CASE REPORT

# Acute Longitudinal Myelitis As The First Presentation in Child with Systemic Lupus Erythematosus Concurrent with Positive Antiphospholipid Antibody

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**ABSTRACT**
Systemic lupus erythematosus (SLE) is a multi-system auto-immune disorder that is characterized by widespread immune deregulation, formation of auto–antibodies, and immune complexes, resulting in inflammation and potential damage to variety of organs. 25-95% it is complicated by neurological or neuropsychiatric symptoms, which is referred to as neuropsychiatric SLE (NPSLE). NPSLE contain both central and peripheral nervous systems, which includes transverse myelitis. We report our experience of concurrent manifestation of transverse myelitis as an initial presentation of SLE, which suggests the common immune-mediated mechanisms of diseases. We here report the case of a 7-year-old girl with SLE who first presented with features of TM. The patient developed ascending weakness starting from low extremities, experienced difficulty in voiding. An initial diagnosis of TM was made on the basis of clinical findings and MRI spine, which displayed T2 weighted high signal intensities at thoracic level. She partially respond to intravenous immunoglobulin therapy, and serological analysis revealed the presence of anti-dsDNA, anti nuclear antibody with decreased level of complements. The diagnosis was revised to acute transverse myelitis resulting from SLE. Additional methylprednisolone pulse therapy led to rapid clinical improvement. This was followed by oral prednisolone and cyclophosphamide pulse therapy. The cross-reactivity of auto-antibodies and increased susceptibility to infection owing to immunologic changes associated with lupus may form the basis of the association. Systemic Lupus Erytheromyitis should consider as an etiology of transverse myelitis. Aggressive treatment may alter the course and lead to a better outcome.

**Keywords:** Auto-immune, SLE, Transverse myelitis

## INTRODUCTION

Systemic lupus erythematosus (SLE) is a rare connective disease affecting 6-19 cases per 100, 000 children. The neurological manifestations are seen in 25-95% of patients with SLE more commonly in the form of headache, psychosis, or cognitive dysfunction.[1,2] In up to 1-2% of patients with SLE it may be complicated by transverse myelitis, but rarely may be the initial manifestation of SLE. We present one such case where ATM was the initial and only manifestation of SLE.

## CASE REPORT

A 7-year-old Saudi girl, previously well, presented with history of pain in the left leg for 10 day weeks progressing to bilateral weakness of legs and sensory loss. She was febrile for 2 days prior to admission. She had constipation and urinary retention. There was no history of trauma, recent vaccination, cough, skin rash, joints pain, oral ulcers, or any other clinical symptoms or signs suggestive of SLE.

On admission to hospital she was afebrile with normal vital observations and blood pressure. Examination of her cardiovascular and respiratory system was unremarkable. Abdominal examination revealed distended abdomen as a result of constipation and urinary retention. Neurological examination suggested normal cranial nerve examination with no bulbar palsy. The motor power in the lower limb at presentation was 3/5 MRC with areflexia. The motor power was 5/5 MRC in the upper limbs with brisk tendon reflexes. Her weakness progress with in 2 days and sensory level, MRI T2 spine showed increased signal at T3 and below. she was diagnosed with Transveres Myelitis started on intravenous-pulsed therapy followed by oral prednisone in tapering dose. She was partially improved. Three days later we notice

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left facial palsy and asymmetric tone with brisk reflex left side upper limb that lower limb, MRI brain support focal stroke at right internal capsule. Diagnosis corrected to Transveres Myelitis and focal stroke, extra dose of methylpredisolone was given, and oral prednisolone in tapering doses continued and intravenous heparin.

Blood results at presentation showed normal biochemistry but elevated ESR of 35 mg/L (normal range 0-10 mg/L). The complete blood count was normal except for low white blood count of 3.4/L and lymphocytes of 0.67 × 109/L. MRI spine (Figure 1) showed multi-focal multi-regional transverse myelitis involving spinal cord from T2 down to the T10 (Figure 1). MRI brain show T2 increase signal in right internal capsule and right restriction diffusion (Figure 2). Extensive investigations were carried to identify the underlying cause. Cerebrospinal fluid (CSF) showed white cell count (WCC) of 1120 × 106/L with predominant polymorphs and elevated protein of 0.67 g/L. Oligloclonal band Negative, immunoglobulin IgG index 26.2, myelin basic protein 1.2 n/L(0.0-2.2 n/L).

