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Lupus-Related Myelitis: Serial MR Findings

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PURPOSE: To correlate the MR findings in transverse myelitis secondary to systemic lupus erythematosus with clinical findings during disease exacerbation and remission. **METHODS:** Four patients (ages 33 to 47 years) with episodes of transverse myelitis secondary to systemic lupus erythematosus were identified. Three patients had recurrent transverse myelitis episodes (one patient with two recurrences), for a total of eight episodes. MR examinations (six after contrast administration) were performed during each transverse myelitis episode, as well as during four periods of remission (in three patients) after therapy with steroids and/or immunosuppressive agents. MR examinations were reviewed for the presence of spinal cord enlargement, intramedullary signal abnormality, and contrast enhancement. **RESULTS:** Prolongation of T1 or T2 signal (or both) was seen in eight episodes (100%). Spinal cord enlargement was seen in six (75%) of eight transverse myelitis episodes, although it was mild during two episodes. Contrast enhancement was seen in three of six transverse myelitis episodes (dense, inhomogeneous enhancement during two episodes in one patient, and a small focus of enhancement in one patient). During periods of remission, spinal cord diameter returned to normal, and no contrast enhancement was seen, although abnormal signal was present in three examinations performed within 2 months of a transverse myelitis episode. **CONCLUSION:** Spinal cord widening and signal abnormalities are common MR findings during episodes of transverse myelitis related to systemic lupus erythematosus, and contrast enhancement is less frequently seen. Improvement or resolution of these findings correlates with clinical improvement.

Index terms: Myelitis; Lupus erythematosus; Spinal cord, magnetic resonance

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Transverse myelitis, rapid onset of motor, sensory, and, usually, autonomic dysfunction at a spinal cord level, is an uncommon but well-recognized complication of systemic lupus erythematosus (1–4). Before the development of magnetic resonance (MR), the diagnosis of systemic lupus erythematosus-related transverse myelitis was one of exclusion, when computed tomographic (CT) myelography in a patient who had systemic lupus erythematosus with myelopathy failed to demonstrate another

cause of spinal cord dysfunction. There have been few reports of MR findings in systemic lupus erythematosus-related transverse myelitis (5, 6). The present study reports serial MR examinations in four patients and further establishes the role of MR in the diagnosis and treatment of this disease.

Materials and Methods

Four patients with a diagnosis of systemic lupus erythematosus (age range, 33 to 47 years, all women) were identified (Table). Two patients were identified by radiologic case material at the Massachusetts General Hospital, and the remainder were identified by survey of neuroradiologists at two other institutions. The diagnosis of systemic lupus erythematosus had been based on a history of arthralgias or myalgias (all four patients), nonerosive arthritis (four patients), malar rash (three patients), anemia or leukopenia (three patients), pleuritis or pericarditis (one patient), and the presence of antinuclear and anti-DNA antibodies (four patients) (Table). One patient was diagnosed with systemic lupus erythematosus only at the time of her first transverse myelitis episode. Evaluation to ex-

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Clinical features of systemic lupus erythematosus (SLE) in 4 women with transverse myelitis

Patient	Age, y	SLE Features	Episodes of Transverse Myelitis	Symptoms	Relapse
1	33	A,N,R,An,PI,PE,Ab	3	Paraparesis, T-3 sensory level	Quadriparesis, C-4 sensory level
2	47	A,N,R,Ab	2	Paraparesis, T-5 sensory level	Paraparesis, T-4 sensory level
3	40	A,N,An,S,NS,Ab	1	Bilateral arm and left leg weakness and paresthesias	None
4	46	A,N,L,Ab	2	Right arm, trunk and leg paresthesias	Paraparesis

Note.—A indicates arthralgias; N, nonerosive polyarthritis; R, malar rash; An, anemia; PI, pleuritis; PE, pericarditis; NS, nephrotic syndrome; S, splenomegaly; L, leukopenia; and Ab, antinuclear antibodies and anti-DNA antibodies.

clude multiple sclerosis as the cause of transverse myelitis included normal brain MR examinations (three patients), normal cerebrospinal fluid IgG, electrophoresis or absence of oligoclonal bands (three patients), and normal brain stem evoked potentials (one patient). Three patients had recurrent episodes of transverse myelitis (patient 1 had two recurrences), for a total of eight transverse myelitis episodes.

