ORIGINAL RESEARCH PAPER
Volume - 10 | Issue - 05 | May - 2021 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

CAVERNOUS SINUS THROMBOSIS IN A CHILD OF NEPHROTIC SYNDROME: A RARE AND LIFE THREATENING COMPLICATION

Paediatrics

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ABSTRACT

Thromboembolic complications are well-documented but rare complications of nephrotic syndrome (NS). NS is a hypercoagulable state associated with venous and rarely, arterial thromboembolism with an estimated risk of up to 1.8–5%. Out of all thromboembolic complications , pulmonary thromboembolism is more than cavernous sinus thromboembolism. Incidence of cavernous sinus thromboembolism is 1.5 per 100000 cases . Here, we describe the subtle presentation of a cavernous sinus thrombosis in a boy with steroid-dependent NS with previous history of massive pulmonary embolism.

KEYWORDS

Nephrotic Syndrome ,Raised ICT , MR Angiography , Cavernous Sinus Thrombosis.

INRODUCTION

Nephrotic syndrome (NS) is a primary glomerular disease. The earliest recorded description of NS was from the 15th century. It was used to describe a major classification of bilateral renal disease. Now, NS is recognized as a common chronic illness in childhood.It is a glomerular disease characterized by altered permeability of the glomerular capillary wall to macromolecules resulting in heavy proteinuria , hypoalbuminemia , hypercholesterolemia , lipiduria and edema. It is characterized by proteinuria defined by urine protein >3.5 g/24 h or urine protein-creatinine ratio >2 mg protein/mg creatinine, edema,hyperlipidemia (>200 mg/dl), and hypoalbuminemia. Hypoalbuminemia causes increased production of coagulation factors in the liver which leads to a hypercoagulable state and an increased risk of thrombotic events, including pulmonary embolism (PE) and cavernous sinus thrombosis[1,2].NS is a hypercoagulable state associated with venous and rarely, arterial thromboembolism with an estimated risk of up to 1.8–5% [3,4]. It involves abnormalities in fibrin formation and platelet aggregation, vascular stasis, loss of antithrombin III, Protein C and S in urine, and increased production of fibrinogen [5]. Thromboembolic events can occur in both venous and arterial territories. The more frequently involved are the cerebral venous sinuses and middle cerebral arteries, deep veins of the lower limbs, vena cava, renal and hepatic veins, pulmonary, and mesenteric artery [6]. Cerebral venous sinus thrombosis in children is a rare complication and only few cases have been reported . As in the present reported case patient presented with symptoms like headache , vomiting , drowsiness , altered behavious .Sometimes the presentation may be very non specific. The CVST should be suspected in any patient with nephrotic syndrome who develops any neurological symptoms [15-17]

CASE REPORT

A 10-year-old male child who was a known case of Nephrotic syndrome.He was diagnosed with NS at 2 years and had started with prednisolone. Since then, the patient had three episodes of relapse , last relapse was 10 days prior to present admission and was started on Tab Omnacortil 20mg at 2mg/kg/day and Tab levamisole 2.4mg/kg/day . Patient also had massive pulmonary embolism one year back for which he was started on low molecular weight heparin for a period of three months and was then stopped .

Now patient presented to our department with complaints of headache and vomiting since one day . Headache was sudden in onset , progressed from left frontal region to entire left half of face associated with photophobia . Vomiting was projectile in nature , 12-14 episodes in one day , containing food particles , non blood tinged , non bile stained. On examination , patient was vitally stable and had mild edema of lower limbs . Detailed systemic examination was done . On Neurologic examination , child was conscious , alert , cooperative , cranial nerve examination was normal , motor examination revealed normal tone and power but reflexes were brisk in all four limbs . Ophthalmic examination was done to rule out signs of raised ICT and it showed no signs of papilledema .

Detailed work up was done. Blood investigations and MRI brain and MR angiography were done simultaneously in view of headache and brisk reflexes . MRI brain showed complete left sided cavernous sinus thrombosis. Blood investigations showed haemoglobin of 13.5gm/dl , hematocrit of 40.1% , total leukocyte count of 9800/mm3 and platelet count of 2.2L/mm3 . Coagulation profile was done which showed prothrombin time of 52.57 and APTT of 72.80 . D-Dimer was done to rule out thrombosis since child had previous history of massive pulmonary embolism which was >4000 .

