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A 40-month Girl with Cerebral Sino-venous Thrombosis: A Very Rare Case Report

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A B S T R A C T

Background and Aim: CSVT in children and neonates is a rare disorder. The incidence is 0.67 per 100,000 children per year. Risk for CSVT in neonatal period is 2.6 to 12 per 100,000 and after the neonatal period is 0.34 per 100,000. Etiology of CSVT can be a function of intravascular factors (prothrombotic states), anatomic factors (damage to venous sinuses from trauma), and many other thrombotic triggers (infection, inflammation, and dehydration). Prevalence of thrombophilias is higher in children with "idiopathic" CSVT (up to 85%) than in those children with a CSVT provoked by a known clinical risk factor. Clinical Features of Childhood CSVT consist of: headache, nausea, vomiting, irritability, lethargy, encephalopathy, visual disturbance, fever and seizures.

Patient History: A 40-month girl was referred to Mofid Children's hospital with abdominal pain from one-month ago. She had diarrhea and fever 1 week before abdominal pain. In the gastrointestinal ward, she suffered from abdominal pain and rectal bleeding. Before the colonoscopy, thrombocytopenia (Plt=37000) was noticed. Two days after platelet transfusion, she complained of a moderate to severe headache in the right frontal region without any associated signs and symptoms of migraine. On another day, transient paresis first at the right side and after 24hr at the left side appeared. The physicians consulted with us because of her headache and transient paresis. On closer examination- it was found that bilateral increased DTR and alternate paresis.

Results: In the brain CT scan, "Empty Delta", a sign in favor of Cerebrosinovenous Thrombosis (CSVT) in the Superior Sagittal Sinus was seen (figure 1-2). lab tests for thrombophilia assessment were performed (table 1), meanwhile subcutaneous enoxaparin(1mg/kg) and intravenous folinic acid (3mg/kg) were started. In post-contrast brain MRV, the impression was confirmed. 5-7 days later, warfarin (0.2mg/kg) was prescribed and when INR reached between 2-3, enoxaparin was discontinued. Findings based on the lab tests, indicated that Pr S, Pr C, Anti-Thrombin III, Homocysteine levels and APA survey were all normal. But there was a homozygous mutation on MTHFR(A1298c) and a heterozygous mutation on PAI-1 & F13. we continued warfarin for 6 months and oral folinic acid for a long time. Now, the child is regularly followed up.

Conclusion: CSVT in children and neonates is a rare disorder. Neuroimaging (MRV) is very important and has a critical role in the diagnosis of CSVT in these patients. Prevalence of thrombophilias is higher in children with "idiopathic" CSVT than in those with a CSVT provoked by a known clinical risk factors.