All patients had MR examinations performed during the early stages of both the initial and recurrent transverse myelitis episodes. Six MR examinations (in three patients) during transverse myelitis episodes were performed after contrast administration. At the time of contrast administration during transverse myelitis episodes, patients were receiving either no steroids or very low steroid doses.

Each episode was treated with high-dose steroids and, in two patients, immunosuppressive agents. Neurologic improvement was seen in each patient during the next few months. Two patients (during a total of three periods of remission) had repeat contrast-enhanced MR examinations within 2 months of the episodes, while undergoing steroid treatment. Another patient had a repeat noncontrast MR examination 4 years after the transverse myelitis episode. All MR examinations were retrospectively evaluated by two neuroradiologists in a nonblinded manner and correlated with the clinical history and neurologic examination findings. MR examinations in three patients were performed on a 1.5-T MR system, and the fourth patient was imaged on a 0.6-T system. Cardiac-gated T1-weighted sequences were obtained with parameters of 400–800/10–20 (repetition time/echo time). T2-weighted sequences were cardiac gated, 2250–2500/16–30, 80–96. All sagittal and axial images were obtained using either 3- or 4-mm noncontiguous scans with a 256 × 192 matrix. Gradient-echo images were obtained using 28–45/15, a flip angle of 6° to 10° degrees, and either a 256 × 192 or 256 × 256 matrix.

Results

MR abnormalities corresponding to the spinal level of clinical involvement were demon-

strated during all eight episodes. T1 and T2 prolongation within the involved region was present during each episode and was more prominent on T2-weighted sequences in all cases. In general, the abnormal signal was homogeneous throughout the entire length of the involved spinal cord. The rostrocaudal extent of the signal abnormality varied between episodes but was generally about four vertebral bodies in length. Spinal cord widening was seen during six episodes, with return to normal size during the period of remission. Contrast enhancement was seen in three of six MR examinations (in two patients) performed during transverse myelitis episodes. In each of the three instances of spinal cord enhancement, the site of enhancement was in a region of abnormal signal on noncontrast images. In two episodes (both in patient 2), almost the entire region of abnormal signal enhanced in a diffuse, inhomogeneous manner (Figs 1A and B). In patient 1, a small portion of the abnormal region enhanced with contrast (Fig 2E).

No contrast enhancement was seen in any of the three episodes of remission during which contrast-enhanced MR imaging was performed. There was resolution of spinal cord enlargement in all cases (Fig 3C). Small regions of residual abnormal signal intensity, however, were present on either noncontrast T1- or T2-weighted images in all cases (Fig 2D). Follow-up MR examination performed 4 years after the initial examination in patient 4 demonstrated atrophy of the previously involved spinal cord segment, with no residual signal abnormality.

