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Case Report

Posterior Reversible Encephalopathy Syndrome in a Child with Steroid Sensitive Nephrotic Syndrome

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Abstract

Introduction: Posterior reversible encephalopathy syndrome (PRES) was described in adults more than children. It was reported in cases of nephrotic syndrome which were mainly on immunosuppressant medications or had severe hypertension.

Case report: We report the case of a 9-years old girl who presented with nephrotic syndrome and moderate hypertension. Nine days after the initiation of steroid therapy she developed disturbed level of consciousness and generalized tonic clonic seizures. Her blood pressure was 145/90 mm Hg and she had normal serum creatinine and electrolytes levels. She was treated with furosemide and convulsions were controlled. After regaining consciousness, the patient complained of loss of vision. Fundal examination was normal. Magnetic resonant imaging, axial FLAIR images and diffusion weighted imaging showed hyper-intensity signal in the parieto-occipital areas. Magnetic resonant arteriography and spectroscopy excluded ischemic insults and neoplastic process. She regained full consciousness and normal vision and was discharged from the ICU four days later. PRES was diagnosed based on the typical pattern of brain imaging and the reversibility of symptoms. .

Conclusion: Nephrotic syndrome in children should be considered a risk factor for developing PRES even without the use of immunosuppressant agents or high doses of steroids.

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Keywords: Nephrotic Syndrome; Posterior Reversible Encephalopathy Syndrome; Steroids

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Introduction

Posterior reversible encephalopathy syndrome (PRES) was described mainly in adults; it has been reported less frequently in children. The imaging pattern is typically seen in patients who develop eclampsia or cyclosporine/FK-506 neurotoxicity after transplantation, but other associations have been reported. These include autoimmune disease (such as SLE or Wegener granulomatosis) and hypertension. The cause of PRES is controversial and unproven [1].

In nephrotic syndrome, hypertension and the administration of calcineurin inhibitors, including cyclosporine, were considered to be the two principal risk factors for developing PRES [2]. We report a case of PRES diagnosed in a patient with steroid responsive nephrotic syndrome, without the use of immunosuppressant agents.

Case report

We report the case of a 9-years old Egyptian girl who presented with generalized body edema, proteinuria of 104 mg/m^2/24 hr, low serum albumin of 2.1 mg/dl and high serum cholesterol level of 432 mg/dl.

She was diagnosed to have nephrotic syndrome. Nine days after the initiation of steroid therapy at a dose of 2 mg/kg/day, the child developed disturbed consciousness level and generalized tonic clonic convulsion. She was not febrile and had no clinical or laboratory evidence of infection. Her blood pressure was 145/90 mmHg, serum albumin was 2.7 g/dl and urinary protein excretion was

Figure-1: MRI axial T2WI image at the level of the lateral ventricles revealing hyper-intensity at the parieto-occipital regions

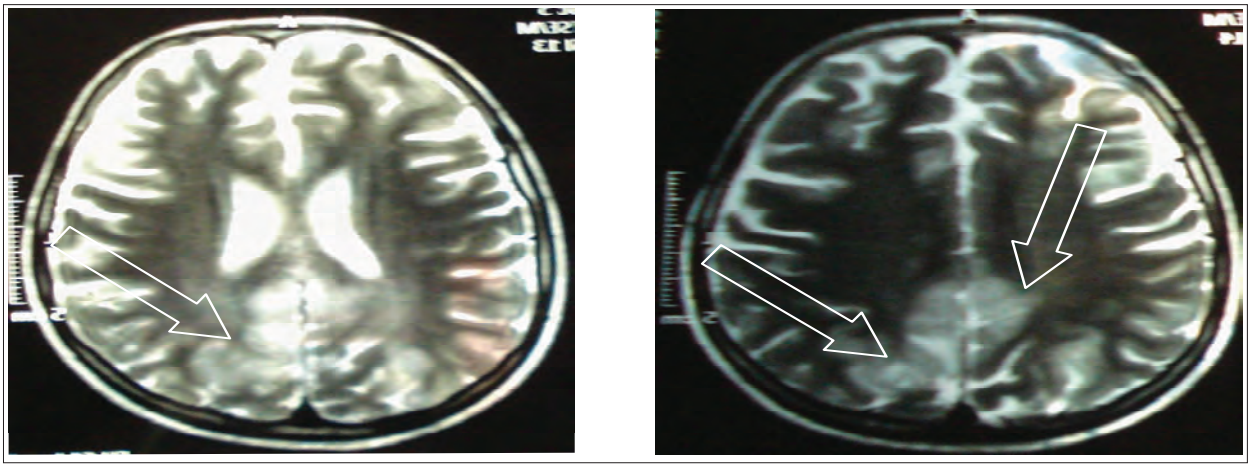


Figure-2: Axial FLAIR image revealing bilateral gyriform cortical and subcortical hyper-intensities at the parieto-occipital regions and subtle hyper-intensity at the right occipital region

