immune complexes.^{5–7 9 14} Many of these abnormalities can be found in infection and demyelinating disease. Cerebrospinal fluid examination in many patients with neuropsychiatric lupus is normal.

Differentiation of central nervous system lupus from demyelinating disease can be extremely difficult. Numerous reports show that 'typical' demyelinating syndromes, such as bilateral internuclear ophthalmoplegia, neuromyelitis optica, and spastic paraparesis, can and do occur in SLE.^{9 15} It was hoped that magnetic resonance imaging might help in the diagnosis of such cases. A number of reports have shown the existence of areas of altered signal intensity in the spinal cords of patients with multiple sclerosis.¹⁶ Recently, abnormal magnetic resonance imaging signals were detected in the spinal cord of a patient with SLE and Devic's disease.¹⁷ Magnetic resonance imaging failed to show any such abnormality in our patient.

Treatment of exacerbations of SLE during pregnancy has been based on increasing steroid dosage.¹ This is not without problems, particularly with respect to the exacerbation of hypertensive disease in pregnancy as occurred in our patient. In the management of myelopathy most authorities would recommend steroids, though there are some authors who maintain that this can actually worsen the condition.¹² Untreated lupus myelitis has an unfavourable outlook; death is common, residual neurological signs almost universal, and progressive spastic paraplegia well documented. Regardless of steroid treatment the prognosis for full neurological recovery is poor.^{12 14}

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