Symptomatic treatment was given . Child was immediately started on low molecular weight heparin . Anticoagulation with low molecular weight heparin was started and was asked to continue for 3 months at 1mg/kg/day dose BID and was asked to follow up every 2 weeks initially for first two months followed by once a month .

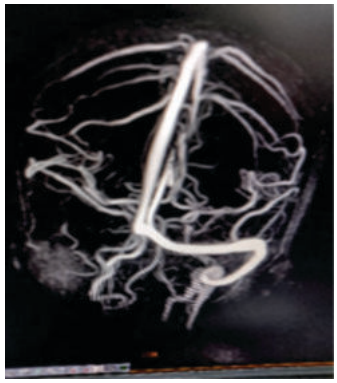

Fig1:Mr Angiography Showing Thrombosis Of Left Side Cavernous Sinus

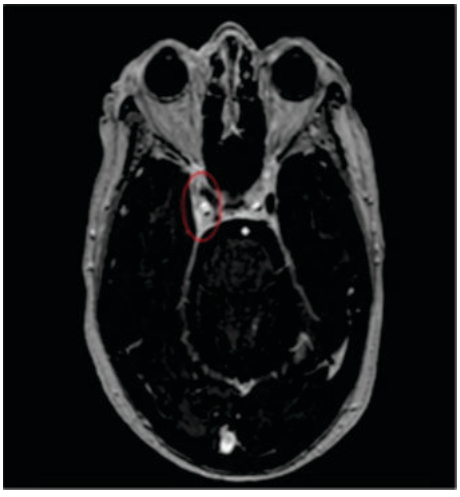

Fig 2 : MRI Brain Showing Left Sided Cavernous Sinus Thrombosis

DISCUSSION

Patients suffering from NS are at a significantly increased risk of thrombosis, with complication rates reported as high as 40% in adults [7]. Thrombosis risk is apparently lower in nephrotic children (18–50%), but events can be much more severe [8].However, these complications in childhood are actually under-diagnosed.

Thrombotic events generally require three basic conditions to be fulfilled : vascular endothelial injury , slow blood flow or stasis and a hypercoagulable state . The pathophysiology of NS involves increased glomerular permeability to large molecules (albumin) which results in loss of albumin through the glomerulus . This leads to hypoalbuminemia which in turn causes the plasma colloid osmotic pressure to drop , producing movement of water from the blood to the tissues . This then decreases the circulating blood volume and leads to increased concerntrations of blood coagulation factors . Concurrently , to combat the hypoalbuminemia the liver increases the production of albumin along with other substances (except albumin) , which results in an imbalance between pro and anticoagulant factors triggering in a thrombosis . [9,10]

Cerebral sinovenous thrombosis is a rare disorder in children , but increasingly diagnosed now because of greater clinical awareness , sensitive neuroimaging techmiques . CVST in nephrotic syndrome is associated with a hypercoagulable state arising due to alteration in blood levels of various factors involved in coagulation ,fibrinolytic system , alteration in platelet fundtion , hemoconcerntration , increased blood viscosity and possibly administration of steroids and diuretics .[14]

In the present case , patient had a previous history of massive pulmonary embolism during a relapse and now presented with symptoms of raised ICT. Although plasma D-Dimers have little evidence in children , the high levels of D-Dimers are markers for thrombosis and provide an important clue to the diagnosis .[11] in the present case , elevated D-Dimers were observed and MR angiography established a definitive and reliable diagnosis of CVST .

The present case was managed by administration of low molecular weight heparin at the dose of 1mg/kg/day BID for a period of 6 months. In patients with a recurrent but reversible risk factor for thrombotic events , it is recommended to use prophylactic anticoagulants for atleast 6 months and also during future relapses [13]

CONCLUSION

Cavernous sinus thrombosis is a known but rare complication of NS in children. The signs and symptoms of cerebral venous thrombosis are occasionally non specific making clinical diagnosis difficult . Hence , a high degree of suspicion is required and a diagnosis of CVST should be considered in any child of nephrotic syndrome presenting with features of central nervous system involvement. Advanced radiological techniques help in confirming the diagnosis. Early aggressive heparin therapy followed by oral anticoagulants is necessary for a favorable outcome.

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