

# An Unusual Presentation of Systemic Lupus Erythematosus with Vasculitic Polyneuropathy

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## Abstract

We report a case of systemic lupus erythematosus (SLE), in a 48-year-old woman, a known hypertensive and recently diagnosed diabetic, with a predominant complain of acute onset bilateral lower limb weakness and loss of sensation of the bilateral palm and soles with bowel incontinence and excessive hair loss. On evaluation, her vitals were stable, cardiovascular, respiratory, and abdominal systemic examination revealed no significant abnormality. Neurological examination revealed a normal tone with distal muscle weakness more in the lower limb than in the upper limb. She was thoroughly evaluated for the above-mentioned complaints and examination findings. Nerve conduction study was done - which revealed upper limb - severe asymmetric motor sensory neuropathy - axonal type and lower limb - axonal symmetrical severe polyradiculoneuropathy. Nerve biopsy revealed a vasculitic neuropathy. Autoimmune workup was positive for antinuclear antibodies, and low C3 levels, with direct Coombs test positive, increased UPCR ratio. According to the Systemic Lupus International Collaborating Clinic criteria, she was diagnosed with SLE. Hence, this is a case of SLE, with a primary presentation of a vasculitic neuropathy, an unusual occurrence.

**Key words:** Atypical presentation of systemic lupus erythematosus, Systemic lupus erythematosus, Vasculitic polyneuropathy

## INTRODUCTION

Systemic lupus erythematosus (SLE) is an autoimmune disorder with a variety of manifestations. These manifestations are brought forth by immune complex deposition in various tissues such as skin, kidney, and vasculitis. This immune complex activation occurs through a variety of pathways, genetic, epigenetic, and environmental factors.

Clinical manifestations of SLE include a wide array of symptoms and signs, which can affect almost every part of the human body.

Approximately 10–20% of patients with SLE show an involvement of the peripheral nervous system.<sup>[1,2]</sup>

Primary presentation of an axonal polyradiculopathy with a vasculitic neuropathy in a patient with SLE has rarely been reported.<sup>[3]</sup>

Involvement of peripheral nervous system in patients with SLE includes non-specific axonal, microvascular changes, and vasculitis.<sup>[2,4]</sup>

## CASE DESCRIPTION AND RESULTS

A 48-year-old woman, hailing from Tambaram, Chennai, presented to us with complaints of acute onset lower limb weakness for 1 week, which was precipitated on exertion, and was bedridden and dependent for daily activities ever since. She also had a loss of sensation of the soles and feet for the past 1 week. She had no previous history of gait instability or lower limb weakness. She had no history of syncope, seizures, loss of consciousness, fever, headache, or vomiting.

She also complained of increased hair loss for 2 weeks and bowel incontinence for 4 days.

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Patient Name:	Mrs. USHA	Sample Collection Date:	19/06/2018 10:23 AM
Age :	48 years	Lab Reference No:	X-3076/18
Gender:	Female	Report Generated Date:	26/06/2018 01:11 PM
Ward Name/Collection Centre:	Biopsy Room	Lab Name:	Neuropathology
<b>Sample Details : H-1806190004 (Nerve)</b>			
<b>NERVE BIOPSY</b>			
<b>Nature Of Specimen:</b> Received a nerve segment measuring 1.8cm. All processed - A1A2.			
<b>Histopathology Report:</b> Section from the nerve segment shows dense perivascular lymphocytic infiltrate around medium sized epineurial vessels with transmural inflammation as well as an occasional small sized vessels. Acute axonal breakdown is present. K-pal myelin stain shows moderate to severe non - uniform loss of myelinated fibers with many regenerating clusters. Perl's stain for hemosiderin is negative.			
<b>Final Impression:</b> Vasculitic neuropathy; Left sural nerve.			
<b>Drafted By:</b> Dr. Anusha S PDF Resident		<b>Reported By:</b> Dr. T C Yasha Professor & Head	

Figure 1: Nerve biopsy report

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<b>DEPARTMENT OF SURGICAL GASTROENTEROLOGY</b> <b>UPPER GI ENDOSCOPY REPORT</b>			
NAME :	Mrs. USHA	PATIENT ID :	1102921
REF BY :	FMW I	SEX :	FEMALE
		AGE :	48
		DATE :	09-Jun-18
Pharynx & Larynx :			
Oesophagus :	Mucosa appears normal.		
QG Junction :	Normal		
Stomach			
Fundus			
Body	Mucosa appears normal.		
Antrum			
Pylorus			
Duodenum Part - I	No abnormality seen.		
Part - II			
Impression	- No abnormality noted in this study.		

