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**Nephrology**

# Acute Flaccid Quadriparesis as an Uncommon Initial Presentation of SLE: A Case Report and Literature Review

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**Abstract** **Case Report**

Systemic lupus erythematosus (SLE) is an autoimmune, multisystem disorder that has a high morbidity and mortality rate involving millions of people worldwide. It has a diverse presentation due to its ability to affect almost all body organs. Here we present a case of a 22-year-old female who presented to us with acute flaccid quadriparesis. Subsequently, there was 6th cranial nerve palsy, proximal muscle weakness, joint stiffness, and proteinuria. Laboratory investigations show positive ANA, Anti-ds-DNA, and Anti Sm Ab. She was diagnosed as a case of SLE and was treated with IV Methylprednisolone. After getting the treatment her symptoms were improved.

**Keywords**: Autoimmune, Multisystem, Flaccid Quadriparesis, Cranial Nerve Palsy, Autoantibodies.



## INTRODUCTION

SLE was first described by the great physician Hippocrates (460–375 bc) as cutaneous ulcers under the heading of herpes esthiomenos. Since then it has been considered a skin disease. Sir William Osler first stated that SLE can involve other organs even without skin manifestation [1]. It is an autoimmune, inflammatory disorder that most commonly affects females of reproductive age. The main pathology is the dysfunction of the immune system. There is production of autoantibodies due to loss of B and T cell tolerance. Antibodies are mainly formed against nuclear antigens resulting in widespread inflammation and ultimately tissue injury [2]. The morbidity and mortality of SLE patients are two to three times higher than the normal population depending upon the organ involvement [3]. The reported incidence and prevalence of SLE differ significantly by geography. Age, gender, and ethnicity are the main factors in determining the clinical outcome and management. Though this disease is more prevalent in the female population, its course is more critical and devastating in men [4]. Though SLE is an incurable disease early diagnosis and proper management can delay the progression of diseases, and major organ damage. The common presentations of SLE are skin manifestation, oral ulcer, musculoskeletal involvement, and serositis. But sometimes it may have a bizarre presentation. A high level of suspicion is necessary regarding SLE if a female of reproductive age presents with multisystem involvement. In this report, we present a case with acute onset flaccid quadriparesis along with cranial nerve palsy subsequently musculoskeletal manifestation, and renal involvement as the early presentation of SLE.

## CASE HISTORY

A 22-year-old female presented to us with complaints of weakness in all four limbs for one month. Initially, she felt numbness and tingling sensation in her upper limbs, gradually spreading to her lower limbs. Two to three days later she noticed heaviness in her lower limbs with difficulties walking. There was decreased sensation in the affected limbs. She also had problems standing from sitting posture without the help of others. Fifteen days later she developed double vision mostly on the right side on the lateral gaze. During this course of

the disease, she gradually developed joint stiffness, especially in the morning mainly involving small joints of her hands which persisted for about 1-2 hours. She also had pain in the elbow joint and shoulder joint which was asymmetrical in distribution and inflammatory in nature. The patient also complained of extreme fatigue and myalgia. Seven days before admission she noticed decreased urine output though the urine color was normal without any dysuria or abdominal pain and gradual periorbital puffiness with bilateral leg swelling. She had no recent history of diarrhea, respiratory tract infection, vaccination, surgery, and trauma. Her father is a known case of Psoriatic arthritis. Clinical examination revealed she was mildly anemic and edematous, her BP was 120/70 mmHg, and her pulse was 90b/min. A nervous system exam showed right 6th cranial nerve palsy, muscle tone reduced, muscle power 3/5 in lower limbs, 4/5 in the upper limbs, jerks are absent in both upper and lower limbs, plantar flexor (bilateral), coordination was impaired due to muscle weakness. A sensory exam revealed a decrease in fine touch and pain sensation. Position and vibration sense were intact. The Gower sign was positive. Other Systemic examinations revealed normal findings.

