Case Report

Bilateral Superior Cerebellar Artery Infarcts: Unusual Presentation in Two Patients of Stroke in Young

R Verma*, R Shukla**

Abstract

Stroke or cerebrovascular 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 bread earners of the family. Ischaemic strokes are increasingly being attributed to causes other than atherothrombotic disease. Disorders of coagulation leading to thrombotic disorders are relatively uncommon conditions which are implicated in approximately 1% of all ischaemic strokes and 4-8% of young strokes. Bilateral superior cerebellar infarcts due to hypercoagulable state are extremely rare situations. Here we present two patients with unusual presentation of stroke in young due to hypercoagulable state.

INTRODUCTION

Stroke is one of the most important causes of high morbidity and mortality all over the world. Stroke in young individual poses a major problem as these young men and women are the major bread earners of the family. Community based survey and data from India is lacking. Abraham et al from Vellore, South India reported that 25% of cases of stroke were of less than 40 years of age.1 Other Indian studies have highlighted a high incidence (24-35%) of stroke in young population.2 Here we present two patients with unusual presentation of stroke in young due to hypercoagulable state.

CASE 1

A twenty four years old young male presented acutely with complaints of recurrent vomiting, vertigo, slurring of speech, unsteadiness of gait, difficulty in walking and tendency to veer on either side. One month prior to present episode patient got attack of right sided ataxic hemiparesis which improved after few days of non-specific therapy. Patient was non-diabetic, non-smoker, non-obese and there was no history of hypertension. Family history was negative for vascular events or other predisposing factors for stroke.

General examination revealed pulse rate of 90/min, regular, synchronous with no radio-femoral delay. Bilateral cerebellar signs were present. Motor system evaluation revealed normal power in both upper and lower limbs.

CASE 2

A thirty-five years male presented with acute onset...
Patient was non-diabetic, non-hypertensive and non-smoker. Patient belongs to non-vegetarian only rarely group Family history was negative. General examination revealed pulse rate of 80/min, regular, synchronous and no radio-femoral delay. Blood pressure was 130/80 mmHg with no evidence of postural hypotension. Cardiovascular system, abdomen and chest examination did not reveal any abnormality.

Central nervous system examination revealed bilateral cerebellar signs. Deep tendon reflexes were normally elicitable and planters were bilaterally down-going. Investigations revealed normal metabolic profile. Hb-14.0 gm/dl, random blood sugar 90 mg% and serum creatinine 0.5 mg/d. Liver function tests were within normal limits. Serum lipid profile did not reveal any abnormality. Transesophageal echocardiography did not delineate valvular abnormality or LA clot. Magnetic resonance imaging of brain revealed multiple areas of signal alteration, hyperintense or T₂ and FLAIR weighted images and hypointense on T₁ were noted in bilateral cerebellar hemispheres and vermis. These areas showed evidence of restriction on diffusion weighted imaging which were suggestive of infarcts mainly in bilateral superior cerebellar artery territories. Magnetic resonance angiography revealed faintly visualized bilateral posterior inferior cerebellar arteries. Antiphospholipid antibodies were negative. Thrombophilia profile revealed protein C functional 82.20% of normal (70.00-140.00), protein S
functional 75.00% of normal (77.00-143.00), antithrombin III functional 107.00% of normal (80.00-120.00). PCR-Lipa based venous thrombosis risk profile revealed factor-V Leiden, methy-lenetetrahydrofolate reductase (MTHFR) 677C→T both mutant positive. Serum homocysteine level was 18.76 µmol/l. So this patient again had procoagulant state and presented with bilateral superior cerebellar artery infarcts.

**DISCUSSION**

Before the advent of magnetic resonance imaging (MRI) many cerebellar infarcts running a benign course were not documented. Small lesions, particularly in the lower half of the cerebellum went unrecognized on computed tomography (CT) scans. With the help of MRI, it is now possible to visualize even very small cerebellar infarcts and to delineate their topography in vivo.

Our both patients were diagnosed as posterior circulation stroke due to hypercoagulable state. Hypercoagulable state is a condition in which a clearly identified alteration of the blood tends to shift the haemostatic balance to excess platelets/fibrin deposition and lead to arterial and venous thrombosis in response to vascular injury that would not trigger thrombosis under normal circumstances (Bick and Pegram 1994).³

Our first patient had gross protein S deficiency even at repeat protein S estimation after three months. Ischemic stroke has also been reported as a rare manifestation of protein S deficiency. Most studies carried out to determine the contribution of protein C and S in ischemic strokes have revealed a meager contribution of these factors in young stroke. Ischemic stroke in association with protein S deficiency has been anecdotal with most studies revealing weak association.

Carod-A FJ et al 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 cause.⁴

The importance of thrombophilic disorders in arterial stroke is debated. Some authors find the frequency of hereditary thrombophilic disorders e.g. factor S as frequent in arterial stroke and controls and some authors found as causative factor. Hereditary thrombophilic disorders are to be considered in venous stroke, recurrent pulmonary embolism, unusual site of venous occlusion, family history of venous infarcts, unusual site of arterial occlusion and stroke in childhood, adolescent or early adulthood.⁵ As this patient is 24 years old and has no other risk factor it is possible that the factor S deficiency played a part and implies he will require long-term anticoagulation.

Our second patient presented with bilateral superior cerebellar artery infarct. Factor V Leiden mutant and MTHF reductase mutant variant 677C→T was positive which were double heterozygotes for the two mutations and patient had moderate hyperhomocysteinemia. Ischemic stroke is a frequent heterogenous multifactorial disease that is affected by a number of genetic mutations and environmental factors. Szolnoki Z et al suggested that genetic risk factors which are minor or insignificant when present alone can not only exert an additive effect, but also facilitate the effects of other clinical risk factors at a clinical phenotypic level. There co-occurrence in the same subject can therefore give rise to a highly significant relative risk of an ischemic stroke event.⁶

Kim RJ, in metaanalysis for association between factor V Leiden, prothrombin G20210A and MTHFR C677T mutation and events of the arterial circulatory system found genetic abnormalities specific to factor V, prothrombin and homocysteine metabolism increase the risk for myocardial infarction and ischemic stroke, particularly among younger patients. The individual propensity for arterial and venous thrombosis is likely influenced by differing local mechanisms, systemic mechanisms, or both.⁷

Data are conflicting concerning ischemic stroke risk associated with a common polymorphism in the gene encoding (MTHFR 677CT) which predisposes to hyperhomocysteinemia. Cronin S et al have done meta-analysis regarding association of MTHFR 677T allele with risk of ischemic stroke. Among 14,870 subjects, the pooled
estimated risk of stroke/TIA associated with the 677T allele increased in a dose dependent manner (T allele pooled OR 1.17, 95% CI 1.09 to 1.26, TT genotype pooled OR 1.37, 95% CI 1.15 to 1.64). A graded increase in ischemic stroke risk with increasing MTHFR 677T allele dose was observed, suggesting an influence of this polymorphism as a genetic stroke risk factor and supporting other evidence indicating a causal relationship between elevated homocysteine and stroke.\textsuperscript{8}

These both cases have been reported as bilateral superior cerebellar artery infarcts are quite rare and no case has been reported of bilateral superior cerebellar artery infarct due to procoagulant state. It has to be emphasized that prothrombotic states should be considered in young ischemic stroke patients when common causes of stroke in young has been ruled out.

**REFERENCES**