Figure 2: Upper gastrointestinal scopy report

She had no history of early morning stiffness of joints, rash, decreased urine output, no history of oral ulcers, headaches, memory loss, seizure episodes, photosensitivity, malar rash, chest pain, breathlessness, or sicca-like symptoms.

She is a known case of systemic hypertension on calcium channel blockers and a recently diagnosed diabetic, on biguanides. She had a history of a bulky uterus for which hysterectomy and bilateral oophorectomy were done 3 years ago, histopathological evidence of which suggested leiomyoma with no malignant etiology.

On examination, she was conscious, oriented, and afebrile, with no signs of neuropsychiatric manifestations. She was thin built and moderately nourished. Essential tremor of

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<b>DEPARTMENT OF SURGICAL GASTROENTEROLOGY</b> <b>COLONOSCOPY REPORT</b>			
NAME :	Mrs. USHA	PATIENT ID :	1102921
REF BY :	FMW I	SEX :	FEMALE
		AGE :	48
		DATE :	25-Jun-18
Rectum			
Sigmoid Colon			
Ascending Colon			
Splenic Flexure			
Transverse Colon			
Hepatic Flexure			
Ascending Colon			
Caecum			
Ileum			
Impression	Scope passed upto caecum. Two nodular granular areas of 1cm size seen in the rectum, 10cms from the anal verge the opposite walls. Both biopsied. Rest of the rectum and colon upto caecum appear normal. Vascular pattern preserved. Biopsy (A) taken from Caecum, ascending colon and Biopsy (B) taken from sigmoid colon.		

Figure 3: Lower gastrointestinal scopy report

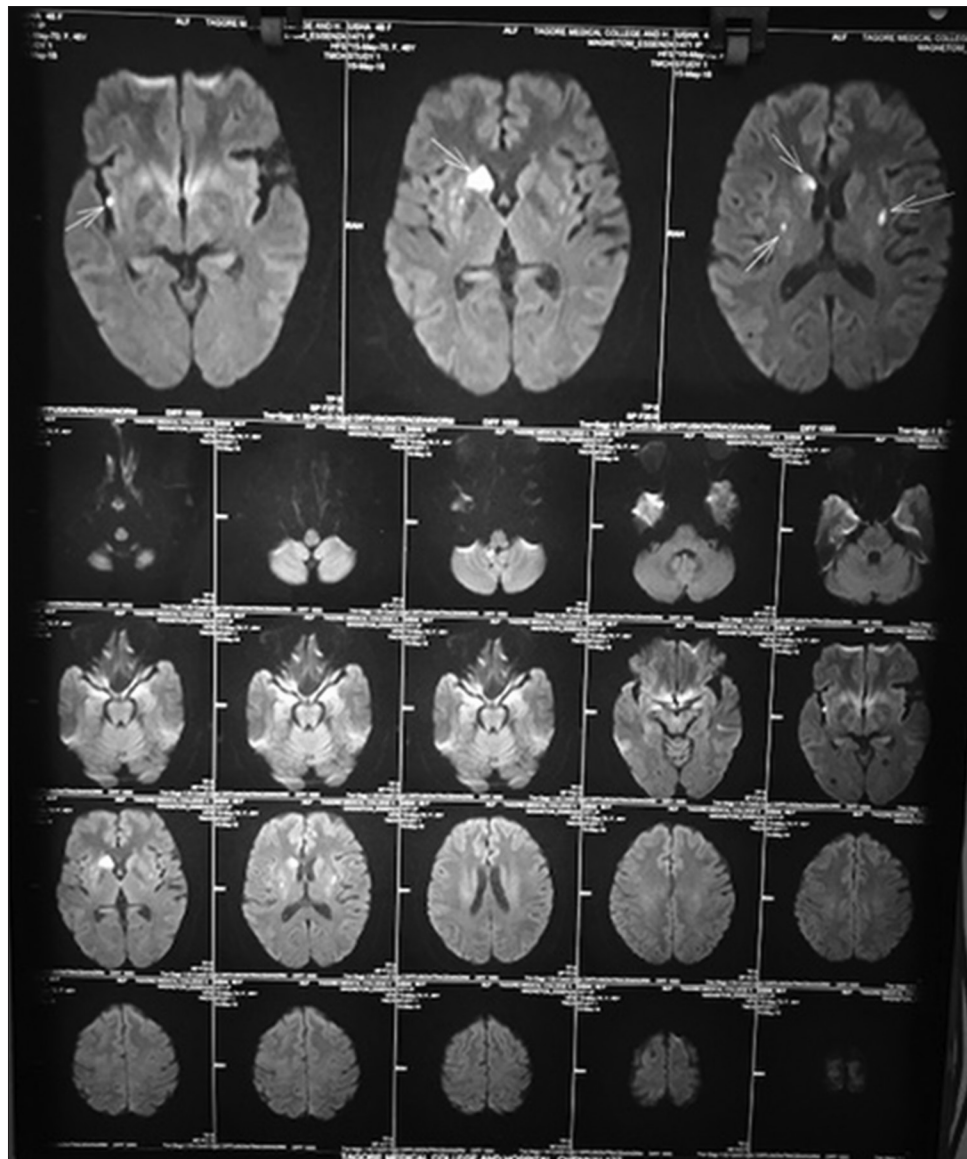
the hands was present and had a left hand hypothenar muscle wasting (h/o trauma, surgical fixation done 5 years back). She was pale, with no signs of icterus, cyanosis, clubbing, pedal edema, or lymphadenopathy. Her pulse rate was 88 beats per minute, regular and normal volume. Blood pressure was 140/100 mm of mercury and she was saturating at 99% in room air.

