Lateral sinus thrombosis and haemorrhagic ischemic stroke with protein S deficiency in a young

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ABSTRACT

Stroke or cerebro-vascular disease is one of the most important causes of high morbidity and mortality throughout the world. Stroke in young individuals poses a major problem as these individuals are the earning members of the family. Ischemic strokes are increasingly being attributed to causes other than athero-thrombotic disease .Protein S deficiency is a disorder with increased risk of venous thrombosis. Our patient, 19 years old lady presented with haemorrhagic venous infarction in right temporo-occipito-parietal region with right sided lateral sinus thrombosis.

Keywords: Haemorrhagic venous infarction, lateral sinus thrombosis, protein S deficiency, young lady.

Protein S is a vitamin K-dependent protein synthesized in the liver, vascular endothelium and megakaryocytes, and plays an important physiologic role in the protein C anticoagulant system. This anticoagulant system is one of the major regulators of haemostasis and acts inhibiting clot formation and by promoting fibrinolysis. Protein S functions as a cofactor for activated protein C on the vascular membrane to facilitate the degradation of clotting factors Va and VIIa, downregulating clot formation. Free protein S itself has an anticoagulant effect: it inhibits the prothrombinase complex (factor Xa, Va and phospholipids) that converts prothrombin to thrombin and the tenase complex (factor IXa, VIIIa and phospholipids), which convert factor X to Xa.Therefore, protein S deficiency either congenital or acquired, may lead to serious thrombotic events such as thrombophlebitis, deep vein thrombosis, pulmonary embolism, or an increased propensity to venous thrombosis.[1] We report one case with haemorrhagic venous infarction and lateral sinus thrombosis secondary to protein S deficiency.

CASE REPORT

A 19 years old female presented with headache, vomiting and drowsiness. The headache started insidiously one and half months back and was exaggerated since the day before admission. There was no history of fever, trauma, convulsion, weakness of any limb, facial deviation, diplopia, dysphagia, nasal intonation, nasal regurgitation neither any sensory involvement. There was no similar episode in the past or any of the family members. Patient is not hypertensive, diabetic and hypothyroid. No past history of Tuberculosis or any STD. Patient is married and no history of OCP intake.

General physical examination was normal. On neurological examination, the patient was drowsy, GCS 12/15, plantar non-responsive bilaterally, power 4/5 in all four limbs. Jerks were normal in all limbs. No sensory loss was present. There was no neck rigidity, Kernigs negative. All other systemic examination were normal

Routine haematological examination, liver function, kidney function test, electrolytes and urine was normal. Thyroid function test were normal. Chest X ray, USG, Electrocardiogram (ECG), echocardiogram was normal. CT scan done on the day of admission showed oedema at right temporo-parietal region. CSF study was unremarkable. MRI showed infarct at splenium of corpus callosum, right posterior parietal lobe & right thalamus, sub acute intraparenchymal haemorrhage in right cerebellar and adjacent parietal lobe with mass effect, subacute subdural hematoma in right cerebellar area and ischaemic lesion in right half of midbrain. Then we decided to do MR-venography (MRV) and MR-angiography (MRA). MR-venography showed subacute haemorrhagic venous infarction in right temporo-occipito-parietal lobe with perifocal oedema with right sided lateral sinus thrombosis. MR-angiography report was normal. Her lipid profile, coagulation profile (PT and APTT) ANA (Hep-2), antiphospholipid antibody (IgM and IgG) and homocystein levels were normal. But her protein S activity was low (30%, normal range 55-123%).The concentration of protein C was within normal range.

The patient was given intravenous heparin with warfarin and her clinical condition improved within two days and was discharged in another seven days with oral anticoagulant.

DISCUSSION

Congenital and acquired protein S deficiency is associated with an increased risk of venous thrombosis.[2] The prevalence of protein S deficiency has been estimated to be less than 1 case per 300 in the general population. Two-thirds of patients with a congenital deficiency of protein S (levels less than 50% of normal) may present with venous thrombosis in young adulthood.[3] Stroke in young population has a high incidence of approximately 24–35%, according to some studies in India.[4] Carod-Artal *et al*[5] studied about ischemic stroke subtypes and prevalence of thrombophilia in Brazilian stroke patients. They examined 130 consecutive young and 200 elderly patients. Prevalence of thrombophilia was, respectively: protein S deficiency (11.5% versus 5.5%), protein C deficiency (0.76% versus 1%). They concluded that prothrombotic conditions were more frequent in stroke of undetermined causes. It has been estimated that protein S deficiency occurs in 1 in 3,000 to 1 in 15,000 people.[6]

In this 19-year-old patient without any risk factors, the acquired factor S deficiency possibly.

A role in the stroke and lateral sinus thrombosis. Factor S deficiency should be considered in venous stroke, recurrent pulmonary embolism, unusual site of venous occlusion, family history of vascular events, and stroke in young population. Aetiology of such vascular events in young must be thoroughly investigated so as to guide prevention and treatment of this devastating disease. Measurement of total and free protein S levels should be a part of the evaluation for any young adult who has had a stroke.[7]

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