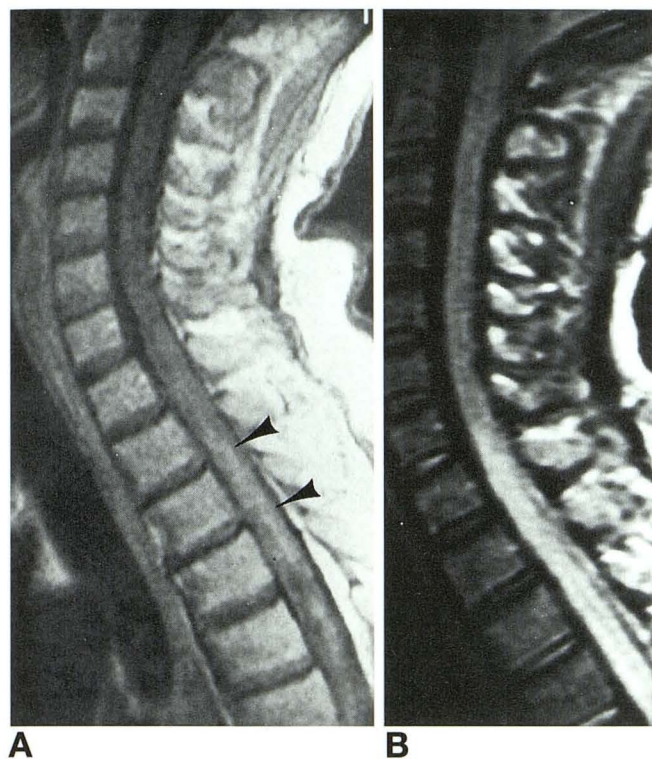


Fig 1. Patient 2, 47-year-old woman with documented systemic lupus erythematosus and a 2-day history of paraparesis and leg paresthesias.

A, Contrast-enhanced T1-weighted (400/22/2 excitations) sagittal image demonstrates diffuse, inhomogeneous enhancement of the upper thoracic spinal cord (arrowheads).

B, Proton-density (2000/60/1) sagittal image shows diffuse hyperintense signal throughout the upper thoracic spinal cord.

Discussion

Central nervous system manifestations of systemic lupus erythematosus are found in 20% to 50% of patients with systemic lupus erythematosus (1, 7). Neuropsychiatric symptoms are particularly common, but other neurologic features can include seizures, cranial neuropathy, hemiparesis or paraparesis, and peripheral nervous system involvement, such as peripheral neuropathy and myopathy (1). Transverse myelitis is one of the least frequent central nervous system complications. The initial clinical features usually include back pain, paraparesis or quadriplegia, and sensory loss caudad to the level of the lesion. A midthoracic or low-thoracic sensory level is usually present, reflecting the most common sites of spinal cord involvement (8). Onset is usually within a few years of the diagnosis of systemic lupus erythematosus (9) but may be delayed many years (10). Transverse myelitis as the first manifestation of systemic lupus erythematosus, seen in one of

our patients, is uncommon (2, 3, 11), as are recurrent transverse myelitis episodes (12). Three of our patients, however, had recurrent episodes, suggesting that transverse myelitis recurrence may be more common than previously reported.

Before the advent of MR, CT myelography was the principle means of neuroradiologic evaluation of patients with transverse myelitis of any cause (13). CT myelography, however, is neither sensitive nor specific for the diagnosis of transverse myelitis. Positive findings in transverse myelitis on CT myelography are limited to the finding of spinal cord widening, reported in only 20% of transverse myelitis cases (13). The lack of sensitivity of this finding is underscored by the fact that spinal cord widening was mild or absent during four of eight transverse myelitis episodes. Furthermore, spinal cord widening is a nonspecific finding on CT myelography, because a neoplasm or infarct could also produce this finding.

MR demonstration of spinal cord widening and prolongation of T1 or T2 signal has been previously described in patients with transverse myelitis in case reports (13–16). Based on these reports and our findings, T1 and T2 signal prolongation seems to be a common finding in transverse myelitis generally. It was the most sensitive finding in this series. In our patients, spinal cord widening was a less-sensitive indicator of spinal cord involvement, being either absent or less severe than the degree of signal abnormality in four transverse myelitis episodes.

Lack of contrast enhancement does exclude disease activity, because it was absent during three of six transverse myelitis episodes in which contrast-enhanced examinations were performed. During one episode (patient 1), the region of contrast enhancement was only a small portion of the area of signal abnormality on noncontrast images (Fig 2E). In patient 2, although a large degree of contrast enhancement was present (Fig 1A), the area of contrast enhancement was less extensive than the region of abnormal signal on T2-weighted images (Fig 1B). The paucity of contrast enhancement in patients 1 and 3 cannot be attributed to steroid treatment, because they were not receiving steroids or were receiving only low doses of steroids when MR imaging was performed. Follow-up contrast-enhanced MR examinations during periods of steroid treatment and remis-