**Investigations showed**

| | |
|---|---|
| CBC | Hb 10.6 gm/dl<br>TWBC: 3200<br>Plt :250000<br>ESR :50mm in 1st hour |
| Urine RE | Protein :4+<br>Pus cell : 12-15/HPF<br>RBC: nil<br>Cast : nil |
| S. Creatinine | 0.5 mg/dl |
| UTP | 13gm/day |
| S albumin | 2.4gm /dl |
| CRP | 1.6mg/dl |
| ANA | Positive |
| Anti dsDNA | Positive |
| Anti Sm Ab | Positive |
| Anti uRNP | Positive |
| CPK | 51.0U/L |
| C3,C4 | Normal |
| RA, Anti CCP | Negative |
| APS panel | Negative |
| liver function test | Normal |
| S vitamin B12, folic acid | Normal |
| USG of whole abdomen | Right kidney 11.3 cm; left kidney 11.4 cm<br>Cortical echogenicity–normal, CMD well maintained |
| chest Xray PA view was | Normal |
| CSF study | Albuminocytological dissociation |
| MRI of the brain with screening of the spinal cord | No abnormalities |
| NCS of all four limbs. | Prolonged distal latency in the right median nerve and left ulnar nerve. Reduced Camp amplitude only in the right ulnar nerve. F response was absent in both ulnar nerves and prolonged in all other nerves |
| Renal Biopsy | Light microscopy: focal mesangial proliferation, endocapillary hypercellularity, wire loop lesion (Figure 1)<br>DIF: Immune deposit of IgG(1+), IgA(1+), C3(1+), Kappa(1+), Lambda(1+), C1q and IgM trace<br>Light microspic and DIF findings were suggestive of class **III** of lupus nephritis |

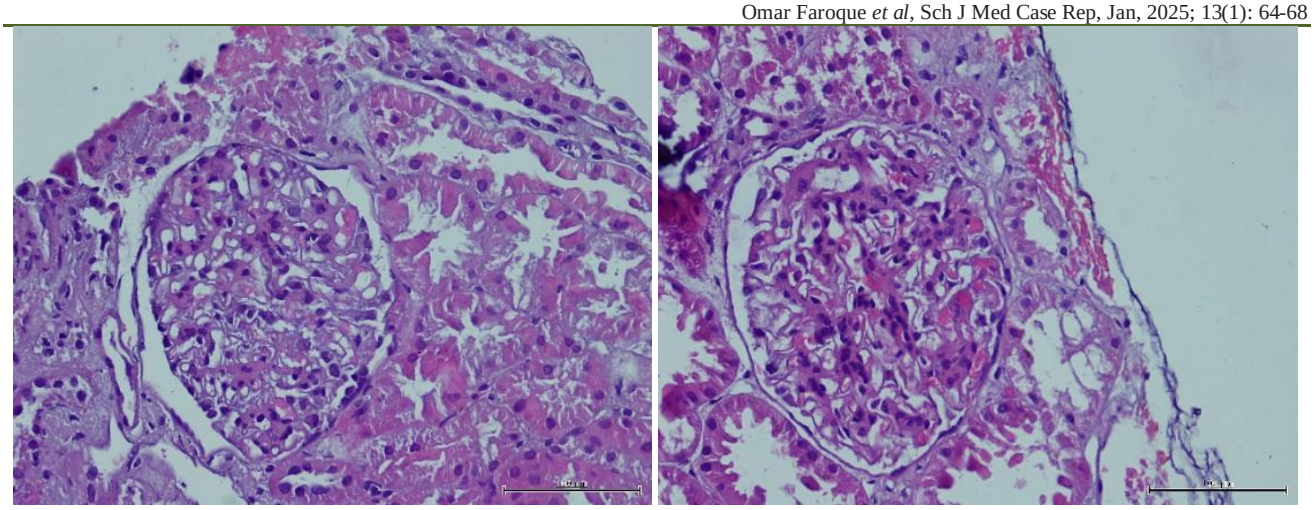
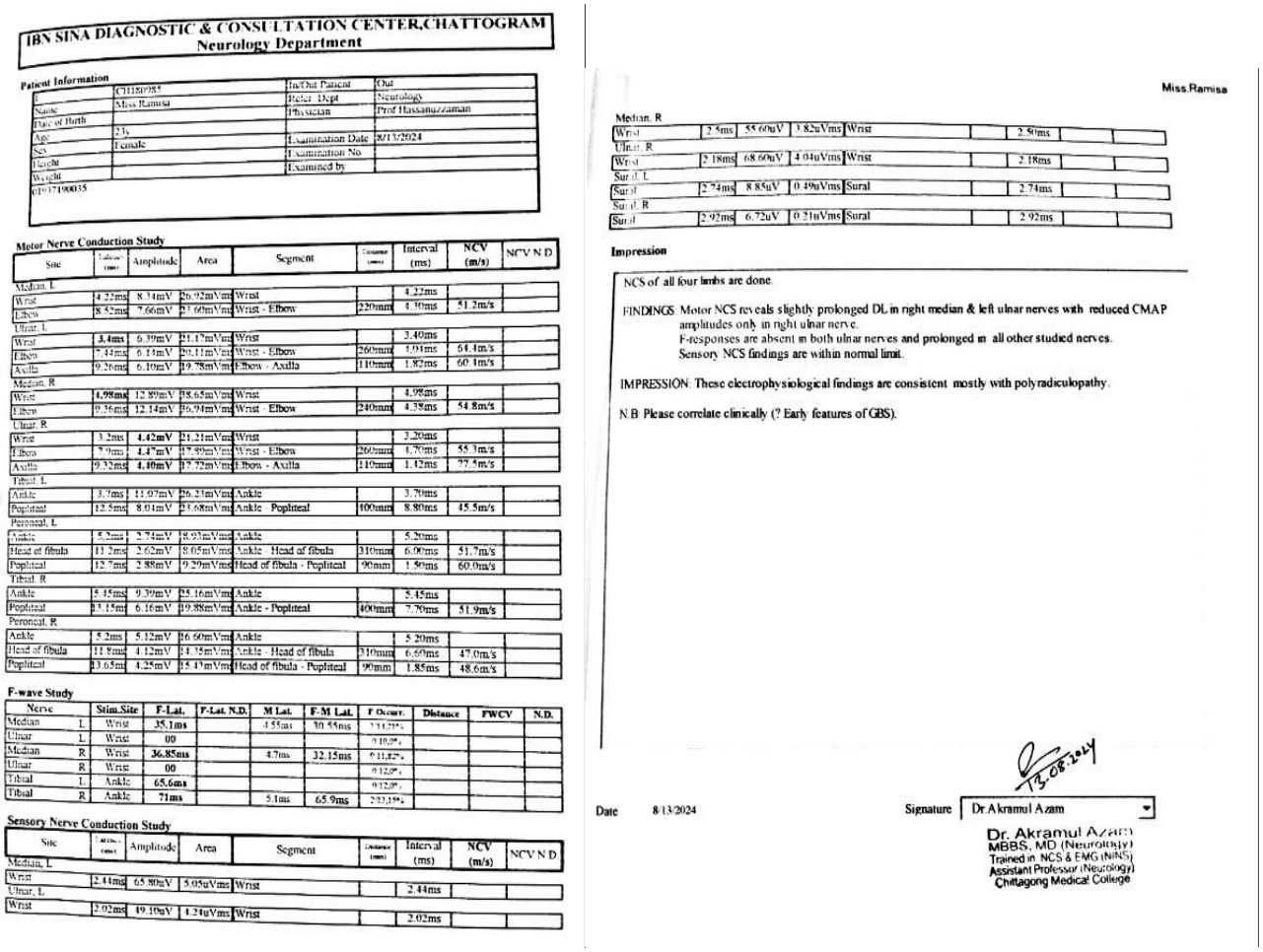


