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Case Report

The unseen onset: myelitis and nephritis as the first clue to juvenile lupus

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ABSTRACT

Juvenile systemic lupus erythematosus (SLE) is a multisystem autoimmune disorder that commonly affects the kidneys and central nervous system. While lupus nephritis and neuropsychiatric manifestations are well-recognized complications, the concurrent onset of lupus nephritis and acute transverse myelitis (ATM) as the initial presentation is exceptionally rare, particularly in the pediatric population. Such an atypical presentation poses significant diagnostic challenges and requires a high index of clinical suspicion to enable early recognition and timely management.

Keywords: Unseen onset, Myelitis, Nephritis, Juvenile lupus

INTRODUCTION

Among pediatric autoimmune diseases, juvenile systemic lupus erythematosus (SLE) stands out for its diverse and often aggressive clinical course, which frequently includes renal and neurological involvement. Compared to adult-onset disease, childhood SLE is associated with earlier organ damage and increased morbidity. The majority of children with SLE are adolescent females.[1,2] Lupus nephritis (LN) is one of the most common and serious complications, affecting approximately 50–80% of children with SLE and often presenting early in the disease course, sometimes even as the first clinical feature.[3] Neurological involvement is also frequently observed in SLE, with a reported prevalence of 25–95%, and affects 13–45% of pediatric patients, commonly manifesting as headache, seizures, psychosis, or cognitive dysfunction.[4] Among the rarer but potentially devastating neurological complications is acute transverse myelitis (ATM), also known as lupus myelitis - an inflammatory disorder of the spinal cord leading to motor, sensory, and autonomic deficits. ATM is estimated to occur in approximately 0.7% of all SLE patients, with only 1–2% of those cases occurring in the pediatric population.[5,6] Lupus myelitis typically appears years after the diagnosis of SLE, but in exceptional cases, it may be the initial manifestation. It often presents as longitudinally extensive transverse myelitis (LETM), where spinal cord involvement spans three or more vertebral segments on magnetic resonance imaging (MRI).[7]

The simultaneous presentation of lupus nephritis and ATM at disease onset in juvenile SLE is extremely rare and has only been reported in isolated case reports, with no clearly defined incidence due to its rarity. Most affected patients are adolescent females, although cases have been described in children as young as five years.[4,5]

We report a case of a 14-year-old girl presenting with concurrent onset of lupus nephritis and LETM, emphasizing the need for a high index of suspicion for systemic autoimmune diseases in children presenting with unexplained renal and neurological symptoms. Early recognition and aggressive immunosuppressive treatment are crucial to improving outcomes in such rare presentations.

CASE REPORT

A 14-year-old girl presented with a two-month history of constitutional symptoms, followed by a three-day history of acute-onset, progressive paraparesis involving the lower

limbs and was associated with sensory deficits in the lower extremities. As the motor weakness worsened and sensory deficits persisted, she was referred to a tertiary care center for further evaluation.

On examination, the patient was alert with normal cranial nerve function and no evidence of bulbar palsy or facial weakness. Vital signs were stable except for elevated blood pressure (150/100 mmHg). Neurological examination revealed paraplegia involving both proximal and distal muscle groups, with exaggerated deep tendon reflexes and extensor plantar responses bilaterally. Sensory examination revealed impaired pain and touch perception below the T6 level, along with bowel and bladder disturbances.

Initial laboratory evaluation showed bicytopenia (anemia and leukopenia), severe hypokalemia (serum potassium 2.0 mmol/l), hypoalbuminemia. Despite appropriate correction of hypokalemia, the motor weakness persisted. MRI of the brain and spine revealed longitudinally extensive T2 hyperintensities spanning T5 to T10 suggestive of acute transverse myelitis (ATM) (Figure 1). Cerebrospinal fluid (CSF) analysis was unremarkable, with normal protein and no pleocytosis or evidence of infection.

Given the persistent neurological deficits despite correction of metabolic abnormalities and absence of infectious or traumatic etiology, an autoimmune cause was suspected. Further evaluation revealed a positive antinuclear antibody (ANA) test (+2 intensity, titer 1:100, nuclear homogeneous pattern), strongly positive anti-dsDNA antibodies (ELISA), and low complement levels (C3 - 57 mg/dl and C4 - 4 mg/dl). Urine studies showed sub-nephrotic range proteinuria (urine protein/creatinine ratio 1.4, 24-hour urinary protein 2.78 g/day) and hypertension, raising suspicion for lupus nephritis. Renal biopsy confirmed ISN/RPS class IV lupus nephritis. Antiphospholipid antibody (APLA) panel was negative.

