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Case Report

Neuropsychiatric systemic lupus erythematosus: a rare case of acute polyneuropathy

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ABSTRACT

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease which presents most commonly in middle aged females and affects multiple organ systems. Amongst the many systems involved, the nervous system generally gets affected later in the course of the disease. We report a case of a male patient who developed quadriparesis as the initial presentation of SLE who progressed to lupus nephritis. The patient was started on methylprednisolone, and later planned on IVIg when he did not respond to the initial treatment. Unfortunately, the patient developed diaphragmatic paralysis and succumbed to the illness.

Keywords: Systemic lupus erythematosus, Quadriparesis, Guillain-Barré syndrome

INTRODUCTION

Neuropsychiatric systemic lupus erythematosus (SLE), as described by American College of Rheumatology (ACR) includes 19 neuropsychiatric syndromes divided into neurologic syndromes of central, peripheral and autonomic nervous system.[1] Peripheral nervous system involvement occurs in 3-18% of patients.[2] Guillain-Barré syndrome (GBS) and its other variants are a part of the 19 syndromes, although are reported only rarely in the course of the disease. The universally accepted treatment of GBS is IVIg ± plasmapheresis which is unlike SLE flares which are treated with high dose intravenous corticosteroids. We report a case of a middle-aged man presenting initially with acute polyneuropathy and progressing to lupus nephritis and pancytopenia.

CASE REPORT

A 36-year-old male, with a history of bronchial asthma, presented to the emergency department with progressive oedema of the lower limbs. The onset of oedema was associated with a gradually progressing weakness of the lower limbs, which in due course also involved the upper limbs. He also complained of a tingling sensation of all four limbs for sixteen days before presentation. He gave no history of skin rashes, mouth ulcers, hair loss, photosensitivity or arthralgia.

On physical examination, vitals were stable and oxygen saturation was ninety seven percent on room air. Bilateral lower extremity pitting oedema was present. Motor examination revealed 5/5 strength in the upper extremities and 3/5 strength in the lower extremities. Reflexes were diminished in all four limbs.

Initial laboratory investigations are given in Table 1 which revealed microcytic anaemia and a slightly decreased platelet count. Kidney function test was suggestive of pre-renal acute kidney injury. Serum electrolytes were suggestive of hyperkalemia, hypocalcemia and mild hyponatremia. Liver function test was within normal limits except low serum protein and low serum albumin. Urine routine examination revealed ++ proteinuria and microscopic hematuria.

Hemogram gradually progressed to a picture of pancytopenia. Serum albumin progressively decreased to

a nadir of 1.61 g/dl. Ascitic fluid analysis was suggestive of low SAAG and low protein, following which a 24-hour urine protein was done which was measured as 1,961 mg in 24 hours. Magnetic resonance imaging (MRI) brain and cervical spine showed no abnormalities. Nerve conduction studies were inconclusive due to anasarca. SLE workup (Table 2) was done due to appearance of pancytopenia with worsening renal function and presence of sub-nephrotic proteinuria which turned out to be positive.

Table 1: Initial laboratory investigations.

Lab parameter	Value
Hemoglobin	10.9 g/dl
Mean corpuscular volume	10.5 fL
Mean corpuscular hemoglobin	26.7 pg
Red cell distribution width	18.3%
Platelet count	120,000 cells/mm^3
Serum urea	127.9 mg/dl
Serum sodium	133.6 mmol/l
Serum potassium	6.86 mmol/l
Serum ionic calcium	3.96 mg/dl
Serum protein	4.7 g/dl
Serum albumin	2.27 g/dl
Urine examination routine-protein	++ 100 mg/dl
Urine examination routine-RBC	5/hpf

Table 2: SLE workup.

Lab parameter	Value
ANA titres	1:100, 4+, coarse speckled
ANTI dsDNA	152 IU/ml (+ve >30 IU/ml)
C3 complement	24.2 mg/dl (normal- 90-180 mg/dl)
C4 complement	7.2 mg/dl (normal- 10-40 mg /dl)
Anti-Smith antibody	+ve (++)
Anti ribosomal P	+ve (+++)
Anti Ro	+ve (+++)

Treatment and outcome

Patient fulfilled ACR SLE criteria and was initiated on pulse methylprednisolone at a dose of 1 gm daily for three consecutive days which was later extended to five days.[3] The patient showed a very mild reduction of oedema with no improvement in power, which over time had progressed to 0/5 in all 4 limbs.

Patient was planned for IVIg therapy at a dose of 0.4 g/kg/day for five days but unfortunately, the patient developed diaphragmatic involvement and went into type 2 respiratory failure. The patient was intubated but could not be salvaged.

DISCUSSION

SLE is a chronic autoimmune disease that most commonly affects young women of reproductive age. It presents with constitutive symptoms, rashes, photosensitivity, oral ulcers, kidney and nervous system involvement. In the aggregate, studies report that approximately one-third to one-half of SLE patients report neurologic or neuropsychiatric symptomatology.[4]

Neuropsychiatric SLE refers to the pathological involvement of central nervous system (CNS) by the disease process. Involvement of the CNS due to infections or due to immunosuppression, or neuropsychiatric symptoms secondary to side effects of drugs is considered as secondary neuropsychiatric SLE. The frequency of peripheral neuropathy in patients with SLE is variably reported but is typically less than 10 to 15 percent.[5] In many cases, the peripheral neuropathy is attributed to a cause other than SLE.[4]

AIDP, also called Guillain-Barré syndrome, and the axonal variant are both described in patients with SLE (and included as 1 of the 19 phenotypic syndromes of neuropsychiatric SLE (NPSLE) by the ACR, but are rare.[6] In one meta-analysis, the prevalence was <0.1 percent, and only two cases of acute inflammatory neuropathy were reported in the international SLICC cohort of 1206 patients.[4,7] In other series, AIDP accounted for only 1.1 percent of peripheral nervous system manifestations in SLE.[8]

SLE flares are usually managed with high dose intravenous steroids however, AIDP and AMAN/AMSAN rarely respond to them. Since a lot of cases of GBS presenting as a manifestation of SLE have not been reported, there is no unified guideline for its treatment. Some were treated with plasmapheresis and steroids with full recovery or steroids, plasmapheresis, and cyclophosphamide with partial recovery or had full recovery to only cyclophosphamide and steroids after failed initial treatment with IVIG and plasmapheresis or had full recovery with cyclophosphamide and steroids after failed treatment with IVIG.[9-12]

CONCLUSION

Guillain-Barré syndrome is a rare but possible presentation of neuropsychiatric SLE. Current treatments for GBS in SLE are not driven by universal guidelines and patient response to medications are very variable.

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