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RESEARCH ARTICLE

NEWLY DIAGNOSED SYSTEMIC LUPUS ERYTHEMATOSUS PRESENTING AS CERVICAL MYELOPATHY: A RARE CASE PRESENTATION

[1]Dr. Ravi Kumar, [2]Dr. Vineender Reddy, B., [3]Dr. Rashi Srivastava, [4]Dr. Ashutosh Chajjed, [5]Dr. Dinesh Saini and [6]Dr. Mannan

[1]Senior resident Department of General medicine, Ananta Institute of Medical Sciences
[2]Final year resident, Department of General Medicine, Ananta Institute of Medical sciences
[3,4,5]Assistant professor, Department of General medicine, Ananta Institute of Medical sciences
[6]Junior resident, Department of General medicine, Ananta Institute of Medical sciences

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***Corresponding author:**
Dr. Ravi Kumar

ABSTRACT

SLE is an autoimmune disease in which organ damage is mediated by auto-antibodies and immune complexes. Around 90% of patients are women of child bearing age. It is ~9 times more prevalent in women than in men. SLE is newly diagnosed in 4 lakh people each year worldwide 1,2. SLE can involve many organ systems likewise musculoskeletal, dermatological, renal, neurological, hematological, cardiopulmonary, ocular and gastrointestinal. Severity of SLE varies from mild and intermittent to severe and fulminant. Neurological manifestations are common in SLE occuring in ~ 60% of patients but Myelopathy is rarest presentation among neurological manifestations (< 1%). Hence we are reporting a rare case of a young female patient of SLE presented with cervical myelopathy Subject Area: Internal Medicine and Rheumatology

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INTRODUCTION

SLE is an autoimmune disease in which organ damage is mediated by auto-antibodies and immune complexes. Around 90% of patients are women of child bearing age. It is ~9 times more prevalent in women than in men. SLE is newly diagnosed in 4 lakh people each year worldwide[1,2]. SLE can involve many organ systems likewise musculoskeletal, dermatological, renal, neurological, hematological, cardiopulmonary, ocular and gastrointestinal. Severity of SLE varies from mild and intermittent to severe and fulminant. Neurological manifestations are common in SLE occuring in ~ 60% of patients but Myelopathy is rarest presentation among neurological manifestations (< 1%). Hence we are reporting a rare case of a young female patient of SLE presented with cervical myelopathy.

CASE REPORT

A 22 year old female patient admitted in medicine ward in Ananta institute of medical sciences, Rajsamand with history of fever and rash over face since 2 years, B/L symmetrical lower limb weakness since 6 months, b/l upper limb Fever was releived with antipyretics but was reappearing again. On general physical examination, patient had non scarring alopecia, facial rash sparing nasolabial folds and pallor. On neurological examination, patient's higher mental functions were normal, there was no cranial nerve involvement. Patient had decreased power (3/5) in all four limbs along with increase in tone in all limbs in the form of spasticity, plantar reflex b/l extensor. All deep tendon reflexes were exaggerated with bilateral sustained ankle clonus, Hoffmann reflex and finger flexion reflex was present. On sensory examination, proprioception and vibration sense was lost, rhomberg's couldn't be assessed because of weakness. So, on the basis of history and examination a provisional diagnosis of non compressive cervical myelopathy was made. MRI cervical spine was done but it was normal. All the infectious causes were ruled out. On CSF examination, csf sugar was 45mg/dl (simultaneous blood sugar was 104mg/dl), csf protein was 106mg/dl, cells were 4/mm^3 (100% lymphocytes).So, csf findings were suggestive of non infectious cause but inflammatory cause couldn't be ruled out. Patient's ESR was 105mm, urinary albumin creatinine ratio was 520.5mg/gm. On the basis of general physical, systemic examination and investigations a diagnosis of connective tissue disorder was thought.

LABORATORY REPORT

Name [redacted]		Sex/Age : **Female / 22 Years**	Case ID : **40301502903**
Ref. By		Dis. At :	Pt. ID :
Bill. Loc. : Ananta medical college and hospital			Pt. Loc :
Reg Date and Time	: 19-Mar-2024 17:44	Sample Type : Serum	Mobile No. :
Sample Date and Time	: 19-Mar-2024 17:44	Sample Coll. By : non	Ref Id1 : 1403207
Report Date and Time	: 20-Mar-2024 14:58	Acc. Remarks : -	Ref Id2 :

TEST	RESULTS	UNIT	BIOLOGICAL REF RANGE	REMARKS

ANTI NUCLEAR ANTIBODY BY INDIRECT IMMUNOFLUORESCENCE

Dilution (D)	**1:100**	
Intensity On IF	**Intensity (++++)**	No Intensity
Pattern On If For ANA	**Nuclear Speckled**	
Suspected Antigen Specificity	**Sm,RNP,Scl-70,SS-b and others**	
Titre	**1:1000 to 1:3200**	<1:32
Clinical Significance	**SLE, MCTD, Scleroderma, Sjogren-Sicca complex syndrome and other connective tissue disease**	
Interpretation	**Positive**	
Follow - Up	**Clinical correlation and/or repeat testing after 6-12 weeks and /or confirmation by ANA profile(Refer interpretive note below)**	
Additional Remark for Liver Substrate	**Non significant**	

ANTI NUCLEAR ANTIBODY (Immunofluorescence, HEp2000-immunoconcepts)

Neuberg Supra

REFERENCE LABORATORIES

(A unit of Neuberg Diagnostics Private Limited)

