INTRODUCTION

Stroke in young population has a high incidence of approximately 24 – 35%, according to various studies worldwide. Martinez-Martinez et al. from Spain reported an incidence of 46% in population less than 50 years of age.1 Hypercoagulable states have been reported as an established risk factor for venous thromboembolism and pulmonary embolism but they have also been proposed as a predisposing factor for cerebral ischaemia of arterial origin, especially among young patients. Causes of inherited thrombophilia include factor V Leiden mutation, prothrombin gene mutation, dysfibrinogenemia and deficiencies of protein C, protein S,2 and antithrombin III. To-date, the exact role of protein C and protein S deficiencies in arterial cerebro-vascular occlusion is controversial.

CASE REPORT

An 18 years old girl presented with acute-onset left-sided hemiplegia, bizarre behaviour, irrelevant talks and urinary/fecal incontinence. Past history of the patient revealed recurrent episodes of severe bilateral or sometimes unilateral headache and often, associated vomiting. There were recurrent episodes of focal (left sided) and sometimes generalized tonic-clonic fits in the past as well. No precipitating factors such as drug abuse, head trauma or others were present. Family history was unremarkable for vascular events or other predisposing factors for stroke. General physical examination was unremarkable. Neurological examination revealed that the patient was fully conscious but higher mental functions were disturbed with bizarre and irritable behaviour. She had Wernicke's dysphasia and was repeatedly uttering few sentences. Primitive/released reflexes were present. Motor system examination revealed generalized hypotonia and hyper-reflexia with clonus, absent movements, and positive Babinski's sign on the left side. Rest of the systemic examination was unremarkable.

CT scan of brain showed low attenuation lesion in left temporal lobe with localized loss of gray/white matter configuration and effacement of sulcal pattern-suggestive of acute non-haemorrhagic infarction predominantly involving left temporal lobe. Low attenuation area with loss of cerebral gray/white matter configuration involving cortical, sub-cortical and deep regions of right frontal, parietal, temporal and occipital lobes with relative loss of volume of right cerebral hemisphere and compensatory dilatation of right lateral ventricle, were more pronounced in the temporal horn-suggestive of chronic infarctions, encephalomalacia and vascular compromise in the supply area of right middle cerebral artery (Figure 1). Asymmetrical lateral ventricles with prominent 3rd and 4th ventricles was also observed.
The ultimate diagnosis of combined protein C and S deficiencies was established and the patient was managed with subcutaneous LMW heparin followed by oral anticoagulants. Patient was discharged on oral anticoagulants and physiotherapy.

**DISCUSSION**

Carod et al. studied about ischaemic stroke sub-types and prevalence of thrombophilia in Brazilian stroke patients. They examined 130 consecutive young and 200 elderly patients of thrombophilia with protein S deficiency (11.5% versus 5.5%) and protein C deficiency (0.76% versus 1%) respectively. They concluded that prothrombotic conditions were more frequent in the stroke of undetermined causes.

Clinically, patients with protein C and S deficiency are at an increased risk for venous thrombo-embolic disease, occasional arterial thrombosis, neonatal purpura fulminant and childhood stroke and even portal vein thrombosis. The underlying mechanism of arterial stroke in thrombophilic disorders is yet to be determined. Ischaemic stroke has been reported as a rare manifestation of protein C and/or S deficiency. Girolami et al. and Sie et al. were among the first who reported the association of familial deficiency of protein S as a cause of ischaemic stroke in young. Wiesel et al. studied 105 patients with protein S deficiency, out of whom 14 had arterial thrombotic accidents involving the central nervous system or the myocardium, while most studies revealed a weaker association between the two. Dovay et al. reported that hereditary deficiencies of coagulation inhibitors are rare in ischaemic stroke patients under 45 years and their systematic investigation seems to be of poor interest.

In this 18-year-old patient without any risk factor, the inherited factor C and S deficiency possibly played a role in the recurrent ischaemic stroke. Factor C and S deficiency should be considered in venous stroke, recurrent pulmonary embolism, an unusual site of venous occlusion e.g. portal vein thrombosis, testicular vein thrombosis etc. family history of vascular events, and stroke in young population. Systematic investigation of such causes of cerebral infarctions will reduce the group of undetermined strokes, and will open the possibility of prophylactic treatment in this group of patients.

**REFERENCES**