nefrología
Revista de la Sociedad Española de Nefrología
www.revistanefrologia.com

Letter to the Editor: Clinical experience

Two children with steroid-responsive nephrotic syndrome complicated by cerebral venous sinus thrombosis

Dos niños con síndrome nefrótico sensible a corticoides complicado por trombosis de seno venoso cerebral

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ARTICLE INFO

Article history:
Available online 21 July 2015

Dear Editor,

Nephrotic syndrome (NS) is a common disorder of childhood. The risk of thromboembolic (TE) phenomena in children with NS is estimated as 1.8–5% with a higher incidence in steroid resistant forms.[1,2]

Venous thrombosis is the most frequent type of thrombosis in childhood NS and occurs mainly in the deep venous system.[3] Kerlin et al.[4] reported 39 TE events among 326 NS patients and 29 of them were deep venous thromboses majority of which were seen in lower extremities. Although infrequent other sites like the cerebral venous system may also be affected and this presentation carries increased morbidity and mortality. Herein we present two children with minimal change disease (MCD) who experienced thrombosis of the cerebral sinovenous system.

Patient 1

A 15-year-old boy was admitted to our clinic with edema and 10 kg weight gain in a month. Physical examination (PE) showed generalized edema and ascites. Urinary examination showed 246 mg/m^2/h proteinuria. Serum albumin level was 0.8 g/dl, cholesterol and triglyceride levels were 375 mg/dl and 190 mg/dl respectively. Hemoglobin (Hb) level was 16.2 g/dl. Renal biopsy was compatible with MCD. He was put on 60 mg/day prednisone treatment and remission was achieved on the 22nd day. Steroid was used as 60 mg/day for a month and tapered over 3 months. The day after cessation of prednisone generalized edema, headache and vomiting started. Laboratory investigations showed severe proteinuria, 1060 mg/m^2/h and a serum albumin of 0.8 g/dl. His Hb

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http://dx.doi.org/10.1016/j.nefro.2015.06.001

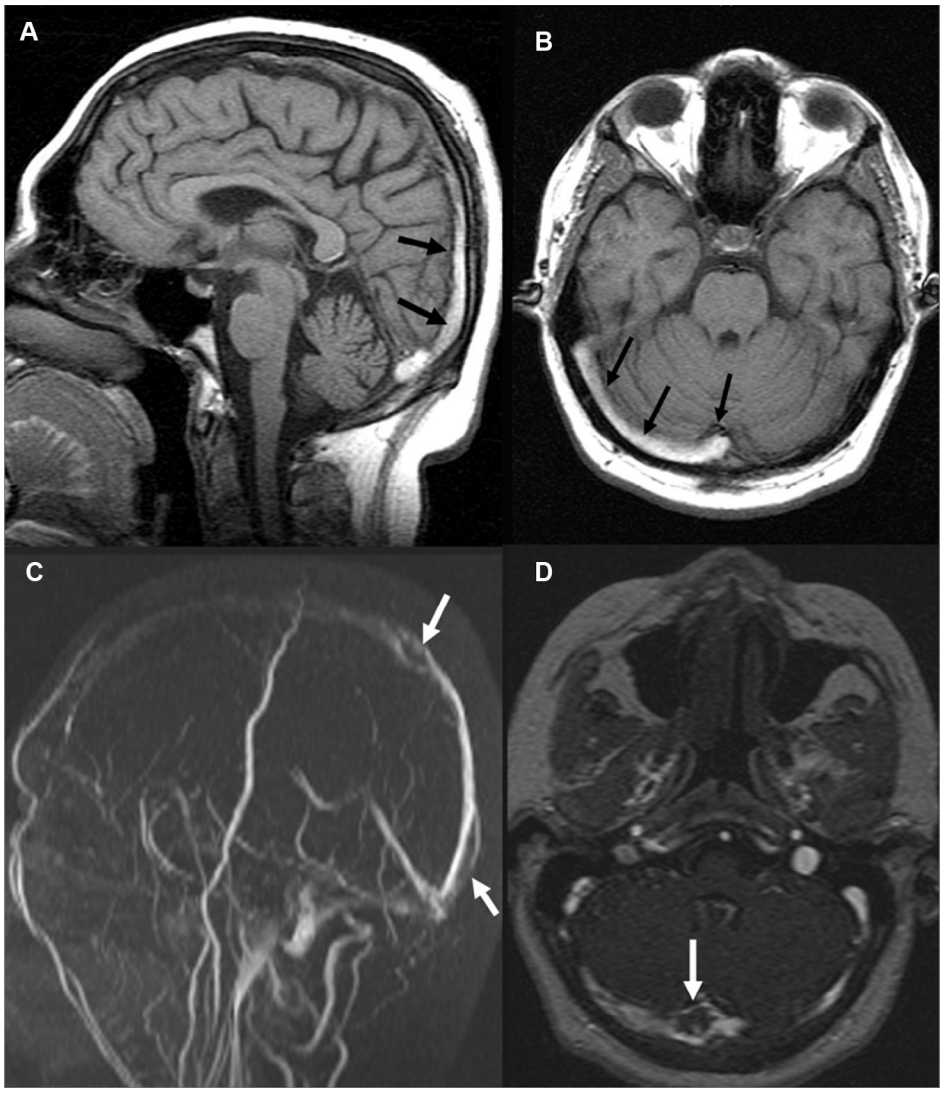

Fig. 1 – Cranial MR on admission; superior sagittal sinus (A) and right transverse sinus (B) thrombosis (arrows). Follow-up MR angiography (C, D); sinuses are partially recanalized with persistant hypointense thrombotic filling defects (arrows).

level was 21.6 g/dl and hematocrit (htc) 63.8%. He was put back on 60 mg/day prednisone and intravenous fluid, albumin replacement, acetylsalicylic acid therapy were started. Cranial computed tomography (CT) was normal. Next day PE revealed lateral gaze palsy of the right eye. Cranial magnetic resonance imaging (MRI) and MR venography (MRV) were performed which showed subacute superior sagittal, right sigmoid and transverse sinus thromboses (Fig. 1A and B). Immediate anticoagulation with heparin was started. Investigations for inherited thrombotic risk factors were negative. On the third day of admission lateral gaze became normal. Two weeks later a repeat MRV showed partial recanalization of the thrombi (Fig. 1C and D). Remission of NS was achieved on the 35th day of steroid treatment, then the dose was decreased and the patient was put on cyclosporine. He had complete neurological recovery and did not experience recurrence of NS or thrombosis during the two-year follow-up.

Patient 2

A 2.5-year-old boy with no significant past medical history presented to our clinic with puffiness of the eyelids, edema of legs and decreased urine output. He had gained 2 kg of weight in a week time. Physical examination revealed facial and pitting edema on extremities in addition to abdominal distension. Laboratory investigations showed Hb 14.9 g/dl, htc 45.9%, platelet count 406,000/μL, total protein 3.7 g/dl, albumin 1.4 g/dl, serum cholesterol 382 mg/dl and normal renal function tests. Urinary examination revealed heavy proteinuria; 244 mg/m^2/h. Serum C3 and C4 levels were normal. Nephrotic syndrome was diagnosed and the patient was put on 2 mg/kg prednisone treatment. On the 4th day of treatment the patient complained of headache. Cranial CT was normal. On the 5th day he developed right 6th cranial nerve palsy. Magnetic resonance imaging and MRV showed thromboses in the