

A case report of nephrotic syndrome with hemorrhage of intracerebral in cerebral venous thrombosis

Case Report

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Abstract

Introduction: Cerebral vein thrombosis is a rare complication of nephrotic syndrome (NS). We report a known case of NS with hemorrhagic thrombosis.

Case report: A boy with previous history of NS was admitted with headache and decrease of level of consciousness and his brain images were revealed hemorrhagic thrombosis

Conclusions: Cerebral vein thrombosis must be considered in patients with history of NS (especially in new cases and during of relapses) and prevention of hemoconcentration is very important to decrease thrombosis risk.

Keywords: Children, Nephrotic Syndrome, Thrombosis, Central Nervous System

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Introduction

Nephrotic syndrome (NS) is defined by heavy proteinuria, hyperlipidemia, hypoalbuminemia, and generalized edema. It is a common nephrologic disorder in childhood [1-2]. One of the major problems in this disorder is hypercoagulopathy and increased risk of thrombosis [3-5]. About ten percent of adult and about 5% of children with N.S have a history of thrombosis [6]. The cause of thrombosis is related to an imbalance of coagulations and anti-coagulations factors. The risk is increased by thrombocytosis, hemoconcentration, hyperviscosity, decrease of physical activity, use of corticosteroids and diuretics [7-10]. The risk of vein thrombosis is more than arterial one [11]. The rate of cerebral venous vessels thrombosis is rare complication especially in children. Because of nonspecific sign and symptom of central nervous system (CNS) thrombosis in children with NS, the diagnosis may be occurred with delay. Therefore, any child with NS and CNS manifestations must be considered as CNS thrombosis [12-13]. For this reasons, we report a 12 year old boy with relapse of NS and CNS manifestations with diagnosis of CNS thrombosis.

Case presentation

An 8 year old boy with history of nephrotic syndrome since 3.5 years old has presented with generalized edema, several times vomiting, decrease of urine output, fever, headache (since 3 days ago) and decrease level of consciousness. At the time of admission, his temperature and blood pressure were 38.5^oC and 70/105 mmHg, respectively. He had abdominal tenderness and pitting edema of lower extremities. He was on remission one week before admission, but after an upper respiratory infection, urinalysis was showed 3 + protein and then treatment with prednisolone was started.

After admission, urinalysis showed 3+ protein, blood urea nitrogen and creatinine were 20 mg/dL and 0.58 mg/dL, respectively. He had hemoconcentration (hemoglobin: 17g/dL, hematocrit: 52.5%). Serum sodium (136 mEq/L) and potassium (3.8 mEq/L) were normal. PT and PTT were in normal ranges.

He was admitted with impressions of pseudotumor cerebri and bacterial infection. Then antibiotic (cefotaxim) was recommended and serum for correction of hypovolemia was started. The ophthalmologic exam didn't show edema of pupil and he had normal exam. An emergency non-contrast Computed tomography scan (CTS) of brain was done and it revealed hyperdense shadow in posterior fossa and suggested sagittal sinus of hemorrhage or thrombosis (Fig 1). Then Magnetic resonance imaging (MRI) of brain was performed and showed left occipital sinuvenous thrombosis (Fig 2).

The patient had no neurologic focal signs and there was not any abnormal neurologic exam. The conservative management was done with correction of water and electrolytes continue of antibiotics. Dose of dexamethasone was decreased to 4mg/kg/BID and conservative management was done for him. He did not have any increase in creatinine in period of admission and his headache was alleviated after two days of admission. He was well one week after admission and discharged with neurologic and nephrologic flow up.

