Case Report

# Myelopathy: A rare presentation of initial systemic lupus erythematous manifestation in a young male

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## ABSTRACT

Acute transverse myelitis (ATM) is a consequence of inflammation of various etiologies resulting in damage to the nerve tracts. Regardless of the etiology, the prevalence of ATM is estimated at 1–4 new cases per million a year. Among connective tissue disorder, systemic lupus erythematous (SLE) is the most common cause of ATM. SLE is complicated by neurological manifestation in 25–80% of the patients. ATM may be a complication in 1–3% of SLE patients, but in some patients, it may be the first manifestation of SLE. We describe the case of a 20-year-old male where longitudinal extensive transverse myelitis was the initial manifestation of SLE involving almost the whole length of the spine. Early diagnosis and aggressive treatment may prevent long-term permanent damage and may have a favorable outcome.

**Key words:** Acute transverse myelitis, American College of Rheumatology, Systemic lupus erythematous

Acute transverse myelitis (ATM), a consequence of inflammation of various etiologies including systemic connective tissue disease, multiple sclerosis, sarcoidosis, neuromyelitis optica, infectious disease (bacteria, viral, and parasitic), and paraneoplastic syndrome resulting in damage to the nerve tracts passing through the spinal cord causing muscle paresis, sensory deficit, and sphincter dysfunction. According to the American Academy of Neurology, the involvement of greater than 3 vertebral segments is classified as longitudinal myelitis. Regardless of the etiology, the prevalence of ATM is estimated at 1–4 new cases per million a year [1]. It has been estimated that in about 10% of ATM cases, etiology remains undetermined [2]. In these cases, the disease is referred to as idiopathic ATM. Among connective tissue disorder, systemic lupus erythematous (SLE) is the most common cause of ATM [3]. SLE is an autoimmune disorder that potentially affects any organ system. In SLE, the neurological manifestation is seen in 25–80% of patients [4,5].

Myelopathy is one of the less common neuropsychiatric manifestations of SLE which occurs only in 1–3% of SLE patients [6-8]. In the majority of cases, myelopathy occurs shortly after SLE is diagnosed usually within the first 5 years of age. In about half of the patients, ATM is the first clinical manifestation of SLE [6,8]. These groups of patients may not fulfill the American College of Rheumatology (ACR) criteria for the diagnosis of SLE which may delay the diagnosis and may affect the outcome [9]. We present one such case where ATM was the initial and early manifestation of SLE involving almost the whole length of the spine.

| Access this article online | |
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| Received - 26 May 2020<br>Initial Review - 10 June 2020<br>Accepted - 06 November 2020 | Quick Response code |
| **DOI:** 10.32677/IJCR.2020.v06.i12.010 | |

## CASE REPORT

A 20-year-old male presented with complaints of tingling sensation in both lower limbs for 3 days that progress to the weakness of all four limbs, reduced sensations below the neck with urinary retention, and constipation. The patient was febrile (100.5°F) for 2 days before the onset of weakness. There was no history of trauma, recent vaccination, tuberculosis, rash, joint pain, oral ulcer, or any other clinical signs and symptoms suggestive of SLE.

On admission, he was afebrile with normal vitals. Cardiovascular system and rheumatic system examinations were unremarkable. Per abdomen examination showed a distended abdomen as a result of constipation and urinary retention. Neurological examination suggested normal cranial nerve examination, power in the lower limb was 0/5 and in the upper limb, it was 3/5 with areflexia and sensory loss below the neck. He was diagnosed with longitudinally extensive transverse myelitis (TM) and was started with IV 1 g methylprednisolone for 5 days followed by oral prednisolone in tapering dose.

Blood reports showed normal complete blood counts and biochemistry, negative C-reactive protein, rheumatoid factor,

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normal thyroid function test, negative serology, normal urine examination, elevated erythrocyte sedimentation rate-26 at the end of 1 h, and elevated serum creatinine phosphokinase-myocardial band-51.2 (2 times of upper normal limit). His chest X-ray and ultrasound abdomen were normal. His electrocardiogram showed sinus bradycardia with a heart rate of 50/min. His 2D echo showed mild mitral valve prolapse. His cerebrospinal fluid (CSF) examination showed clear fluid, no cellular element on microscopy, CSF protein was 20 mg/dl, CSF glucose was 56 mg/dl, and Pandy's test was negative. His fundus was normal. Magnetic resonance imaging (MRI) spine showed a hyperintense signal on T2-weighted image from C2 to conus (Fig. 1) suggestive of acute longitudinal TM. MRI brain showed T2 fluid-attenuated inversion recovery bright lesion in the left frontal and right gangliocapsular parenchyma suggestive of inflammatory edema. A further autoantibodies screen revealed positive anti-double-stranded deoxyribonuclease (anti-dsDNA) and antimitochondrial antibodies.

Based on the clinical picture, radiology, and immunological marker, he was diagnosed with SLE-related acute longitudinal TM which is an extremely rare condition. Only a few cases have been reported throughout the world.

