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Original Research Paper

General Medicine

A RARE CASE OF CNS VASCULITIS IN SLE WITH SECONDARY APS – A CASE REPORT

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ABSTRACT Systemic lupus erythematosus is a chronic autoimmune disease that can affect any organ , including nervous system. The presentation of neurological symptoms in SLE presents a distinct clinical challenge. The clinical spectrum include stroke (ischemic and hemorrhagic) , seizures, altered mental status, cognitive impairment , inflammatory and demyelinating diseases. Here we are presenting a case of 25 year old female with no comorbidities presented with complaints of shortness of breath, yellowish discolouration of eyes and pedal edema since one month. On evaluation, diagnosed as SLE WITH SECONDARY APS who later developed signs of CNS vasculitis. Patient was treated with steroids , cyclophosphamide and supportive therapy, discharged and advised follow-up on OPD basis.

KEYWORDS : VASCULITIS, SLE, APS, MAGNETIC RESONANCE IMAGING, CYCLOPHOSPHAMIDE

INTRODUCTION

The brain is a common target in connective tissue diseases. In systemic lupus erythematosus (SLE), CNS involvement occurs in 14% to 80% of adults and 22% to 95% of children. Multifocal microinfarcts, cortical atrophy, gross infarcts, hemorrhage, ischemic demyelination, and patchy multiple-sclerosis-like demyelination are typical findings in neuropsychiatric lupus. Rheumatoid arthritis, Sjögren's syndrome, and mixed connective tissue disease rarely affect the CNS in a vasculitic pattern. CNS vasculitis is typically a late occurrence in these diseases.

Patients with SLE are at increased risk of stroke, with ischemic stroke being more common than intracerebral hemorrhage. between the presence of antiphospholipid antibodies (aPL) and/or valvular heart disease and the risk of stroke. Brain MRI is a critical test in the diagnosis of ischemic or hemorrhagic stroke, and magnetic resonance angiography (MRA) can detect vessel aneurysms. Echocardiography, carotid ultrasound, and electrocardiography are important diagnostic tests in the setting of suspected thromboembolic cerebrovascular disease.

CASE PRESENTATION

A 25 year old female with no comorbidities presented with complaints of shortness of breath, yellowish discolouration of eyes and bilateral leg swelling since one month. They went to local hospital where diagnosed as severe anaemia and referred to tertiary hospital for evaluation.

Patient had history of similar complaints in the past- one year ago, two PRBC transfusions were done. She also had bad obstetric history – P2L1D1A1 First pregnancy was uneventful; 2nd Pregnancy - still born, 3rd pregnancy - abortion at 26 weeks. Patient had history of recurrent oral ulcers, arthralgias since two years.

ON EXAMINATION:

Non scarring alopecia present
Pallor, icterus present;
Oral ulcers - present
Bilateral pitting pedal edema present
JVP- elevated
CVS- s1 s2 heard, no murmurs
Respiratory system - bilateral fine crepitations in Infra scapular regions

INVESTIGATIONS –

Hemoglobin – 3.6 g/dl
RBC – 3.02 MILLION/CUMM
WBC – 3640 cells/cumm
Platelets- 0.84 lakhs/cumm
Reticulocyte count – 3.5%
Total Bilirubin = 3.3; Direct -1.1; Indirect - 2.2
SGOT, SGPT- normal
CUE- within normal limits
Sr. Creatinine: 1.1 mg/dl
LDH, Ferritin increased
Direct coombs test positive
Anti ds DNA positive 240IU/ml
ANA positive 3+ (Homogeneous pattern)
C4 low (<0.0642)
C3 low (0.437)
Anticardiolopin antibodies – IgM , IgG Positive

She was diagnosed as SLE with secondary APS and started on pulse therapy.

After 10 days of treatment, she complained of numbness of right upper and lower limbs then MRI brain was suggested.

MRI BRAIN CONTRAST WITH MRA : Multiple areas of altered signal intensity appearing hypointense on T2/FLAIR with patchy T1 hyperintensity involving the left high fronto parietal lobe and splenium on the left side showing patchy and peripheral restricted diffusion with blooming on SWI associated with significant perilesional edema causing mass effect suggestive of acute to early subacute hematoma.

After ruling out all the possible causes, diagnosis was made as CNS vasculitis and started on Inj. Cyclophosphamide after adequate hydration and measures to prevent hemorrhagic cystitis.

DISCUSSION

Here We Discuss A Case Of 25 Year Old Female Who Was Diagnosed As Sle Based On Systemic Lupus International Collaborating Clinic Criteria For Classification Of Systemic Lupus Erythematosus – Scored 9 Positive Points. Diagnosed As Apla Based On International Consensus Statement On Revised Criteria For Classification Of Antiphospholipid Syndrome (aps) – Scored One Point Each With Pregnancy Morbidity And Anticardiolipin Antibodies. Therefore

Diagnosed As Acute Flare Of Sle With Secondary Aps And Started On Pulse Therapy – Inj. Methylprednisolone 1g Iv Od For 3 Days Followed By 1 Mg/kg Bw Oral Supplementation.

After Initiating Steroid Therapy, Patient Responded Well. Patient Suddenly Complained Of Paraesthesia Of Right Upper And lower limb after 10 days. On examination- tone was normal but power was 4/5 in both upper and lower limbs. MRI BRAIN revealed acute heamatoma in left frontoparietal lobe. Following this, her BP is found to be on higher side . patient was evaluated – her fundoscopy, renal doppler studies, renal parameters were normal. Then she was started on anti hypertensives. Bp was stabilized and neurologist consultation was taken. All the possible causes like hypertensive bleed, aneurysmal bleed, hypercoaguable states are ruled out and CNS VASCULITIS diagnosis was made. Then adequate hydration was given following which inj.cyclophosphamide was given. Patient improved gradually and was discharged and followed up on OPD bases.

Gold standard test for diagnosis is brain biopsy but it is not done due to the risk associated with procedure.

Other treatment option left is IVIG which is not considered in view of financial constraints.

CONCLUSION

CNS vasculitis is a rare and challenging diagnosis. A combination of clinical, serological and imaging findings have diagnostic value to start the treatment. Presentation of CNS vasculitis with hemorrhagic stroke is a rare entity. So we presented this case to bring it to the notice of the clinicians. Treating CNS vasculitis in patients with SLE remains difficult. In our case, high dose steroids with cyclophosphamide was associated with good clinical outcome.

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