**Figure 1: Mesangial proliferative changes and wire loop lesion (right) in renal biopsy**



IBN SINA DIAGNOSTIC & CONSULTATION CENTER,CHATTOGRAM
Neurology Department

Patient Information

| | | | |
|---|---|---|---|
| ID | 01118095 | In/Out Patient | Out |
| Name | Miss Ramisa | Refer. Dept | Neurology |
| Date of Birth | | Physician | Prof Hassanuzzaman |
| Age | 23y | | |
| Sex | Female | Examination Date | 8/13/2024 |
| Height | | Examination No | |
| Weight | | Examined by | |

01917100035

Miss.Ramisa

Motor Nerve Conduction Study

| Site | Amplitude | Area | Segment | Interval (ms) | NCV (m/s) | NCV N.D |
|---|---|---|---|---|---|---|
| Median, L | | | | | | |
| Wrist | 4.22ms 8.34mV | 26.92mVms | Wrist | 4.22ms | | |
| Elbow | 8.52ms 7.66mV | 27.60mVms | Wrist - Elbow 220mm | 4.30ms | 51.2m/s | |
| Ulnar, L | | | | | | |
| Wrist | 3.40ms 6.39mV | 21.17mVms | Wrist | 3.40ms | | |
| Elbow | 7.44ms 6.14mV | 20.11mVms | Wrist - Elbow 260mm | 4.04ms | 64.4m/s | |
| Axilla | 9.26ms 6.10mV | 19.78mVms | Elbow - Axilla 110mm | 1.82ms | 60.4m/s | |
| Median, R | | | | | | |
| Wrist | 4.98ms 12.89mV | 38.65mVms | Wrist | 4.98ms | | |
| Elbow | 9.36ms 12.14mV | 36.94mVms | Wrist - Elbow 240mm | 4.38ms | 54.8m/s | |
| Ulnar, R | | | | | | |
| Wrist | 3.2ms 4.42mV | 21.21mVms | Wrist | 3.20ms | | |
| Elbow | 7.0ms 4.47mV | 17.89mVms | Wrist - Elbow 260mm | 4.70ms | 55.3m/s | |
| Axilla | 9.32ms 4.40mV | 17.72mVms | Elbow - Axilla 110mm | 1.42ms | 77.5m/s | |
| Tibial, L | | | | | | |
| Ankle | 3.7ms 11.07mV | 26.23mVms | Ankle | 3.70ms | | |
| Popliteal | 12.5ms 8.01mV | 21.68mVms | Ankle - Popliteal 400mm | 8.80ms | 45.5m/s | |
| Peroneal, L | | | | | | |
| Ankle | 5.2ms 2.74mV | 8.92mVms | Ankle | 5.20ms | | |
| Head of fibula | 11.2ms 2.62mV | 8.05mVms | Ankle - Head of fibula 310mm | 6.00ms | 51.7m/s | |
| Popliteal | 12.7ms 2.88mV | 9.29mVms | Head of fibula - Popliteal 90mm | 1.50ms | 60.0m/s | |
| Tibial, R | | | | | | |
| Ankle | 5.15ms 9.39mV | 25.16mVms | Ankle | 5.45ms | | |
| Popliteal | 13.15ms 6.16mV | 19.88mVms | Ankle - Popliteal 400mm | 7.70ms | 51.9m/s | |
| Peroneal, R | | | | | | |
| Ankle | 5.2ms 5.12mV | 16.60mVms | Ankle | 5.20ms | | |
| Head of fibula | 11.8ms 4.12mV | 14.35mVms | Ankle - Head of fibula 310mm | 6.60ms | 47.0m/s | |
| Popliteal | 13.65ms 4.25mV | 15.47mVms | Head of fibula - Popliteal 90mm | 1.85ms | 48.6m/s | |

F-wave Study

| Nerve | | Stim.Site | F-Lat. | F-Lat. N.D. | M Lat. | F-M Lat. | F Occurr. | Distance | FWCV | N.D. |
|---|---|---|---|---|---|---|---|---|---|---|
| Median | L | Wrist | 35.1ms | | 4.55ms | 30.55ms | 7/11.2% | | | |
| Ulnar | L | Wrist | 00 | | | | 0/10.0% | | | |
| Median | R | Wrist | 36.85ms | | 4.7ms | 32.15ms | 9/11.82% | | | |
| Ulnar | R | Wrist | 00 | | | | 0/12.0% | | | |
| Tibial | L | Ankle | 65.6ms | | | | 0/12.0% | | | |
| Tibial | R | Ankle | 71ms | | 5.1ms | 65.9ms | 2/12.5% | | | |

Sensory Nerve Conduction Study

| Site | Amplitude | Area | Segment | Interval (ms) | NCV (m/s) | NCV N.D |
|---|---|---|---|---|---|---|
| Median, L | | | | | | |
| Wrist | 2.44ms 65.80uV | 5.05uVms | Wrist | 2.44ms | | |
| Ulnar, L | | | | | | |
| Wrist | 2.02ms 49.10uV | 4.24uVms | Wrist | 2.02ms | | |

Median, R

| | | | | | | | |
|---|---|---|---|---|---|---|---|
| Wrist | 2.5ms | 55.60uV | 3.82uVms | Wrist | | 2.50ms | |
| Ulnar, R | | | | | | | |
| Wrist | 2.18ms | 68.60uV | 4.04uVms | Wrist | | 2.18ms | |
| Sural, L | | | | | | | |
| Sural | 2.74ms | 8.85uV | 0.49uVms | Sural | | 2.74ms | |
| Sural, R | | | | | | | |
| Sural | 2.92ms | 6.72uV | 0.21uVms | Sural | | 2.92ms | |

Impression

NCS of all four limbs are done.