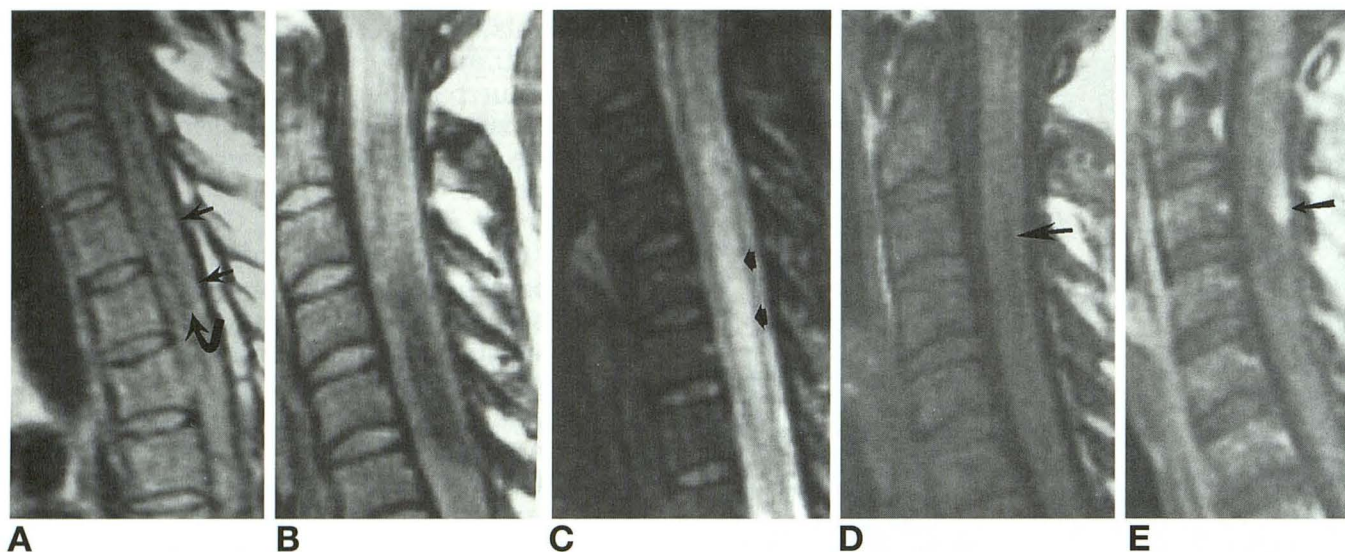


Fig 2. Patient 1, 33-year-old woman diagnosed 4 years earlier with systemic lupus erythematosus, with a 2-month history of progressive paraparesis.

A, Noncontrast T1-weighted (400/20/2) sagittal image demonstrates widening of the upper thoracic spinal cord (arrows) with abnormal central hypointense signal (curved arrow). She was treated with steroids, with relatively good recovery of motor strength.

B, Noncontrast T1-weighted (600/25/2) sagittal image performed 11 months after that in A, after steroid taper and new onset of quadriplegia. There is widening of the cervical spinal cord with central regions of hypointense signal.

C, T2-weighted (2000/80/1) sagittal image demonstrates hyperintense signal (arrows) within the spinal cord.

D, Noncontrast T1-weighted (400/11/2) sagittal image taken 1 month after that in C, during a period of remission after steroid therapy. The spinal cord diameter is now normal, although a central region of hypointense signal (arrow) remains.

E, Contrast-enhanced T1-weighted (400/11/2) sagittal image taken 6 months after that in D. The symptoms had continued to diminish during steroid treatment but again worsened during steroid taper, prompting this MR examination. A hypointense region is present in the spinal cord, which partially enhances with contrast material (arrow).

sion within 2 months of the transverse myelitis episode demonstrated that a marked decrease in both the signal abnormality and spinal cord widening accompanies clinical improvement, although residual regions of abnormal signal can still be seen (Fig 2D). The absence of contrast enhancement during the periods of clinical improvement may reflect disease remission or be secondary to the stabilizing effect of high doses of steroids.