Cardiovascular, respiratory, and abdominal examination revealed no significant abnormality.

Central nervous system examination revealed a normal higher motor function, with bilaterally equal and reactive pupils. Essential tremor of the hands was present. Cranial nerve examination was normal. Motor system examination revealed a normal tone, with a 5/5 power of the upper limb and lower limb power being bilaterally 4/5 proximally and 3/5 power distally. Deep tendon reflexes revealed a normal reflex of the biceps, triceps, supinator, and knee. Bilateral ankle jerk was absent with plantar reflex being flexion bilaterally. Sensation, joint position sense, and vibration sense were intact. Cerebellar signs on examination were negative, and Romberg's test could not be assessed.

She was thoroughly evaluated for the above-mentioned complaints and examination findings.

Her blood investigations showed hemoglobin of 8.0, mean corpuscular volume of 81, mean corpuscular



**Figure 4: Magnetic resonance imaging brain**

hemoglobin of 25, total counts of 14,300, neutrophils - 67, lymphocytes 26, eosinophils 01, prothrombin time 13, international normalized ratio 1.0, activated partial thromboplastin time 31, platelet counts of 290,000, HbA1c 6.5, urea 49, creatinine 0.9, sodium 137, potassium 4.5, chloride 100,  $\text{HCO}_3$  30, calcium 8.1, phosphorus 3.5, uric acid 2.6, magnesium 1.9, total bilirubin 0.7, direct/indirect bilirubin - 0.2/0.5, alanine aminotransferase 27, aspartate aminotransferase 16, total protein 5.2, albumin 2.8, globulin 3.5, alkaline phosphate 103, thyroid-stimulating hormone 1.30, and free t4 2.05.

Urine analysis showed proteinuria with spot urine PCR of 1.21. Creatine kinase total was 08, lactate dehydrogenase 147, C-reactive protein positive (12), rheumatoid factor negative, HIV negative, HBsAg negative, iron 34, TIBC 175,

percentage transferrin saturation 19, cholesterol 61, TGL 75, HDL-c 21, LDL-c 29, and VLDL 15.

