

Case reports

Acute transverse myelopathy complicating systemic lupus erythematosus

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SUMMARY A sixteen year old girl with systemic lupus erythematosus developed acute transverse myelopathy. She was treated with high dose steroids, cyclophosphamide, and plasma exchange and regained partial neurological function. Previous descriptions of transverse myelopathy complicating systemic lupus erythematosus are reviewed, with particular reference to the efficacy of high dose steroid treatment.

Transverse myelopathy is a rare complication of systemic lupus erythematosus, and the prognosis is generally poor.¹ Although it has been suggested that steroids confer no benefit,² there are several reports of complete or partial recovery associated with high dose steroid treatment. We describe a patient with transverse myelopathy associated with systemic lupus erythematosus treated, uniquely, with steroids, cyclophosphamide, and plasma exchange. Moreover, we review the published work concerning the treatment and prognosis of this condition, and the usual cerebrospinal fluid findings.

Case report

A sixteen year old negroid girl presented with a three month history of polyarthralgia and Raynaud's phenomena. Examination was normal apart from cyanotic mottling over the finger tips and florid splinter haemorrhages affecting the fingernails.

Initial investigations were as follows: antinuclear antibody titre 1/1024, DNA binding 21% (normal <20%), C3 0.59 g/l (normal 0.7-1.7 g/l), C4 0.12 g/l (normal 0.15-0.55 g/l). Despite treatment with prednisolone 60 mg daily the digital vasculitis progressed. Hence treatment with intravenous cyclophosphamide 1 g weekly was started, and she underwent three plasma exchanges, totalling nine litres. The digital lesions subsequently healed. Over the next eight months apart from mild arthralgia she was well, and treatment was reduced to prednisolone 2.5 mg daily. Subsequently she developed active

synovitis, associated with rising antinuclear antibody titres and falling complement concentrations, and was readmitted. Three days after admission she complained of mid-scapular pain and numbness affecting the legs, and became paraplegic over four hours. She had a sensory level at T7, below which pain and temperature sensation were absent, though vibration sense and proprioception were preserved. She developed urinary retention and faecal incontinence. Lumbar puncture showed normal cerebrospinal fluid pressure. Cerebrospinal fluid analysis showed protein 1100 mg/l (normal <600 mg/l), glucose 2.0 mmol/l (normal 3.9-6.1 mmol/l), and white cell count $21 \times 10^9/l$. Myelography was normal. A diagnosis of transverse myelopathy was made and on the day of onset she received methylprednisolone 1 g and cyclophosphamide 1 g, both intravenously. Prednisolone was increased to 80 mg daily. Over the next three days she underwent three plasma exchanges, totalling nine litres.

No further neurological deterioration occurred. After two months movement returned, initially to the right foot, and after four months bladder and bowel control returned. Eighteen months after the onset she had sufficient lower limb power to walk with the aid of crutches. Throughout the illness antiphospholipid antibodies were not detected in the sera.

Discussion

There are 44 reported cases of transverse myelopathy occurring in SLE,¹⁻³⁰ though the prevalence is probably higher. Twenty six,¹⁻¹⁸ including the present case, are reported in sufficient detail to enable meaningful comparison (Table 1). We wish

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Table 1 Comparison of treatments and outcome in transverse myelopathy associated with systemic lupus erythematosus

| Reference number | Age and sex | Diagnosis before onset of myelopathy | Time from onset of myelopathy to start of treatment | Drug treatment | Neurological outcome | Time from onset of myelopathy to neurological outcome |
|------------------|-------------|--------------------------------------|---|---|-------------------------|---|
| 1 | 24 F | Rheumatoid disease | 1 day | Decadron 8 mg/day | No change | 3½ months |
| 1 | 22 F | None | 1 day | Prednisolone 100 mg/day | No change | 3 months |
| 1 | 56 F | SLE* | 1 day | Prednisolone 60 mg/day | Death | 40 days |
| 2 | 24 F | SLE | Not known | Prednisolone 20 mg/day | No change | 9 years |
| 2 | 35 M | SLE | Not known | Prednisolone 20 mg/day | No change | 12 years |
| 3 | 47 F | SLE | Same day | Prednisolone 120 mg/day | Partial recovery | 4 months |
| 4 | 21 F | SLE | Same day | Prednisolone 20 mg/day, azathioprine 100 mg/day | Death | 1 day |
| 5 | 16 F | SLE | Same day | Prednisolone 100 mg/day | Partial recovery | 4 weeks |
| 6 | 27 F | None | 3 weeks | Prednisolone | No change | 5 months |
| 6 | 53 F | None | 3 weeks | Prednisolone | No change | 10 months |
| 7 | 35 F | SLE | None | None | Death | 4 days |
| 8 | 19 F | SLE | Receiving treatment before | Cortisone 75 mg/day | Death | 45 days |
| 9 | 40 M | None | Not known | Antibiotics | Recovery | — |
| 10 | 28 | SLE | Not known | Cortisone 200 mg/day | Recovery | 80 days |
| 11 | 26 M | SLE | Receiving treatment before | Steroids, ACTH* | Death | 3 days |
| 12 | 43 F | SLE | Same day | Cortisone 100 mg/day | Death | 1 year |
| 13 | 11 F | SLE | Receiving treatment before | Cortisone 50–100 mg/day | Death | 14 days |
| 13 | 12 F | Pyrexia of unknown origin | 4 days | Dexamethasone 24 mg/day | Partial recovery | 6 months |
| 13 | 35 F | None | 8 months | Prednisolone 80 mg/day | No change | 2 years |
| 14 | 33 M | Idiopathic thrombocytopenic purpura | 20 days 30 days | Steroids, cyclophosphamide | Death | 35 days |
| 15 | 41 M | Rheumatoid disease | Not known | Prednisolone | Death | 20 days |
| 16 | 51 F | SLE | Not known | Prednisolone | Death | 20 months |
| 17 | 23 F | SLE | Same day | Steroids-high dose | Recovery | 1 month |
| 18 | 29 F | None | — | None | Continued deterioration | — |
| 18 | 33 F | SLE | Not known | Not known | Continued deterioration | — |
| This case | 16 F | SLE | Same day | Prednisolone 80 mg/day, cyclophosphamide 1 g, plasma exchange | Partial recovery | 18 months |

