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SCIENTIFIC LETTER

Nephrotic Syndrome Associated with Multicystic Kidney Disease with Concomitant Cerebral Venous Thrombosis

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To the Editor: Nephrotic syndrome is known to be associated with various renal malformations, renal cystic diseases being one of them. Autosomal recessive polycystic kidney disease (ARPKD) and autosomal dominant polycystic kidney diseases (ADPCKD) are reported to be associated with nephrotic syndrome both in adults as well children [1].

We report an 18-mo-old girl who presented with generalized swelling for 5 d. Child was irritable, had generalized pitting edema. Child was diagnosed as nephrotic syndrome based on hypoproteinemia (total protein 3.88, albumin 1.37), hypercholesterolemia (serum cholesterol 352 mg/dl, triglycerides 398 mg/dl), though Up: Uc ratio on day 1 was 0.028 (probably because of severe hypoproteinemia). USG abdomen showed left cystic renal disease with right sided enlarged kidney with cystic changes. Child was started on oral prednisolone and albumin infusion, after which she started having 3+ proteinuria. On day three, child became more irritable, lethargic (Glasgow coma scale- $E_3V_5M_5$) and had one episode of generalized tonic clonic seizure. Child was loaded with phenytoin. Fundus examination revealed bilateral papilledema. Urgent MRI brain with venogram was done to rule out intracranial infection and cerebrosinovenous thrombosis (CSVT) and it demonstrated complete thrombosis of all major dural venous sinuses. MRI abdomen showed multiple non-communicating cysts and minimal intervening renal tissue suggestive of multi cystic kidney disease (MCKD).

She was started on enoxaparin; adequate hydration was maintained with albumin and intravenous fluid. Child started improving from 3rd day onwards and was shifted to oral anticoagulants. She went in to remission in second week and was discharged on warfarin and steroids. She is doing well without any neurological complications.

The case highlights associated renal anomalies with nephrotic syndrome. In pediatric patients, 4 cases have been reported with ADPCKD and nephrotic syndrome. Out of them 3 were steroid sensitive and 1 child had steroid resistant nephrotic syndrome (SRNS) [1–3]. CSVT is a well known complication in nephrotic syndrome because of hypercoaguability due to imbalance between thrombotic and antithrombotic mechanisms [4]. High index of suspicion of CSVT in nephrotic syndrome, can recognize it early and prompt therapy can be started which will reduce further complications and lead to better outcome.

Compliance with Ethical Standards

Conflict of Interest None.

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