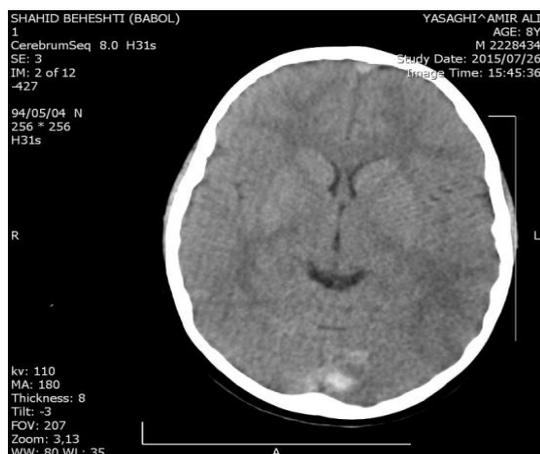


Fig 1: Brain CTS of patient with hemorrhagic thrombosis and nephrotic syndrome

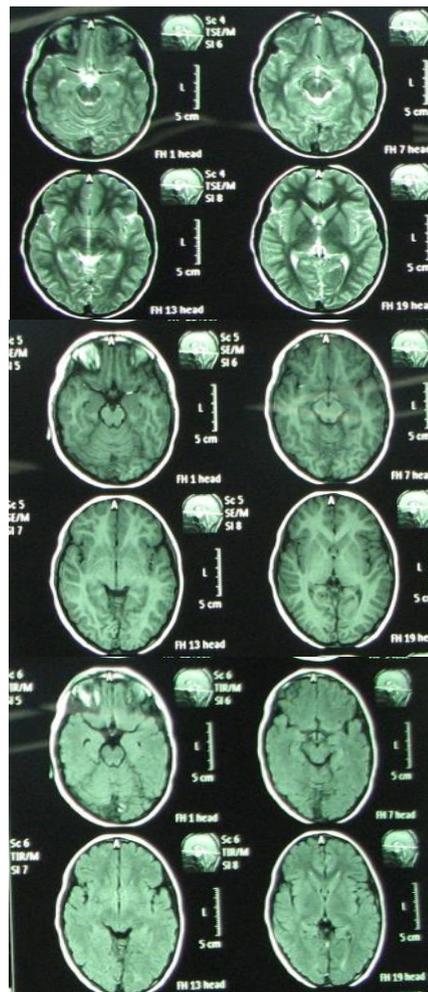


Fig 2: Brain MRI of patient with hemorrhagic thrombosis and nephrotic syndrome

Discussion

Decrease of intra vascular volume due to edema of nephrotic syndrome, vomiting and fever may causes of hemoconcentration (hemoglobin: 17 gr/dL) in this patient and increase the risk of thrombosis. Thrombosis of cerebral vessel is very rare in nephritic syndrome especially in children [13]. The risk of thrombosis is higher in new cases and during relapse for decrease of intravascular volume [14]. The other factor suggested to increase the risk of thrombosis is hypercoagulability [13, 15-16]

Decrease serum level of low molecular weight coagulation factors (factors IX, XI) resulting from increase urine loss has been reported [16-17]. Of course, serum level of high molecular precouagulation factors (factor V, VII, VIII, X) and fibrinogen were increased. The low plasma level of antithrombin III more than 70% of normal was reported by Kauffmann [13]. This

factor has a urinary loss and its plasma level may be related to serum albumin level. Increase platelet activation and then thromboembolic complication have been reported in children with new cases or relapse of nephrotic syndrome [18-19]. The other factor that was suggested to play of role in increase thrombosis is protein Z (PZ). PZ is a single chain vitamin k – dependent glycoprotein. Therefore, PZ deficiency was reported in nephrotic syndrome and might increase the risk of thrombosis. [20-22]

The clinical presentation of cerebral venous thrombosis may be different in sign and symptoms. Our patient had low level of consciousness and severe headache. However, he did not have neurological exam abnormality that probably may be due to small lesion. He had occipital lobe involvement that was very small.

Central vein thrombosis may be seen with early diagnosis and treatment [23-24]. The first investigation is CTS and then MRI and MRV (Magnetic Resonance Venography) [25].

In our patient, the CTS indicated hyperdense lesion in posterior sagittal sinus and thrombotic hemorrhage in left occipital lobe. In addition, in his brain MRI, a small thrombotic hemorrhage was seen in occipital lobe (fig 1, 2). Our patient was treated by dexamethasone and correction of hypovolemia and his condition was better and he was discharged without any neurologic deficit. There is controversy on anticoagulant therapy in children with NS and thrombosis [9]. Heparin is recommended for these patients and may be safe in children [26]. Because antithrombin III is decreased via urinary loss and increase catabolism, the replacement of these factors may be helpful [27].

In conclusion, cerebral vein thrombosis must be considered in any patients with the history of NS and especially in new cases and during of relapses, and the prevention of hemoconcentration is very important to decrease the thrombosis risk.

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