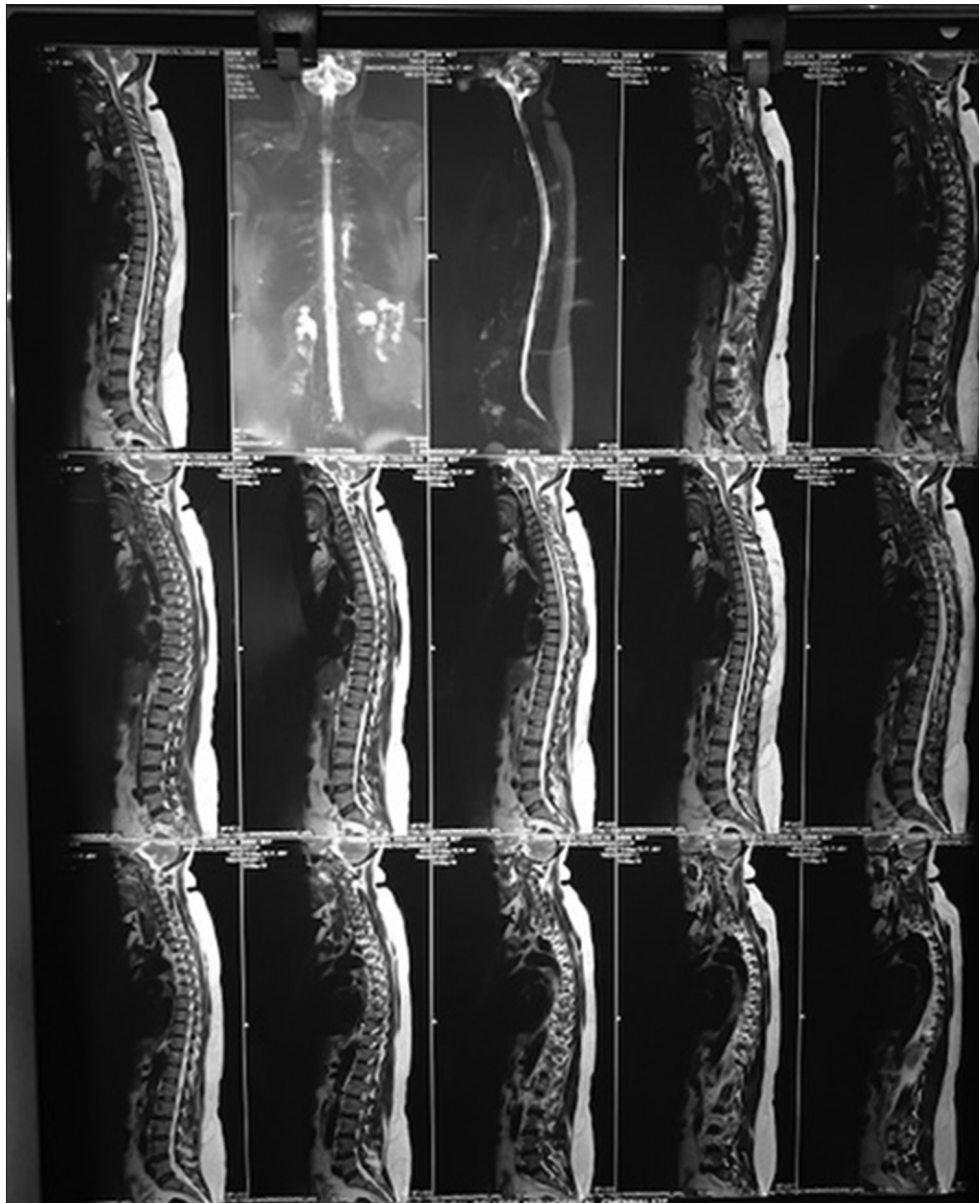
Antinuclear antibodies (ANAs) were 2+ with a positive direct Coombs test.

USG abdomen showed medical renal disease and left renal cortical cyst.

Echocardiogram showed Grade 1 diastolic dysfunction, normal left ventricular function, ejection fraction = 65%, and no regional wall motion abnormality.