All the cultures including blood, CSF, and urine were reported as no growth. The virology screen was negative including Lyme's serology. Anti nuclear antibodies (ANA) was positive with the titres being and show 1:2560. At this point pediatric rheumatology opinion was sought to rule out an auto-immune condition or a connective tissue disease leading to transverse myelitis and stroke. Rheumatology evaluation did not reveal any other signs suggestive of SLE or any other connective tissue disease. She did not satisfy the Americal College of Rheumatology (ACR) criteria for diagnosis of SLE.

Further study was done Anti dsDNA 28.7 u/ml (normal range 0.0-20 u/ml), and Anti B2– gly 1 19.9 (range 0-10 u/ml). C3 and C4 were low. Immunoglobulin profile showed elevated IgG and IgM levels. Anti Cardiolipin antibody and lupus anticoagulant were negative. Aquaporin IgG antibody was negative. Echocardiogram normal study.

The lab markers were suggestive of SLE but with no convincing clinical features to correlate. She was transferred to local pediatric rheumatology unit. Based on the overall clinical picture and immunology markers, she was commenced on IV cyclophosphamide which was continued for seven cycles. After completion of IV cyclophosphamide cycles, she was commenced on azathioprine, oral prednisolone 10 mg once daily along with hydroxychloroquine. Initially, she needed s/c heparin therapy which was changed to Aspirin once she started mobilizing.

She had neuro rehabilitation in the form of intensive physiotherapy, support from the occupational therapy. Over the following few months she made significant recovery. Her repeat MRI of spine (Figure 3) done 12 months later has shown complete resolution of the inflammatory changes. She was able to walk with very minimal hemiparesis. She has normal bladder but has abnormal bowel movements use intermittent laxative. She has normal motor power in the upper limbs and she has normal sensations both in upper and lower limbs. She has re-integrated back at her

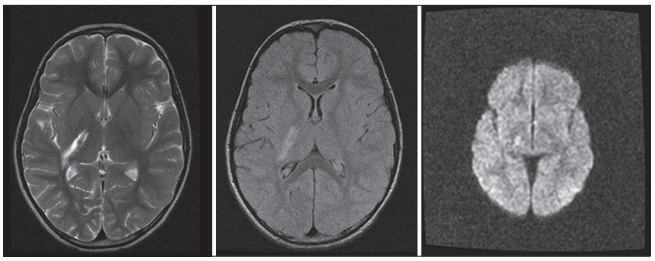

**Figure 2:** MRI brain T2 (left) showed increase signal at lower part of internal capsule. Same finding at T1 MRI brain (right). At the bottom DWI showes restrection diffusion same area suggest focal stroke

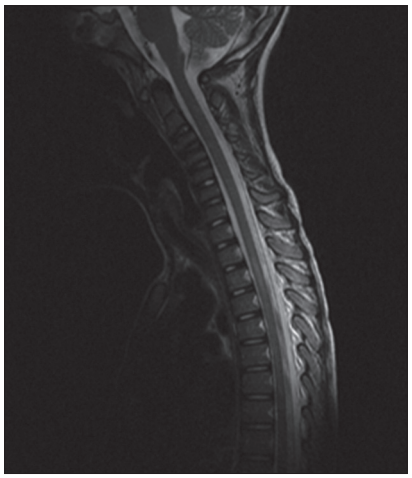

**Figure 1:** T2-weighted sagittal image of the crevice-thoracic spine demonstrating enlargement of the spinal cord below T3 with high signal within it

