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Case Report

A Rare Case of Spinal Cord Infarction in Young Female

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Abstract

Background: Systemic Lupus Erythematosus (SLE) with underlying undiagnosed Anti phospholipid antibody (APLA) syndrome may have rare catastrophic cardiac and neurological complications. These complications must be kept in mind while evaluating patients with these disorders so that prompt action must be taken.

Case: Most patients presented with Paraplegia with sphincteric involvement of acute onset are evaluated on an urgent basis in view of spinal cord compression. In our case, young women of reproductive age group, unmarried having SLE presented with acute paraplegia with urinary retention without sensory level were evaluated for aortic dissection first, then for Guillain Barre syndrome, Demyelinating disorder as Transverse myelitis as per top differentials then spinal cord infarction.

Keywords: APLA syndrome; Aortic dissection; SLE; Spinal cord infarction

Introduction

Being a thrombo -inflammatory disorder, Anti-Phospholipid syndrome complicates systemic lupus erythematosus. [1] It causes vascular thrombosis and obstetrical complications by circulating antiphospholipid antibodies [2]. Regarding pathogenesis, these antibodies engage phospholipids and phospholipid-binding proteins at cell surfaces to activate the endothelium, platelets, and leukocytes—thereby tipping the circulating intravascular milieu toward in situ thrombosis while also promoting other autoimmune and inflammatory complications [3,4]. Cerebral vessels along with a deep venous system of lower limbs are frequently involved in thrombosis [5,6]. It may be Primary or Secondary means associated with other rheumatological disorders.

Neurological manifestations of Anti Phospholipid Syndrome include chorea/ athetosis, cognitive decline, longitudinal myelitis, migraine headache, retinal ischemia, seizures, and white matter hyperintensities.

Spinal cord infarction is a rare and catastrophic neurological complication. Either ischemic or hemorrhagic, these have apoplectic onset evolving mostly over minutes. Other confounding diagnoses, including acute transverse myelopathy, Guillain Barre syndrome, and mass lesions in the spinal canal develop over 24-72 hours with an acute but slower evolution than a vascular phenomenon [7,8].

We are reporting this rare arterial neurological complication of undiagnosed Anti Phospholipid Antibody syndrome in a young female with Systemic Lupus Erythematosus.

Case Presentation

A female of 28 years of age, a teacher by occupation, unmarried, diagnosed as having Systemic lupus erythematosus for 10 years (non-compliant with medications), presented in emergency with a history of sudden onset bilateral lower limb weakness upon awakening 10 days back then develop urinary retention 2 days after weakness. There was a history of mild backache and one episode of hypotension documented up to 80 mmHg systolic blood pressure, not associated with chest pain or sweating. There was no history of fall or trauma. No history of preceding febrile illness. Although she had diarrhea 2 weeks before it. No history of diplopia, dysarthria, dysphagia, or dyspnea. No history of headache, fits, paraesthesias, episodes of such illness in the past, TB contact, recent vaccination, or surgery. There was a history of hair fall, oral ulcers, and arthralgias.

On examination, she was lying on a bed with no obvious signs of respiratory distress with a Foley catheter in place. Vital signs were Blood pressure of 100/70 mmHg in both arms, Pulse of 75 beats per min, respiratory rate of 18 breaths/min, and Temperature of 37 degrees Celsius. A general physical examination revealed bilateral periorbital puffiness, mild pallor, and grade 2 clubbing. On neurological examination, Glasgow coma scale (GCS) of 15/15, Pupils were bilaterally equal and reactive, and cranial nerves were intact. Dilated Fundoscopic examination of the eyes was unremarkable. According to MRC grading, power in muscles was 5/5 in bilateral upper limbs with 1/5 in the right lower limb and 2/5 in the left lower limb both proximally and distally. Sensations of both pinprick, temperature, and joint position were intact. Deep tendon reflexes were intact in both upper limbs with depressed in the lower limbs. Plantar reflexes were bilateral equivocal. Cerebellar functions were intact in upper limbs and cannot be assessed in lower limbs due to reduced muscle power.

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Spine examinations were unremarkable. Gait cannot be assessed due to profound muscle weakness of the lower limbs. The rest of the systemic examination was unremarkable. Preliminary Investigation done (shown in table 1).