Initial differentials thought of were a spinal pathology, causing a predominant lower limb weakness with bowel incontinence. Magnetic resonance imaging (MRI) spine was done which revealed a diffuse disc bulge at L3-L4,





**Figure 5: Magnetic resonance imaging spine**

L5-S1, with nerve root compression of L3-L4. However, this did not explain the symptoms manifested. MRI brain was also done which showed - acute non-hemorrhagic infarcts involving head of right caudate nucleus, anterior limb of right internal capsule, and bilateral basal ganglia.

Hence, nerve conduction study was done - which revealed - upper limb - severe asymmetric motor sensory neuropathy - axonal type - lower limb - axonal symmetrical severe polyradiculoneuropathy. She was also evaluated for her bowel disturbance - contrast-enhanced computed tomography showed transverse colon and ileocecal region wall thickening - Crohn's etiology/malignancy. Hence, colonoscopy and biopsy were done which showed an

ulcer, biopsy of which showed non-specific granulation tissue.

Nerve biopsy was done from the sural nerve which showed a vasculitic neuropathy.

Hence, complete autoimmune workup was done which showed positive - ANA 3+, AB RO-52, SS-A AB, AB TO NUCLEOSOMES, and LOW C3 [Figures 1-5].

## DISCUSSION

Thus, according to the Systemic Lupus International Collaborating Clinic criteria (1), she had clinical symptoms

**Table 1: Systemic Lupus International Collaborating Clinic criteria for classification of systemic lupus erythematosus**

Clinical manifestations	Immunologic manifestations
Skin	ANA > reference negative value
Acute, subacute cutaneous LE	Anti-dsDNA
Chronic cutaneous LE	Anti-Sm
Oral ulcers	Antiphospholipid
Alopecia	Low serum complement
Synovitis	Positive direct coombs test
Renal	
Prot/Cr $\geq 0.5$	
RBC casts	
Biopsy <sup>a</sup>	
Neurologic	
Seizures, psychosis, mononeuritis, myelitis, peripheral or cranial neu-ropathies, acute confusional state	
Hemolytic anemia	
Leukopenia (<4000) or lymphopenia (<1000)	
Thrombocytopenia (<100,000)	

<sup>a</sup>Renal biopsy read as systemic lupus qualifies for classification as SLE even if none of the other above features are present. Interpretation: Presence of any four criteria (must have at least one in each category) qualifies patient to be classified as having SLE with 93% specificity and 92% sensitivity. ANA: Antinuclear antibody, Cr: Creatinine, LE: Lupus erythematosus, Prot: Protein, SLE: Systemic lupus erythematosus

such as - neurologic manifestation involving peripheral neuropathy, renal involvement involving an elevated protein-creatinine ratio, immunological signs of a positive ANA, low serum complement, and positive direct Coombs test. Hence, having met the criteria, a diagnosis of SLE was made [Table 1].

## CONCLUSION

Hence, a diagnosis of SLE was made, with an unusual presentation of a vasculitic neuropathy.

The patient was pulsed with steroid therapy, Solu-Medrol for 5 days after which she was started on maintenance doses of steroids and mycophenolate mofetil.

The patient symptomatically improved, regained power eventually with steroid therapy, and is currently ambulant for less than normal daily activities.

## ACKNOWLEDGMENT

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## REFERENCES

- Feinglass EJ, Arnett FC, Dorsch CA, Zizic TM, Stevens MB. Neuropsychiatric manifestations of systemic lupus erythematosus: Diagnosis, clinical spectrum, and relationship to other features of the disease. *Medicine (Baltimore)* 1976;55:323-39.
- Johnson RT, Richardson EP. The neurologic manifestations of SLE: A clinical-pathological study of 24 cases and review of the literature. *Medicine* 1968;47:337-9.
- Stefurak TL, Midroni G, Bilbao JM. Vasculitic polyradiculopathy in systemic lupus erythematosus. *J Neurol Neurosurg Psychiatry* 1999;66:658-66.
- McCombe PA, McLeod JG, Pollard JD, Guo YP, Ingall TJ. Peripheral sensorimotor and autonomic neuropathy associated with systemic lupus erythematosus. Clinical, pathological and immunological features. *Brain* 1987;110 (Pt 2):533-49.

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