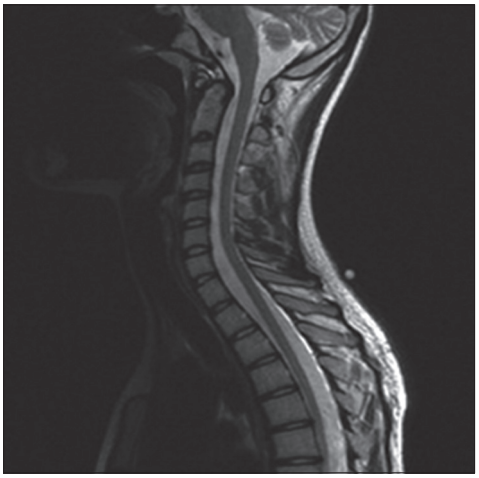

**Figure 3:** Normal MRI spine

normal mainstream school. After 18 months of treatment with immunosuppressants and oral steroids, she has not developed any other clinical features of SLE.

## DISCUSSION

SLE is an auto-immune disease affecting various organ systems. It is complicated by neurological symptoms in 25-95% of the cases.[1,2] The common symptoms are headache, seizures, or psychosis. ATM is seen in 1-2% of SLE patients, but in one adult study it is reported to be a presenting feature in up to 39% of SLE patients.[1,3,4] Most patients who develop complications of ATM do so within 5 years of SLE diagnosis. ATM in SLE may present with the classical picture of motor weakness, sensory disturbance, and sphincter disturbances. Our patient presented with all the classical symptoms of transverse myelitis but with no clinical signs or symptoms suggestive of SLE. The diagnosis of SLE in our patient was made based on positive immunological markers, lymphopenia, and unusual clinical presentation on of longitudinal myelitis. The pathogenesis of ATM in SLE is unclear but most likely the process is immune complex mediated vasculitis or thrombosis leading to ischemic spinal lesions or anti-phospholipid antibodies cross-reacting with spinal cord phospholipids.[1,5]

In those cases where ATM is the presenting feature of SLE, many may not fulfil the criteria for the diagnosis of SLE but over the course of the disease they may eventually present with other signs and symptoms of SLE.[3] In our reported case although the ANA and double-stranded DNA antibodies are positive, the patient has not shown any other clinical features of SLE. It is likely that the clinical course has been altered with on-going immunosuppression. Investigation may reveal raised inflammatory markers in blood as well as CSF.[3,4,6,7] The auto-antibody screen may show antibodies positive suggesting SLE but with no clinical features to correlate. MRI is very useful as it shows the extent of involvement and also helps rule out other causes of myelopathies. It usually shows high signal in T2-weighted images.

A majority of them are positive for anti-phospholipid syndrome, although it is not very clear whether those with APLs do worse in terms of long term recovery.[2,4,8] Transverse myelitis with spastic paraparesis and sensory loss at a given level is a rare but severe complication of SLE or antiphospholipid antibody syndrome. Stroke and transient ischemic attack (TIA) may be related to antiphospholipid antibody syndrome or SLE vasculitis.

Episodes of cerebral ischaemia, mainly focal, can be transient or permanent. Recurrent disease often leads to multifocal deficits. Amaurosis fugax,[9] transient paraesthesias, motor weakness, vertigo and transient global ischaemia[10] can all be expressions of TIAs.

Patients with the highest IgG anticardiolipin titers had the shortest times to subsequent thrombo-occlusive events. The most common recurrent event was cerebral infarction, often occurring within the first year of follow-up during a mean prospective follow-up of 3 years.[11]

Our patient presented with neurological signs suggestive of acute longitudinal myelitis and transient ischemic stroke secondary to positive anti-phospholipid but no clinical markers which would suggest SLE. She fulfilled the ACR criteria for 4 out of 11 criteria but immunological markers were highly suggestive of SLE. CSF showed non-specific inflammatory changes with raised cell count and protein. She was diagnosed early and treated aggressively which may have helped the outcome. Recurrence risk of stroke as the Anticardiolipin was negative.

The involvement may be limited to one particular segment or involve a long length of the spinal cord. Our patient had mainly thoracic involvement of spinal cord from T3 to T10. There have been reports that in those where more than four segments are involved, a high proportion of them have varying degree of disability after treatment. They also may show greater degree of inflammation in CSF and sensory involvement is more frequent.[5,9] In our patient showed good recovery following aggressive treatment. MRI scan done 16 months after presentation showed complete resolution of T2 changes in the spinal cord and brain.

