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Juvenile Systemic Lupus Erythematosus Presenting as Diffuse Lymphadenopathy and Mononeuritis Multiplex

Dr. L. Ratna Neelima[1], Dr. Veena Narisetty[2], Dr. G. Sree Ranga Lakshmi[3], Dr. Akhilesh Kumar[4], Dr. A. Goutami Priyadarshini[5], Dr. Ramya Lavu[6]

[1]Department of Neurology, Osmania Medical College, Hyderabad
Corresponding Author Email: *neelimaratna[at]yahoo.in*,

[2]Department of Neurology, Osmania Medical College, Hyderabad
Email: *veenanarisetty[at]yahoo.com*,

[3]Department of Neurology, Osmania Medical College, Hyderabad
Email: *rangalakshmi2000[at]gmail.com*,

[4]Department of Neurology, Osmania Medical College, Hyderabad
Email: *mailyourdoc[at]gmail.com*,

[5]Department of Neurology, Osmania Medical College, Hyderabad
Email: *docgoutami[at]gmail.com*,

[6]Department of Neurology, Osmania Medical College, Hyderabad
Email: *ramyalavu[at]gmail.com*

Abstract: ***Background: Systemic lupus erythematosus (SLE) is an autoimmune disorder characterized by multisystem involvement with a wide range of clinical manifestations. Lymphadenopathy may be seen in children with SLE but is only occasionally reported as a presenting feature. Neuropsychiatric manifestations occur in a substantial proportion of pediatric patients[3], with isolated involvement of the peripheral nervous system being distinctly rare[4]. Case Presentation: We report a 14-year-old boy with generalized lymphadenopathy of one year's duration who had been extensively investigated without diagnostic clarification, and who presented to the neurology department with bilateral ulnar neuropathy. Despite an initial suspicion of lymphoma, an autoimmune panel sent as part of the workup for mononeuritis multiplex established a diagnosis of juvenile SLE. Discussion: This case highlights the need to consider SLE in the differential diagnosis of generalized lymphadenopathy, alongside infections and malignancies, and illustrates mononeuritis multiplex as a rare presenting feature of the disease[5-7]. Conclusion: Early recognition of atypical presentations of SLE- including lymphadenopathy and mononeuritis multiplex- allows timely immunosuppressive treatment and avoids delays caused by pursuing a malignancy workup alone.***

Keywords: Juvenile SLE, generalized lymphadenopathy, mononeuritis multiplex, neuropsychiatric SLE

1. Introduction

SLE is an autoimmune disorder with manifestations that can involve virtually any organ system [1]. Lymphadenopathy is a frequent and nonspecific feature of SLE, and patients with lymphadenopathy have been found to have higher disease activity[8]. Neuropsychiatric SLE (NPSLE) encompasses a spectrum of neurological and psychiatric signs and symptoms that are often difficult to distinguish from events unrelated to SLE[2]. Headache, mood disorders, anxiety, and mild cognitive dysfunction are the most frequent neuropsychiatric complaints in patients with SLE, and neuropsychiatric involvement has been reported in a substantial proportion of pediatric patients[3]. Guillain-Barré syndrome, autonomic dysfunction, polyneuropathy, and mononeuritis multiplex are recognized peripheral nervous system manifestations of SLE[4], though mononeuritis multiplex as a presenting feature of SLE is only rarely reported[5-7].

2. Case Report

A 14-year-old boy, born of second-degree consanguineous parentage, presented with a one-year history of recurrent febrile episodes, occurring intermittently over weeks to months with intervening asymptomatic periods. He had lymph nodal swelling in multiple non-contiguous areas involving the cervical, axillary, and inguinal regions. He gave a two-month history of painless bluish discoloration of the fingertips on cold exposure, and presented to us with paresthesias and weakness involving the bilateral medial two fingers.

On examination, the patient was conscious and coherent, afebrile, with a pulse rate of 86/min and blood pressure of 110/80 mmHg, with no murmurs or added breath sounds. Abdominal examination revealed

hepatosplenomegaly. He had generalized lymphadenopathy, with nodes that were soft and mobile, and a maculopapular rash over the thenar eminence of the right hand.