Investigations	Results	Normal ranges
TLC	10660/ul	[140000-400000]
Hb	8.6g/dl	[12-15g/dl]
Platelets	256000/ul	[4000-10000/ul]
PT	13	[10-14]
APTT	23.8	[28-42]
INR	0.68	[0-1.1]
Bilirubin total	0.182	[0.3-1.2]
ALT	39.1	[4-42U/L]
ALP	138	[35-105]
Urea	67.9	[13-43]
Creatinine	0.535	[0.5-1.1]
Sodium	136.4	[136-146]
Potassium	4.48	[3.5-5.1]

Chest X-ray and electrocardiography were unremarkable. A differential diagnosis of the para-paretic variant of Guillain Barre syndrome was made based on history and examination. Cerebrospinal fluid studies (CSF) revealed shown in table 2.

CSF RE	RESULTS	Normal ranges
Color	Colorless	colorless
Turbidity	Clear	clear
WBCs	10/mm3	<5/mm3
Xanthochromia	Absent	absent
Neutrophils	10%	
Lymphocytes	90%	
Protein	36mg/dl	<45
Glucose	42mg/dl	40-70
Fluid LDH	34 IU/L	
Gram stain	Negative	
AFB stain India	Negative	
ink stain Gene	Negative	
Xpert	Negative	

HIV screening was negative. Nerve conduction studies (NCS) came out to be normal. Computed tomography Aortogram was done to rule out aortic dissection also came out normal. Magnetic resonance imaging of the thoracic spine with contrast was done that showed a long segment central intramedullary abnormal MR signal within the central gray matter of the cord extending from the level of C6 vertebral body superiorly down to the level of T9 vertebral body inferiorly giving differentials of transverse myelitis, spinal cord infarction(Figure 1).

MRI Thoracic spine with Diffusion-weighted images shows acute spinal cord infarction with characteristics of owl eye appearance on T2WI (Figure 2).

Initial treatment of IVMP was given

Her workup was extended. Antinuclear antibody (ANA) by Immunofluorescence and Rheumatoid factor (RF) came positive. Serum

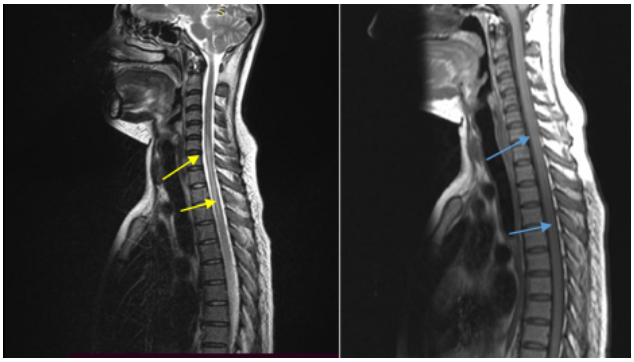

Figure 1: MRI CERVICODORSAL Spine T2(A) sagittal view showed hyperintense signal from C6 to T9 pointed out by yellow arrows (B) T1sagittal view showed abnormal signal from C6 to T9 indicated by blue arrows.

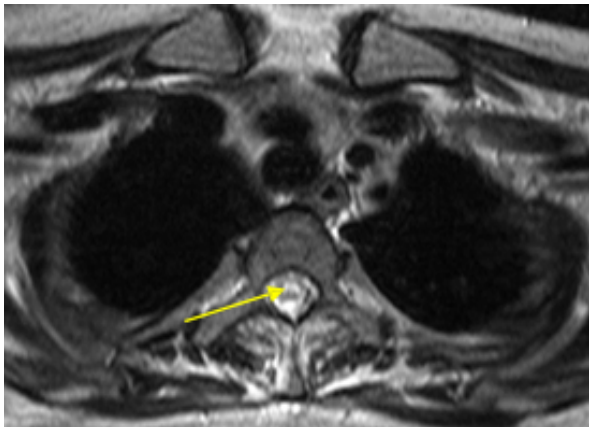

Figure 2 : T2W1 axial view showed hyperintense signal (classical owl's eye appearance involving central anterior cord substance indicated by yellow arrows).