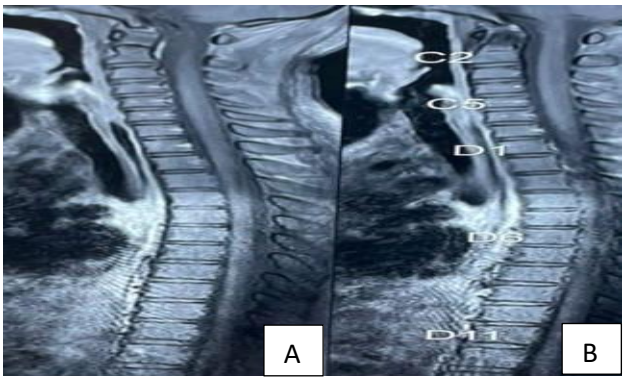

Figure 1 (A and B): T2 hyperintensities suggestive of LETM.

Based on clinical, serological, and histopathological findings, a diagnosis of juvenile SLE with concomitant transverse myelitis and class IV lupus nephritis was established. The patient was initiated on intravenous pulse methylprednisolone, followed by oral prednisolone and intravenous cyclophosphamide as induction therapy. She was subsequently transitioned to maintenance therapy with mycophenolate mofetil. Over follow-up for 6 -12 months, the patient demonstrated complete remission of nephritis and significant neurological improvement, with progressive recovery of motor function.

DISCUSSION

The clinical heterogeneity of SLE often complicates early diagnosis, particularly in pediatric patients where classical features may be absent. When rare manifestations such as transverse myelitis coincide with lupus nephritis at presentation, the diagnostic challenge is even greater. Juvenile-onset SLE (jSLE), tends to have a more aggressive course than adult-onset SLE, with higher frequencies of major organ involvement such as lupus nephritis (LN) and neuropsychiatric lupus.[8]

Lupus nephritis is one of the most frequent and severe complications in jSLE, affecting 50–80% of children, and can be the first manifestation in many cases.[9,10] Class IV lupus nephritis, as seen in our patient, represents a diffuse proliferative form that requires early diagnosis and aggressive immunosuppression due to its strong association with poor renal outcomes.[11]

ATM in SLE is characterized by acute or subacute onset of motor weakness, sensory disturbances, and autonomic dysfunction, and its presentation may mimic other causes of myelopathy, making diagnosis challenging. While ATM often occurs within the first 5 years of SLE diagnosis, in rare instances such as our case it may be the initial presenting feature, even in the absence of other classic clinical signs of lupus.[13]

The pathogenesis of ATM in SLE is complex and incompletely understood. Proposed mechanisms include immune complex-mediated vasculitis, cytokine-driven neuroinflammation, and thrombotic events secondary to antiphospholipid antibodies (APLA).[14,15] In our patient, APL antibodies were negative, making a vasculitic or inflammatory etiology more likely. Notably, in some cases where ATM precedes other SLE manifestations, patients may not initially fulfill classification criteria for SLE but may do so over time. MRI is the cornerstone for evaluating myelitis. T2-weighted hyperintensities, indicating demyelination or inflammatory edema, are typically observed, particularly in LETM. The involvement of more than four spinal segments has been associated with greater sensory involvement, CSF inflammation, and worse long-term outcomes. However, in our case, CSF analysis was unremarkable.

The concurrent occurrence of LETM and class IV LN at presentation in jSLE is extremely rare, with only isolated pediatric cases reported in the literature. This overlap phenotype represents a severe disease variant and warrants

urgent immunosuppressive intervention. Prognosis in SLE-associated ATM is variable: approximately one-third of patients recover completely, one-third partially, and the remaining may suffer significant disability.[14] Poor prognostic indicators include hyper acute onset, positive APLA, delayed treatment, and poor early response to therapy. Early diagnosis and prompt initiation of high-dose corticosteroids, followed by cyclophosphamide or mycophenolate, are critical in controlling both renal and neurological inflammation.[16] The absence of APLA, early diagnosis, and timely therapy likely contributed to the favourable outcome in our case.

This case highlights the importance of maintaining a high index of suspicion for systemic autoimmune disease in children presenting with unexplained multi systemic symptoms, especially in the absence of typical features. Moreover, it underscores the need for long-term follow-up to monitor disease activity, prevent relapses, and ensure adherence to maintenance immunosuppression.

CONCLUSION

Unusual multi systemic involvement in young patients, should raise suspicion for systemic autoimmune rheumatic diseases (AIRD), as they often present with atypical features and pose a diagnostic challenge, leading to delayed treatment and increased morbidity and mortality. This case highlights the importance of early recognition, prompt diagnosis, and a high index of suspicion for SLE, even in the absence of classic manifestations especially in pediatric age group to initiate prompt therapy and thereby preventing long term morbidity and mortality.

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