In general, the appearance of transverse myelitis on any single MR examination in this series was indistinguishable from an intramedullary tumor. Lack of contrast enhancement has been proposed as a feature that is helpful in distinguishing transverse myelitis from a neoplasm (16), but as the findings in our second patient illustrate, transverse myelitis can, indeed, enhance with contrast material. Serial MR examinations were important in making the distinction and provided a measure of specificity not available with a single MR examination. Transverse myelitis can be distinguished from an in-

tramedullary tumor by a rapid and prolonged response to steroids (15). Serial MR examinations provided objective evidence of a treatment response of a degree and duration greater than that expected with a spinal cord neoplasm. The lack of ionizing radiation or need for introduction of intrathecal contrast agents made serial MR examinations possible at low risk to the patient.

Multiple sclerosis is another cause of transverse myelitis from which systemic lupus erythematosus must be distinguished. Multiple sclerosis was excluded or considered highly unlikely in each of our patients on the basis of a normal brain MR examination, normal cerebrospinal fluid examination, or normal brain stem evoked potential study. In patients with an established diagnosis of multiple sclerosis or systemic lupus erythematosus, transverse myelitis can usually be presumed to be caused by the known underlying disease. However, difficulty may arise when there is no preexisting diagnosis of either disease. Because lesion enhance-

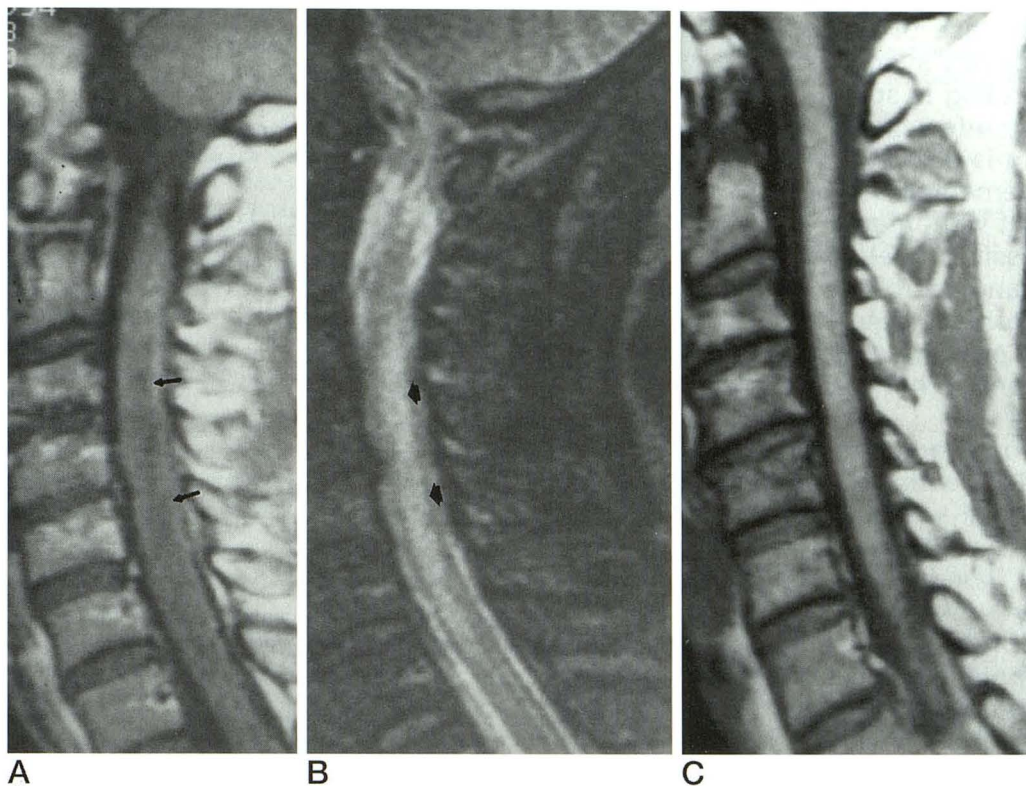


Fig 3. Patient 3, 40-year-old woman with known history of systemic lupus erythematosus and a 2-month history of bilateral arm and left-leg weakness and paresthesias.

