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CORRESPONDENCE

Cerebral sinovenous thrombosis in a child with steroid sensitive nephrotic syndrome

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Dear Editor,

Fluss et al. [1] reported 21 pediatric patients with the nephrotic syndrome (NS) and sinovenous thrombosis (SVT). In most of these patients, SVT presented during the first flare or within 6 months of the onset of the NS. Among those 21 patients, only one had SVT at the initial presentation of the NS. Furthermore, only one patient of that series had FV Leiden (heterozygous for a mutation) and presented within 3 weeks of the onset of the NS.

We would like to report a 5-year-old boy with FV-Leiden heterozygous for G1691A mutation, who presented with cerebral SVT followed by the NS. To the best of our knowledge this is the first patient with NS, FV-Leiden and NS presenting with cerebral SVT as the initial symptomatology.

The patient presented with 2 days of headache and vomiting, followed by swelling of the eyelids and legs. There was neither neurological deficit nor neck stiffness. In the laboratory screening, proteinuria was detected. Other laboratory results were as follows: normal blood count serum total protein 4.7 g/dL (N 6–8.7), albumin 2.2 g/dL (N 3.2–4.8), cholesterol 406 mg/dL (N<200), and triglyceride 209 mg/dL (N<200). Protein C, S, antithrombin III, homocysteine, lipoprotein a, and factor II, V, VII, VIII, IX, XI, XII levels were within normal range. Lupus anticoagulant, antiphospholipid, and anticardiolipin antibodies were negative. Prothrombin G20210A and methylenetetrahydrofolate reductase (MTHFR) gene mutations were not found, while FV Leiden heterozygous mutation was positive. Cranial magnetic resonance imaging (MRI) angiography revealed a thrombosis in the superior sagittal sinus and right transverse sinus.

A treatment for NS and thrombosis comprising prednisolone (2 mg/kg/day) and unfractionated heparin (20 U/kg/hour) was started. One week after the prednisolon treatment, full remission of NS occurred. After the patient had received unfractionated heparin for 10 days as an initial anticoagulant therapy, low molecular weight heparin (LMWH) was administered for 2 months. At the end of the 2nd month of diagnosis, cranial MRI angiography did not show any thrombi in dural sinuses. He is currently well and still on low dose prednisolon and LMWH treatments.

Nephrotic syndrome (NS) is a common renal childhood disease. Its relation with hypercoagulability and thromboembolic complications is well-known [3]. Hypercoagulation in NS has been accounted for by the loss of anticoagulant factors with urine, increased platelet aggregation with higher plasma fibrinogen concentration, and insufficient intravascular volume. In children, the incidence of NS associated with thromboembolic complications vary between 1.8 and 5.3% [2]. However, cerebral SVT accompanied by NS is extremely rare with few reported cases in the literature.

In our patient, thrombosis was the initial finding and was followed by NS diagnosis, which suggests that thrombosis may develop even before the full blown clinical picture of NS develops. Furthermore, hereditary thrombotic risk factors, especially FV Leiden mutation, should also be investigated in patients with NS associated with thrombosis because NS patients with FV Leiden heterozygous mutation may be at increased risk for developing SVT at early phases of the disease.

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