Primary Antiphospholipid Antibody Syndrome Presenting with Cortical Venous Sinus Thrombosis

To the Editor,

We report the case of a 22-year-old unmarried female patient with severe diffuse headache, bilateral vision loss for two days, and multiple episodes of generalized tonic–clonic seizures with alteration of sensorium on the day of presentation to the casualty. There is no preceding history of fever, arthritis, or skin rash. There was no past medical history of any chronic comorbid illness. On physical examination, she was unconscious with normal pupillary size and reaction; bilateral fundi showed papilloedema. The neck was supple; Kernig's sign was negative. Flexor plantar response was present bilaterally with no lateralizing sign on neurological examination. Blood investigations including blood counts, hepatic, renal, thyroid, lipid, and glycemic profile were within normal limits; inflammatory markers including erythrocyte sedimentation rate and C-reactive protein were raised. HIV test by Enzyme-Linked Immunosorbent Assay (ELISA), antinuclear antibody and antineutrophil cytoplasmic antibody test by indirect immune fluorescence assay, and urine pregnancy test were negative. Urine and blood culture yielded no organism. Magnetic resonance (MR) imaging of the brain showed parasagittal T2/fluid-attenuated inversion recovery hyperintensity with diffusion restriction and hemorrhagic transformation suggesting cerebral venous infarcts [Figure 1]. MR venogram confirmed superior sagittal sinus thrombosis. Antiphospholipid antibody (immunoglobulin G) level by ELISA was very high (186.6 U/L; N < 12 U/L), done in view of venous thrombosis in a young patient. The patient was treated with subcutaneous low-molecular-weight heparin at a dose of 1 mg/kg twice daily and oral warfarin titrated according to international normalized ratio target value of 2.0–3.0. The patient gradually improved over the next few weeks and was discharged on warfarin and oral antiepileptic drugs.

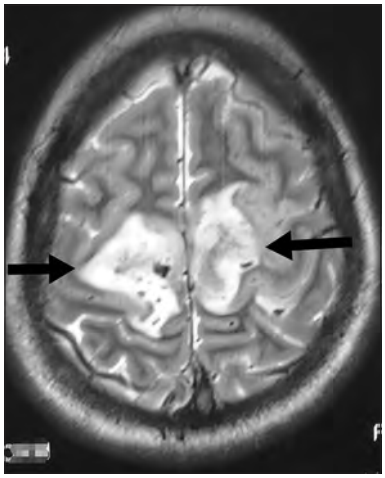

Figure 1: Magnetic resonance imaging brain T2 sequence showing bilateral parasagittal hyperintensity suggestive of cortical venous infarcts

Antiphospholipid antibody syndrome (APS) is a thrombophilic disorder. It usually occurs in young females. It is diagnosed based on the presence of antiphospholipid antibodies.[1] It causes venous or arterial thrombosis and obstetric complications.[1] Venous thrombosis is more common compared to arterial thrombosis in APS. Venous thrombosis commonly presents as deep vein thrombosis in the legs or livedo reticularis in the skin.[1,2] Primary APS presenting with cerebral venous sinus thrombosis (CVT) is a rare complication in primary APS. Central nervous system (CNS) complications usually occur in secondary APS with systemic lupus erythematosus (SLE).[2,3] We excluded other common causes of CVT such as drugs, pregnancy or postpartum state, SLE, and immune-mediated vasculitis by history, examination, and relevant investigations. The treatment of APS mainly relies on anticoagulation and managing CNS complications including seizures and raised intracranial tension, if any. We suggest that APS should be kept in mind while managing a case of CVT, especially in young women of childbearing age to prevent fatal complications.

Declaration of patient consent

In the form the patient has given her consent for her images and other clinical information to be reported in the journal. The patients understand that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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