Case Report:

Status epilepticus: A rare presentation of catastrophic secondary antiphospholipid antibody syndrome with SLE

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Date of submission: 07 July 2014; Date of Publication: 22 October 2014

Abstract:
We reported a case of 16 year old girl who presented as status epilepticus with multiorgan involvement. Based on clinical picture and results of diagnostic tests positive antibodies against β2 glycoprotein and cardiolipin with speckled ANA and negative DS DNA and Anti-Smith antibodies, we finally diagnosed the case as Catastrophic Secondary Antiphospholipid Antibody Syndrome(APLA) with Systemic lupus Erythematous (SLE)

key words: antiphospholipid antibody syndrome(apla), systemic lupus erythematous(sle), lupus anticoagulant(la).

Introduction:
APLA is an autoantibody mediated acquired thrombophilia characterised by recurrent arterial or venous thrombosis and/or pregnancy morbidity in presence of autoantibodies against phospholipid and Lupus. Catastrophic APLA is rapidly progressive thromboembolic event involving three or more organs/systems or tissue causing functional defects. APS occurring alone-primary with other autoimmune diseases secondary.¹,⁴

Case report:
16 yrs old girl came to our emergency department with status epilepticus, she had no previous history of convulsion, fever, palpitation, joint pain, rash, swelling, drug intake.
She had darkening of little toe of right leg since one year, for which no medication was taken.
On examination, patient was in stuporous state with tachycardia and accelerated blood pressure (180/100) no pulse difference in both limbs.
Patient regained consciousness on 4th day and complained of diminished vision upto fingers counting.

Investigations:
Anemia Leucocytosis predominantly neutrophilia, Platelets – adequate, ESR- 34mm.
Urine routine microscopy suggestive of hematuria with proteinuria.
Mildly elevated serum creatinine and urea.
CT brain (P+C)- diffuse vasculitis with punctuate bleeding.
Color doppler of both UL AND LL – mild vasculitis of all vessels.
Renal doppler with USG abdomen pelvis- no significant abnormality.
2D echo- moderate MR with mild pulmonary hypertension.

Ophthalmic examination shows decreased field of vision with optic atrophy.

Patient was positive for antiphospholipid $\beta_2$ glycoprotein and cardiolipin antibodies titre > 300 and ANA speckled pattern negative for DsDNA, anti Smith antibodies and other autoimmune disease antibodies.

Patient was treated with Inj. Heparin followed by Oral warfarin, Inj. Methylprednisolone, Inj. cyclophosphamide in cycles, Anticonvulsants, Antihypertensives. After 15 days patient was discharged with vision upto 6/24 and no other focal neurological deficit.

**Discussion:**

APLA is associated with thromboembolic events rather than clinical bleeding.

Sydney Criteria-2006 for its diagnosis is as follows:

1) One or more clinical episodes of arterial/venous/small vessel thrombosis in any tissue or organ.

And

2) Pregnancy morbidity defined as:

   a) One/more unexplained deaths of normal fetuses at or beyond 10th week or

   b) Premature birth of morphologically normal fetus before 34th week or

   c) Three/more unexplained consecutive spontaneous abortions before 10th week

LAB criterias include:

- Lupus anticoagulant
- Anti cardiolipin with or without anti $\beta_2$ GPI antibodies at intermediate/high titres on two occasions 12 weeks apart.

For diagnosis one clinical and one lab criteria is required.

Clinical manifestations are as a result of thrombosis of all kind of vessels. Major neurological manifestations include stroke, transient ischaemic attacks, multiinfarct dementia, and corticovenous sinus thrombosis. The prevalence of APLA among SLE patients is variable in different geographical regions and ranges from 22% to 61%. High index of suspicion for APLA in young patients with convulsion may prove regarding in timely diagnosis and instituting appropriate treatment thus decreasing mortality and morbidity associated with this syndrome.

**Conflicts of interest:** none

**References:**