CNS examination showed normal higher mental functions and cranial nerves, with bilateral weakness of the ulnar-innervated muscles, decreased sensation over the medial one-and-a-half fingers bilaterally, and bilateral thickening of the ulnar nerves. Deep tendon reflexes were preserved, with bilateral flexor plantar responses.

In view of persistent fever associated with lymphadenopathy and mononeuritis multiplex, lymphoma was considered a strong possibility, and a workup for other causes was conducted in parallel. Investigations revealed hemoglobin 9.6 g/dL, ESR 73 mm/hr, and a negative CRP. Nerve conduction studies showed bilateral ulnar sensorimotor axonopathy, and ultrasound of the bilateral ulnar nerves was normal. The Mantoux test measured 5 mm, and chest radiography was normal. Axillary lymph node biopsy showed reactive follicular hyperplasia without atypical cells. PET scan demonstrated metabolically active lymphadenopathy in multiple stations on both sides of the diaphragm, with diffuse splenic involvement.

As no evidence of malignancy was found, ANA testing was performed. ANA blot was negative, but ANA by immunofluorescence was positive with a mixed pattern (homogeneous 4+, cytoplasmic 2+). Anti-dsDNA was positive, with C3 22 mg/dL (normal 90–180 mg/dL) and C4 2.7 mg/dL (normal 10–40 mg/dL); p-ANCA and c-ANCA were negative. Following rheumatology consultation, a diagnosis of juvenile SLE was made, with a Systemic Lupus Erythematosus Disease Activity Index (SLEDAI) score of 13. The patient was started on low-dose oral prednisolone, hydroxychloroquine, and mycophenolate mofetil for immunosuppression.

On follow-up, the patient showed steady improvement in ulnar motor and sensory function, with resolution of paresthesias and progressive recovery of hand-grip strength over subsequent visits. Prednisolone was gradually tapered according to clinical and serological response, while hydroxychloroquine and mycophenolate mofetil were continued as maintenance immunosuppressive therapy. The patient remains on regular follow-up, with periodic monitoring of disease activity (SLEDAI), complement levels, anti-dsDNA titres, and renal function, in keeping with standard long-term management of childhood-onset SLE.

3. Discussion

Lupus lymphadenopathy has an estimated prevalence of 5–7% at disease onset and 12–15% at any stage of the disease[8,9]. Lupus patients with lymphadenopathy carry an increased risk of coexisting lymphoma, and because the clinical presentation can closely mimic malignancy, diagnosis and treatment of the neoplasm may be delayed; complete differentiation ultimately depends on immunohistochemistry[8]. Kikuchi-Fujimoto disease (KFD), or histiocytic necrotizing lymphadenitis, occurs in young people and commonly involves the cervical lymph nodes; it should also be considered and can usually be distinguished on histology[9].

The differential diagnosis of mononeuropathy multiplex includes vasculitis, autoimmune disorders, infectious diseases, sarcoidosis, amyloidosis, cryoglobulinemia, and paraneoplastic disease[10]. Recognizing mononeuropathy multiplex may play a crucial role in the prompt diagnosis of SLE and the timely initiation of treatment, as mononeuritis multiplex in SLE is uncommon and, when it occurs as a presenting feature, is particularly liable to diagnostic delay[5-7]. Diagnosis of SLE requires a positive ANA together with clinical and immunological features fulfilling the classification criteria[1]. Early diagnosis allows prompt initiation of immunosuppression once infections and malignancies, which commonly present with lymphadenopathy, have been excluded.

4. Conclusion

Lymphadenopathy in SLE is commonly seen in young patients, often accompanied by cutaneous and constitutional symptoms, and generally responds well to corticosteroids. Early diagnosis can be difficult,

since autoantibodies and low complement levels may not yet be evident on initial evaluation, and lymphadenopathy can precede the diagnosis of SLE by years, before the development of other clinical and laboratory abnormalities. Evaluating for autoimmune disease in patients presenting with generalized lymphadenopathy therefore facilitates early treatment and helps prevent complications.

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