*SLE=systemic lupus erythematosus; ACTH=corticotrophin.

to review two aspects of this condition: firstly, the effect of high dose steroid treatment and, secondly, cerebrospinal fluid glucose concentrations at the onset of myelopathy.

Of the 26 cases, seven recovered either full or partial neurological function, in nine the neurological deficit remained static or deteriorated further, and the remaining 10 patients died. For

eight patients treatment with high dose steroids was started at the onset of myelopathy, for seven of these within 24 hours and for the eighth within four days. Of these, five recovered either full or partial neurological function, one died, and in two the neurological deficit was unchanged. Eighteen patients were treated at the onset of myelopathy with either low dose steroids or a variety of drugs, or given no drug treatment at all. Of this group, two recovered, seven experienced no change in neurological deficit, and nine died.

Although it has been suggested that steroids are ineffective in transverse myelopathy due to systemic lupus erythematosus,² this review suggests that significant benefit may be gained from their early administration. Furthermore, the published work contains a further four cases where high dose steroids were administered with subsequent improvement in neurological function,²⁵⁻²⁷ but as insufficient detail was given in these reports they are not included in this review. We can only postulate the mode of action of high dose steroids in this condition. Vasculitis is a prominent feature in the spinal cord at postmortem examination,¹ which suggests that steroids may act by suppressing the vasculitic process. If this is the case then plasma exchange is a logical treatment as it may reduce

concentrations of circulating immune complexes and inflammatory mediators.

A review of the cerebrospinal fluid findings in patients with transverse myelopathy due to systemic lupus erythematosus shows a significantly reduced glucose concentration in many of the cases (Table 2), though it must be noted that simultaneous blood sugars were not measured. In fact of the 10 patients known to have undergone lumbar puncture on the day of onset of myelopathy, all had low cerebrospinal fluid glucose concentrations. Low cerebrospinal fluid glucose is not, however, a common feature of central nervous system lupus. Gibson and Myers, on reviewing the cerebrospinal fluid findings of 80 patients with central nervous system lupus, found only four instances, one of whom had a transverse myelopathy.²⁸ Duffy *et al* reviewing 21 cases of central nervous system lupus found one instance,³¹ and Feinglass found none in 44 cases.²⁴ Furthermore, low cerebrospinal fluid glucose concentrations are not noted to accompany idiopathic transverse myelopathy.³² As 10 of the 26 cases in this review were not known to have systemic lupus erythematosus at the onset of transverse myelopathy the finding of a low cerebrospinal fluid glucose in cases of transverse myelopathy arising afresh may have important diagnostic implications.

In conclusion, we suggest that the early administration of high dose steroids for transverse myelopathy due to systemic lupus erythematosus may decrease mortality and reduce the eventual neurological deficit in survivors.

Table 2 Summary of cerebrospinal fluid findings in transverse myelopathy associated with systemic lupus erythematosus

| Reference number | Time from onset of myelopathy to lumbar puncture | Glucose (mmol/l) | Protein (mg/l) | Number of cells ($\times 10^6/l$) |
|------------------|--|------------------|----------------|-------------------------------------|
| 1 | Same day | 0.8 | 1150 | — |
| 1 | Same day | 1.0 | 3250 | — |
| 1 | Same day | 1.4 | 1150 | — |
| 2 | 24 hours | Normal* | 1280 | 430 |
| 2 | Unknown | Normal | — | 1600 |
| 3 | Same day | 2.2 | 4050 | 940 |
| 4 | Same day | 1.2 | 1200 | 48 |
| 5 | Same day | 1.3 | 220 | 0 |
| 6 | Same day | 1.6 | 880 | 29 |
| 6 | Same day | 1.8 | 5850 | 3 |
| 7 | Unknown | — | 400 | 7 |
| 8 | Unknown | 0.9 | 1750 | 16 |
| 11 | Same day | 1.1 | 530 | 39 |
| 12 | Unknown | — | 400 | 7 |
| 13 | Unknown | 3.8 | 430 | 16 |
| 13 | Unknown | — | 3400 | — |
| 15 | Unknown | — | 1400 | 413 |
| 16 | Unknown | — | Normal* | 27 |
| 17 | 1 month | — | 1500 | 49 |
| This case | Same day | 2.0 | 1100 | — |

*Normal ranges: glucose 3.9-6.1 mmol/l; protein <600 mg/l.

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