## DISCUSSION

SLE is an autoimmune disease affecting various organ systems. It is complicated by neurological symptoms in 25–80% of the cases [4,5]. In 1999, the ACR defined 19 neuropsychiatric syndromes that may occur in the course of SLE, collectively known as neuropsychiatry systemic lupus erythematous syndrome [10]. The most common neuropsychiatric forms include headache, cerebrovascular accident, mood change, cognitive disorder, and convulsion [4]. ATM is seen in 1–3% of SLE patients [6-8]. Most patients who develop ATM do so within 5 years of SLE diagnosis.

ATM can be the first manifestation of SLE. In a study conducted by Kovacs *et al*., they reported ATM as a presenting feature in 39% of the total patient population [6]. ATM in SLE may present with a classical picture of muscular weakness, sensory deficit, and sphincter dysfunction. Our patient presented with all the clinical symptoms of the TM but without clinical signs and symptoms suggestive of SLE. In those cases, where ATM is the presenting feature of SLE, many patients may not fulfill the ACR criteria for the diagnosis of SLE but over the course of the disease, they may eventually develop other signs and symptoms of SLE [9]. In our patient, although the anti-dsDNA antibody was positive, he has not shown any other clinical features suggestive of SLE. It is likely that the clinical course has been altered with ongoing immunosuppressive therapy [9].

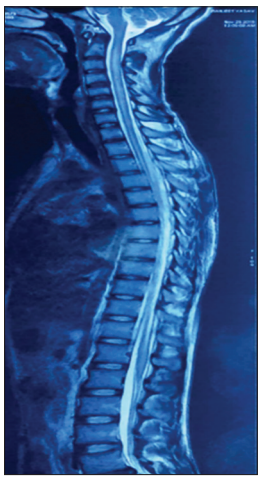

**Figure 1: Magnetic resonance imaging spine sagittal section T2-weighted image showing hyperintensity from C2 to conus**

The sensitivity of antinuclear antibody (ANA) in SLE is 95% [11]. Anti-dsDNA antibodies are found in 70% of SLE patients at some point during the course of the disease and are 95% specific [11]. The ANA-negative lupus is usually associated with the presence of other cytoplasmic autoantibodies [11]. In our patient, ANA was negative, anti-dsDNA and antimitochondrial antibodies were positive suggestive of SLE but no clinical feature to correlate. The prevalence of antiphospholipid antibody (aPL) is higher in SLE patients with ATM compared to SLE patients in general. Lavalle *et al*. reported 10 out of 11 SLE patients with ATM as having aPL antibody [12]. Kovacs *et al*. reported 64% of SLE patients with ATM as having aPL [6]. This might be important for the etiology of TM because the spinal cord necrosis secondary to arterial thrombosis might be a pathological factor. The early use of anticoagulants in these patients might be important for the outcome of the disease.

MRI is very useful as it shows the extent of involvement and also helps to rule out the other causes of myelopathies. It usually shows a high signal in T2WI. The involvement may be limited to one particular segment or involve a long length of the spinal cord. Our patient had almost complete involvement of the spinal cord from C2 to conus (Fig. 1). There have been reports that in those greater than four spinal segments are involved, a higher proportion of them have varying degrees of disability after treatment. They also may show a greater degree of inflammation in CSF and sensory involvement is more frequent.

There are no precise recommendations for standard treatment for ATM in SLE because of its very low prevalence. At present, the use of glucocorticoids and cyclophosphamide is standard therapy [3]. Sole infusion of glucocorticoids was effective in some cases of ATM. A repeated cycle of immunosuppression is accepted in therapy-resistant cases [13]. The effectiveness of plasmapheresis is not clear although it has been used in some patients [6]. Successful treatment with rituximab and autologous bone marrow transplantation also has been used in refractory cases [14,15]. In our patient, we gave IV 1 g methylprednisolone infusion for 5 days then oral prednisolone in tapering dose.

The time taken for the complete to partial improvement in neurological function may be variable, ranging from several days and many months [2,16]. Our patient showed complete sensory recovery and significant improvement in motor function on follow-ups. In general, one-third of the patients with TM recover completely, one-third recover partially but one-third left with a severe disability. Those patients with hyperacute symptoms at

onset, positive ANA, or those caused by connective tissue have poor outcome [16,17].

## CONCLUSION

Acute longitudinal myelitis can be a devastating condition as it remits one's quality of life. Rarely, longitudinal myelitis can be the first presentation of SLE. It may occur irrespective of the lupus activity. Therefore, when a patient presented with unexplained neurological symptoms and longitudinal myelitis on MRI, SLE should be considered as part of differential even though there may not be only clinical signs/symptoms suggestive of SLE. Early detection and aggressive treatment with steroid and cyclophosphamide can prevent long-term permanent damage and may have a favorable outcome. This case report emphasizes the need for multicentric trials for SLE patients with ATM to help study the optimal management option.

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*Funding: None; Conflicts of Interest: None Stated.*

**How to cite this article:** Gupta R, Warkade D, Singh S. Myelopathy: A rare presentation of initial systemic lupus erythematous manifestation in a young male. Indian J Case Reports. 2020;6(12):703-705.