Anti-dsDNA (IgG) was negative. The erythrocyte sedimentation rate (ESR) was 140 mm/1st hour. Urine routine examination was normal. 24 hours urine protein was 295 mg/24 hrs. (<140). Ultrasonography renal tract was normal. Serum Complement levels (C3 and C4) were reduced. Serum lipid profile (cholesterol-284mg/dl, triglycerides-417 mg/dl, HDL-29.7 mg/dl, LDL-171 mg/dl). Echocardiography was normal. Holter monitoring was unremarkable. Thyroid function test and serum vitamin B12 were normal. Serum Vitamin D3 levels were reduced. MRI brain with contrast was unremarkable. Serum Anti-Aquaporin-4 Antibody was negative. Antiphospholipid antibodies (anti- cardiolipin and beta 2 glycoproteins) came out positive but Lupus anticoagulant antibody was Negative. Anti- MOG Antibodies were negative.

Initially, a High dose of Intravenous Methyl Prednisolone 1g was given for 7 days along with vitamin D replacement. Hydration with stress ulcer and DVT prophylaxis was given. She was diagnosed as having a spinal cord infarction due to underlying Secondary Anti Phospholipid Antibody syndrome The rheumatology, cardiology, and Nephrology department were kept on board. The rheumatologist started medications like hydroxychloroquine 200mg 1 tablet once daily and Atorvastatin 10mg 1 tablet nightly. Physiotherapy done. She has shifted to oral Steroids. She was started on vitamin K antagonist warfarin with a target INR of 2-3. According to the National Institute

of Health (NIH) protocol of Cyclophosphamide Pulse therapy, the standard treatment was initiated. The protocol consists of intravenous cyclophosphamide (0.5-1 gm/m2, adjusted to white blood cell (WBC) nadir, given monthly for the first six months and then quarterly for at least 12 months.

After receiving the first pulse therapy of cyclophosphamide with MESNA, her power of lower limbs improved to 4/5 both proximally and distally bilaterally. She was discharged on oral steroids with the plan of tapering along with warfarin, vitamin D supplements, Hydroxychloroquine, and statin therapy with advice to keep regular follow up with the rheumatology department for her long-term management.

Discussion

An antiphospholipid syndrome is an autoimmune disorder driven by autoantibodies disrupting coagulation and fibrinolysis resulting in an increased risk of thrombotic events, pregnancy morbidity, and other autoimmune or inflammatory complications. For most patients with thrombotic antiphospholipid syndrome, Vitamin K antagonists remain the most appropriate treatment and, appear superior to direct oral anticoagulants. Beyond anticoagulants and anti-aggregants, recent research has highlighted additional potential therapeutic targets within the innate immune system, including the complement system and NETs. The potential role of immunomodulatory treatments in antiphospholipid syndrome management is rightly receiving increased attention.

Being associated with other rheumatological disorders, classified as Secondary APLA Syndrome. Diagnosis of this disorder was made based on classification criteria of APLA syndrome based on clinical and laboratory criteria. Treatment and management are according to EULAR recommendations.

Conclusion

Neurological manifestations of this syndrome range from headaches to stroke, seizures, transverse myelitis, and even catastrophic spinal cord infarction. We report this case to emphasize keeping our horizon broad. As in this case, we ruled out almost all the possible important differentials of the presenting symptoms of a patient. Early recognition and diagnosis leading to prompt treatment of the underlying disorder are necessary for good prognosis and less morbidity.

List Of Abbreviations: SCI: Spinal cord infarction , APLA: Anti phospholipid antibody syndrome , EULAR :European Alliance of Associations for Rheumatology , NIH : National institute of Health ,WBC :White blood cell , ANA : Anti Nuclear antibody , IVMP : intravenous methylprednisolone , NCS : Nerve conduction Studies , CSF : Cerebrospinal fluid.

Declaration

Ethical approval

It has been approved by Departmental Review Board and Ethics Committee ,Neurology Department of Pakistan Institute Of Medical Sciences.

Consent to participate and publication

Consent to participate and publication has been taken by participant .

Competing/conflict of interest

The authors declare that they have no competing interests.

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Author's contribution

All authors participated in data collection, manuscript writing .All authors read and approved manuscript.

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