LETTER TO EDITOR (VIEWERS CHOICE)

CEREBRAL VENOUS THROMBOSIS DUE TO ANTITHROMBIN III DEFICIENCY IN ADOLESCENT

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Key words: Cerebral Sino-venous thrombosis (CSVT), Anti Thrombin III (AT-III)

A 17 years old girl presented with headache & vomiting for 3 days, left sided weakness, drowsiness and altered behaviour for 1 day. Examination revealed hypertension, left side hemiparesis, extensor planters and left sided brisk deep tendon reflexes. MRI and MR venogram showed cerebral superficial and deep venous thrombosis, venous infarcts in bilateral basal ganglia and thalami with hemorrhagic foci in right thalamus, high parietal regions and choroid plexus. Hemogram revealed thrombocytopenia (85,000/cumm) and prolonged partial thromboplastin time. ANA, C3, anti-phospholipid antibodies, thyroid functions, protein S, protein C levels were normal. Anti Thrombin III function was 51% (Normal 80 - 120%). There was no mutation in Factor V leiden. Prothrombin gene (G20210A), MTHFR (C677T) was detected. Child was started on subcutaneous low molecular weight (LMW) heparin followed by warfarin after 48 hours. By day 5 of hospital stay, child had complete resolution of neurological deficits although headache and hypertension persisted till day 14 of hospital stay. Child was discharged on warfarin therapy to be continued for three months and to be followed with a brain scan.

Different pediatric series have shown that children with cerebral sino-venous thrombosis (CVST) have underlying co-morbid conditions and prothrombotic abnormalities in 90% of cases. (1) Antithrombin (AT) III deficiency has been reported as highest risk for venous thromboembolism (VTE) including CSVT in various studies. (2) AT-III deficiency is a heterogeneous disorder, usually inherited in an autosomal dominant fashion. There are different AT deficiencies based on the subtypes. Type I results from reduced synthesis of biologically normal protease inhibitor molecules. Type II is produced by a discrete molecular defect within the protein with normal AT immunologic activity, plasma AT functional activity is markedly reduced leading to risk of thrombosis. Type III is characterized by normal functional and antigenic antithrombin levels but impaired interaction between AT and heparin. (3) Mainstay of treatment due to AT-III deficiency are anti-thrombotics, symptomatic and AT concentrate. Aim of treatment is to recanalise the occluded sinus or vein, to prevent the propagation of the thrombus, and to treat the underlying prothrombotic state in order to prevent venous thrombosis in other parts of the body,and prevent recurrence of CSVT. (4) The goal of treatment for these patients is the initial increase in AT activity to >120% of normal levels followed by maintenance of AT activity at >80% of normal. Anticoagulation is done with LMW heparin, it acts by potentiating the activity of AT-III to more than 1,000 fold by causing conformational change in AT-III. (5)

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