A, Noncontrast T1-weighted (600/20/4) sagittal image demonstrates diffuse widening of the cervical spinal cord with hypointense signal abnormality (*arrows*). There was no enhancement on images obtained after contrast administration (not shown).

B, T2-weighted (2340/80/1) sagittal image demonstrates hyperintense signal (*arrows*) throughout the cervical spinal cord.

C, Contrast-enhanced T1-weighted (600/20/4) sagittal image performed 1 month after beginning steroid treatment. The spinal cord diameter is now normal. No abnormal signal or contrast enhancement are seen. There was a small region of residual abnormal signal on T2-weighted images (not shown).

ment can be seen in both diseases, contrast enhancement is not a useful distinguishing feature. The presence of asymptomatic white matter lesions in the brain can provide supportive evidence for the diagnosis of multiple sclerosis (17), but these can also be seen in systemic lupus erythematosus (18) and are, therefore, not helpful in determining the cause of a spinal cord lesion. The MR findings that are most helpful are the lesion number and rostrocaudal extent. Although two different spinal cord levels were involved in one of our patients at different times (patient 1), multiple coexistent lesions were not seen in any of our patients, or noted in previous reports (5, 6). Therefore, the finding of multiple simultaneous lesions should favor the diagnosis of multiple sclerosis. The rostrocaudal extent of lesions in this series of patients was, on average, four to five vertebral bodies in length, much longer than typical spinal cord multiple sclerosis plaques (17, 19, 20).

The cause of the MR signal abnormalities in systemic lupus erythematosus-related transverse myelitis is not known with certainty. Three main pathologic findings have been reported. The most common finding is vacuolar degeneration of the peripheral spinal cord white matter, with relative sparing of gray matter (8). Patchy areas of axonal degeneration and ballooning of myelin sheaths are seen at many spinal cord levels. Spinal cord T1 and T2 prolongation may, therefore, be caused by intravacuolar water. Possible causes of this vacuolar degeneration include an autoimmune mechanism (21) or ischemia (22). Similar vacuolar changes have been noted in patients with acquired immunodeficiency syndrome, and are most prominent in patients with have acquired immunodeficiency syndrome who have severe myelopathy (23). Furthermore, spinal cord expansion and hyperintense signal on T2-weighted images similar to those seen in our patients have been

reported in a case of myelopathy in acquired immunodeficiency syndrome (15). The second pathologic finding is spinal cord infarction, reported in only a few cases (24–26). The cause of this finding is also not fully understood. The rapid improvement in our patients in this series is inconsistent with infarction. The third finding is a compressive myelopathy with regions of hemorrhage and necrosis caused by spinal subdural hematoma, described in two cases (27, 28) and presumed to be caused by a systemic lupus erythematosus-related coagulopathy (29).

Systemic lupus erythematosus-related transverse myelitis is commonly thought to be secondary to a small-vessel vasculitis (26), but this is probably true in only a minority of cases (8, 30). We found a few reports of vascular infiltration by lymphocytes and other mononuclear cells (25, 26), as well as scattered reports of acute fibrinoid necrosis of small or large vessels with severe intimal thickening (22, 24). More commonly, however, there is specific mention of the absence of vasculitis and fibrinoid necrosis (1, 8, 10, 27, 30). Spinal cord vasculitis, therefore, seems to be the exception, rather than the rule, in systemic lupus erythematosus-related transverse myelitis.

Treatment of systemic lupus erythematosus-related transverse myelitis usually consists of high-dose corticosteroid therapy within the first few days after symptom onset (4, 31). Other immunosuppressive agents, such as cyclophosphamide, have also been advocated (32). Clinical improvement, which was accurately reflected by MR imaging, was seen in all our patients after treatment. The clinical course is, however, in general quite variable. Incomplete recovery, significant permanent neurologic disability, or death may result, even in treated cases.

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