The general approach to treatment includes[4,12] steroids in the form of IV PULSE methylprednisolone and immunosuppressants such as cyclophosphamide. This combination of treatment seems to show better response. But the prognosis is generally perceived to be poor in those with SLE. The role of plasmapharesis is not clear although it has been used in some patients.[5]

In general, in those patients with transverse myelitis, 1/3$^{rd}$ recover completely, 1/3$^{rd}$ recover partially but 1/3$^{rd}$ are left with severe disabilities. Those with hyper acute symptoms at onset, positive ANA, or those caused by connective disease have poor outcome.[4,5]

## CONCLUSION

Acute longitudinal myelitis can limits one's quality of life. Rarely ATM with sensory loss at a given level is a rare. SLE should be consider as a cause of TM. Early detection and aggressive treatment can prevent long-

term permanent damage and may even have a favorable outcome.

## REFERENCES


1. Avcin T, Benseler SM, Tyrrell PN, Cucnik S, Silverman ED. A followup study of antiphospholipid antibodies and associated neuropsychiatric manifestations in 137 children with systemic lupus erythematosus. Arthritis Rheum. 2008;59:206–13. [PubMed].
2. Silverman E, Eddy A. Systemic lupus erythematosus. In: Cassidy JT, Petty RE, Laxer RM, Lindsley CB, editors. Textbook of Pediatric Rheumatology. 6th ed. Philadelphia: Saunders Elsevier; 2011: 328–30.
3. D'Cruz DP, Mellor-Pita S, Joven B, Sanna G, Allanson J, Taylor J, et al. Transverse myelitis as the first manifestation of systemic lupus erythematosus or lupus-like disease: Good functional outcome and relevance of antiphospholipid antibodies. J Rheumatol. 2004;31:280–5. [PubMed].
4. Vieira JP, Ortet O, Barata D, Abranches M, Gomes JM. Lupus myelopathy in a child. Pediatr Neurol. 2002;27:303–6. [PubMed].
5. al-Mayouf SM, Bahabri S. Spinal cord involvement in pediatric systemic lupus erythematosus: Case report and literature review. Clin Exp Rheumatol. 1999;17:505–8. [PubMed].
6. Espinosa G, Mendizábal A, Mínguez S, Ramo-Tello C, Capellades J, Olivé A, et al. Transverse myelitis affecting more than 4 spinal segments associated with systemic lupus erythematosus: Clinical, immunological, and radiological characteristics of 22 patients. Semin Arthritis Rheum. 2010;39:246–56. [PubMed].
7. Lopez Dupla M, Khamashta MA, Sanchez AD, Ingles FP, Uriol PL, Aguado AG. Transverse myelitis as a first manifestation of systemic lupus erythematosus: A case report. Lupus. 1995;4:239–42. [PubMed].
8. Linssen WH, Fiselier TJ, Gabreëls FJ, Wevers RA, Cuppen MP, Rotteveel JJ. Acute transverse myelopathy as the initial manifestation of probable systemic lupus erythematosus in a child. Neuropediatrics. 1988;19:212–5.
9. Asherson RA, Khamashta MA, Gil A et al. Cerebrovascular disease and antiphospholipid antibodies in systemic lupus erythematosus, lupus-like disease, and the primary antiphospholipid syndrome. Am J Med 1989;86:391–9.
10. Kovacs B, Lafferty TL, Brent LH, DeHoratius RJ. Transverse myelopathy in systemic lupus erythematosus: An analysis of 14 cases and review of the literature. Ann Rheum Dis. 2000;59:120–4.
11. Levine SR, Brey RL, Sawaya KL Recurrent strok and thrombo-occlusive events in antiphospholipid syndrome. Ann Neurol. 1995;38(1):119-24.
12. Chen HC, Lai JH, Juan CJ, Kuo SY, Chen CH, Chang DM. Longitudinal myelitis as an initial manifestation of systemic lupus erythematosus. Am J Med Sci. 2004;327:105–8.


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