FINDINGS: Motor NCS reveals slightly prolonged DL in right median & left ulnar nerves with reduced CMAP amplitudes only in right ulnar nerve.
F-responses are absent in both ulnar nerves and prolonged in all other studied nerves.
Sensory NCS findings are within normal limit.

IMPRESSION: These electrophysiological findings are consistent mostly with polyradiculopathy.

N.B: Please correlate clinically (? Early features of GBS).

Date 8/13/2024

Signature Dr.Akramul Azam

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**Figure 2: NCS of all four limbs**

After clinical and laboratory evaluation the patient was diagnosed as a case of SLE with lupus nephritis with AIDP. She was treated with IV methylprednisolone. After getting treatment her 6th nerve palsy improved, and muscle power became 5/5 in the upper limb and 4/5 in the lower limbs. Sensory abnormalities resolved. UTP decreased to 3 gm/day, leukopenia improved (TWBC 9000), and proximal myopathy partially improved. She was put on oral prednisolone 0.5mg/kg/day and Tab Mycophenolate Mofetil 2gm/day.

## DISCUSSION

Study shows that in Bangladesh Fever (71.0%), joint pain (60.0%), and alopecia (28.0%) are the most common initial presentations. Constitutional symptoms such as fatigue, malaise and weakness (23.5%), malar rash (21.0%), oral ulcer (20.0%), photosensitivity

(15.0%), and Raynaud's phenomenon (13.0%) are also present in variable proportions [5]. The prevalence of Neuropsychiatric lupus (NPSLE) in SLE patients is 30-40% [6].

NPSLE means neurological and psychiatric disorders. It has a high heterogeneity of clinical phenotypes, including headaches, psychiatric symptoms, and peripheral neuropathy [7]. The symptoms of NPSLE have been classified into 19 neuropsychiatric (NP) manifestations by the American College of Rheumatology (ACR). These manifestations are again classified into two major divisions based on the involvement of central and peripheral nervous systems. Among the peripheral nervous system involvement Guillain-Barre syndrome, autonomic dysfunction, mononeuropathy, polyneuropathy, myasthenia gravis, and cranial nerve palsy are reported [8]. Among the autoantibodies Anti-Sm antibodies (anti-Sm) and anti-U1-ribonucleoprotein antibodies (anti-RNP) are frequently found in patients with NPSLE [7]. Anti-Sm recognizes the U1, U2, U4/U6 and U5 small nuclear RNPs (snRNPs), and anti-RNP recognizes the U1snRNPs [8, 9]. The presence of serum anti-Sm is associated with the prevalence of diffuse NPSLE [10], and high mortality in NPSLE [11]. High anti-RNP levels were also found in patients with NPSLE in both serum and cerebrospinal fluid [12]. Matsueda *et al.*, showed that anti-Sm and anti-RNP bind on the cell surface of monocytes and synergistically enhance the production of IL-6 by human monocytes causing axonal degeneration, demyelination [13] the incidence of SLE in patients with GBS is less common about 0.6% to 1.7% [14]. Initial presentation as AIDP or cranial nerve involvement is very rare. It is uncertain whether the pathogenic mechanism of SLE is inflammatory, thrombotic, or mixed. The most widely accepted pathogenesis is antibody-mediated neuronal involvement, which causes vasculopathy, intrathecal production of proinflammatory cytokines, and accelerated atherosclerosis [15]. In our case patient had both AIDP and 6th cranial nerve palsy both are very rare manifestations. Gradually there was involvement of the musculoskeletal system and kidney. Anti-Sm antibody is associated both with peripheral nerve involvement and renal involvement and it is positive in our patient. Immunosuppressive therapy like high-dose steroids, Cyclophosphamide, Mycophenolate Mofetil, and antimalarial drugs can be used to treat this condition. Rituximab is also a very good option. In our case, we treated the patient with high-dose steroids and MMF. Cyclophosphamide was refused by the patient due to fertility issues. After getting treatment with IV methylprednisolone patient was improved evidenced by improvement of muscle power and resolution of 6th nerve palsy, reduction of proteinuria.

## CONCLUSION

SLE is a disease with diverse presentation. Initial presentation may be with widespread organ damage. Early initiation of immunosuppressive may be life-saving for this patient as every hour matters in this case. For early initiation of therapy, early diagnosis is mandatory. So high level of clinical suspicion is necessary when a young female presents with multi-organ involvement within a very short period.

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