LABORATORY REPORT

Name [redacted]		Sex/Age : **Female / 22 Years**	Case ID : **40301503424**
Ref. By		Dis. At :	Pt. ID :
Bill. Loc. : Ananta medical college and hospital			Pt. Loc :
Reg Date and Time	: 22-Mar-2024 18:20	Sample Type : Serum	Mobile No. :
Sample Date and Time	: 22-Mar-2024 18:20	Sample Coll. By : non	Ref Id1 : 1403207
Report Date and Time	: 23-Mar-2024 20:10	Acc. Remarks : -	Ref Id2 :

TEST	RESULTS	UNIT	BIOLOGICAL REF RANGE	REMARKS
F-Actin Ab EIA	<6	AU/mL	0 - 6 Negative 6 - 12 Intermediate >12 Positive	
DsDNA antibody IgG CLIA	H 52.10	IU/mL	< 20 IU/ml : Negative 20-25 IU/ml: Equivocal >25 IU/ml : Positive	

The blot includes various anti nuclear antibodies of IgG type detected in human sera against Sm, RNP, Sm/RNP, SSA/Ro60kD, SSB (La), Jo-1 (histidyl-t-RNA synthetase), Scl-70 (DNA topoisomerase I), PM-Scl 100, Ku, CENP-A/B(centromere A/B proteins), PCNA and Ribosome P0 antigens. The blot is carried out on automated dot blot analyzer (Blue-Diver) and offers reproducibility of > 90% and following sensitivity and specificity:

Antigen	Sensitivity	Specificity	Disease association with prevalence (%)
Nucleosome	67%	98%	Early marker of SLE (50-90%)
Ds-DNA	100%	100%	SLE (40-90%)
Histones			

Pt. Loc :

Reg Date and Time	: 22-Mar-2024 18:20	Sample Type : Serum	Mobile No. :
Sample Date and Time	: 22-Mar-2024 18:20	Sample Coll. By : non	Ref Id1 : 1403207
Report Date and Time	: 23-Mar-2024 20:10	Acc. Remarks : -	Ref Id2 :

TEST	RESULTS	UNIT	BIOLOGICAL REF RANGE	REMARKS

BIOCHEMICAL INVESTIGATIONS

Anti Nuclear Antibody by BlueDiver Quantirx(Quantitative IgG)

Nucleosome Ab	H 23	AU/mL	0 - 6 Negative 6 - 12 Intermediate >12 Positive	
Histones Ab	<6	AU/mL	0 - 6 Negative 6 - 12 Intermediate >12 Positive	
Sm Ab	H 89	AU/mL	0 - 6 Negative 6 - 12 Intermediate >12 Positive	
Ribosomes P0 Ab	<6	AU/mL	0 - 6 Negative 6 - 12 Intermediate >12 Positive	
PCNA Ab	<6	AU/mL	0 - 6 Negative 6 - 12 Intermediate >12 Positive	
Cenp-A/B Ab	<6	AU/mL	0 - 6 Negative 6 - 12 Intermediate >12 Positive	
Jo-1 Ab	<6	AU/mL	0 - 6 Negative 6 - 12 Intermediate >12 Positive	
RNP 68kd/A/C Ab	H 72	AU/mL	0 - 6 Negative 6 - 12 Intermediate >12 Positive	

So, ANA by indirect immunoflourescence was done which was positive with titre 1:1000 to 1:3200 and intensity was 4+. Further, ANA profile by immunoblot was sent and on the basis ANA blot along with low complement C3 levels a diagnosis of non – compressive type of cervical myelopathy secondary to SLE with? lupus nephritis was made. Patient was treated with MPS pulse therapy 1gm IV once daily for 3 days and patient showed drastic improvement in power (now 5/5)and also in bladder symptoms. Later patient was started on Tab. Prednisolone 40mg (1mg/kg) once daily followed by tapering of doses weekly over 1 month. Patient was kept on maintenance therapy with Tab, Azathioprine 25mg once daily and Tab. HCQ 200MG twice daily. Now, patient is on regular follow up and improved symptomatically with no further episodes of fever.

DISCUSSION

The pathogenesis of SLE involves systemic inflammation with elevated levels of type 1 interferon and auto-antibodies against nuclear antigens, such as double stranded DNA and nucleic acid binding proteins[3]. Development of clinical disease is thought to be because of exposure to environmental risk factors, such as UV light, cigarette smoking, EBV or silica from occupational exposure such as painting, foundry work or sand blasting in an individual with genetic [4-7]. SLE affects multiple organs. Although there are no formal diagnostic criteria, for clinical practice, the 2019 EULAR/ACR criteria designed for scientific investigation had 96% sensitivity and 93.4% specificity. For an SLE diagnosis by an expert [8], a positive Anti-nuclear antio body test result at a titre of >or= 1:30 is required. Typical clinical manifestations of SLE include fever, alopecia, skin exanthem, oral ulcers, joint pain and swelling. Neuropsychiatric symptoms consist of central and peripheral nervous system conditions, including headache, mood disorders, cognitive impairment, seizures, psychosis, myelopathy and polyneuropathy. In a meta-analysisof 22 studies that included 6055 patients with SLE the pooled prevalence of neuropsychiatric SLE was 52% (10.6% - 96.4%)[9], among which myelopathy is the rarest manifestation.

CONCLUSION

Systemic lupus erythomatosus is associated with immune mediated damage to multiple oragans and increased mortality. HCQ is first line therapy and reduces disease activity, morbidity and mortality. When needed, additional immunosuppressionand biologic therapies include Azathioprine, Mycophenolatemofetil, Cyclophosphamide, Belimumab, cyclosporine and anifrolumab.

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