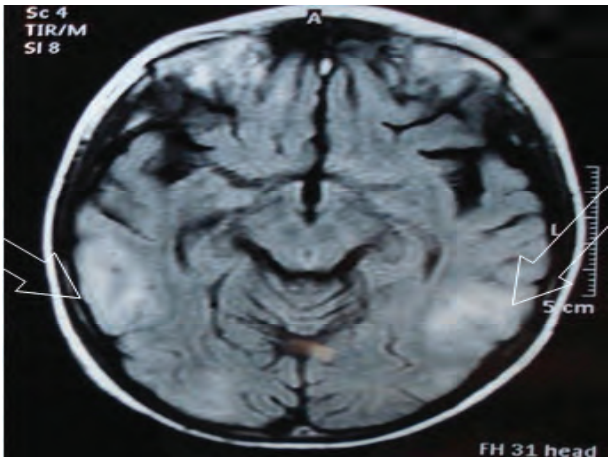
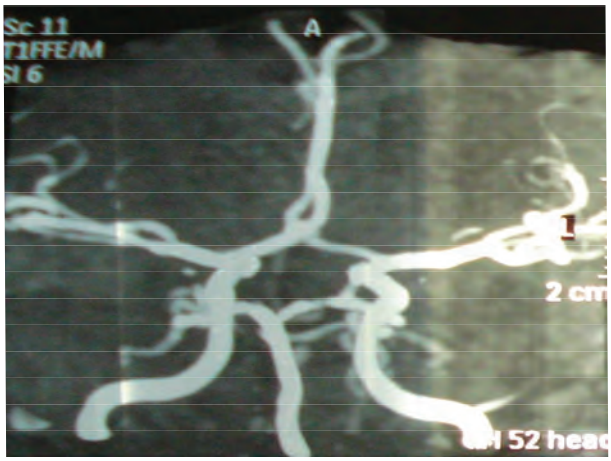


Figure-3: Magnetic resonance arteriography (MRA) with normal display of the circle of Wills



65 mg/m^2/24 hr. She had normal serum urea at 34 mg/dl, normal serum creatinine at 0.7 mg/dl and normal serum electrolytes levels.

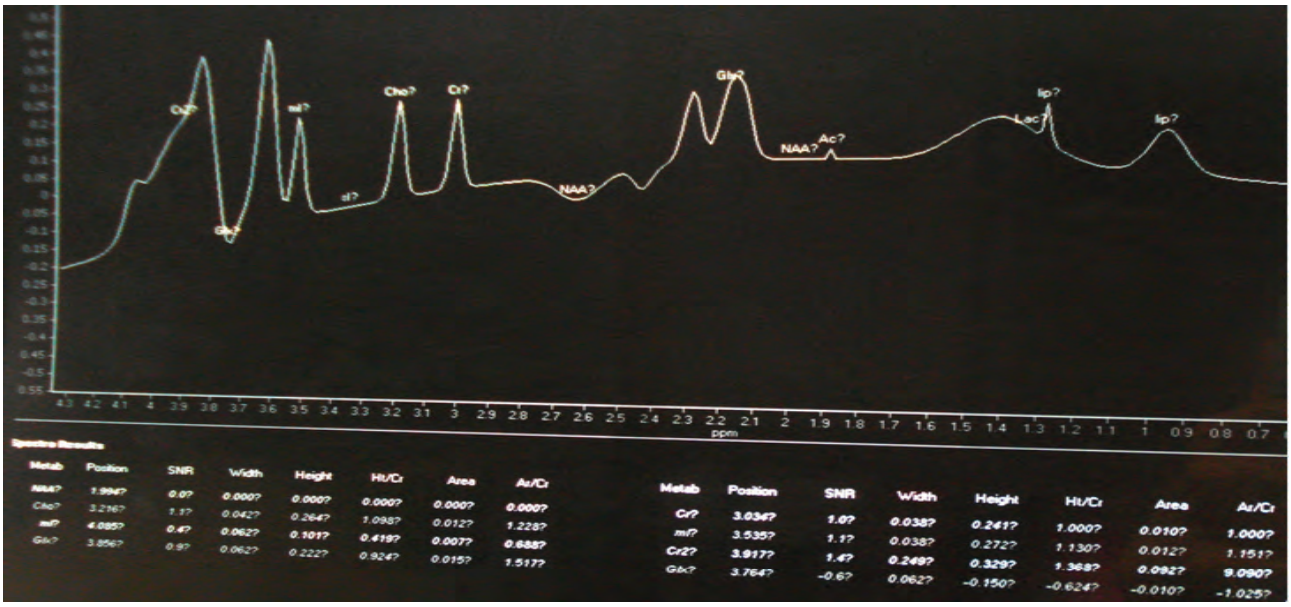
She received emergency antihypertensive treatment; blood pressure and convulsions were controlled after 36 hours. After regaining consciousness, the patient complained of loss of vision. Fundus examination was normal.

Magnetic resonant imaging (MRI) showed areas of hyper-intensity in the parieto-occipital regions Figure-1. Axial FLAIR images revealed bilateral gyriform cortical and subcortical hyper-intensity in the parieto-occipital regions and subtle hyper-intensity in the right occipital region Figure-2.

Based on these imaging findings we suspected ischemia as a complication of nephrotic syndrome. We performed magnetic resonant arteriography (MRA) to exclude arterial occlusion and it revealed normal circle of Wills Figure-3. Magnetic resonant spectroscopy (MRS) was done; it excluded the presence of underlying ischemic insult as there was no evidence of lactate doublet and excluded the presence of neoplastic process by the presence of normal Cho/NAA and Cho/Cr ratios Figure-4.

The patient regained full consciousness and was discharged from the ICU four days later on captopril (1.5 mg/kglday). Her blood pressure was controlled at 100/65 mmHg and she regained her visual acuity. She achieved remission of her nephrotic syndrome after 17 days of starting steroids with urinary protein excretion of 38mg/m2/24 hr and serum albumin level of 3.6 g/dl.

Figure-4: Magnetic resonance spectroscopy (MRS) showing normal MR spectrum with normal Cho/NAA and Cho/Cr ratios



Discussion

PRES was first described in 1996, denoting a reversible, predominantly posterior encephalopathy in patients who had renal insufficiency, hypertension or were maintained on immunosuppressants [3]. Although reversible by definition, secondary complications, such as status epilepticus (SE), intracranial hemorrhage, and massive ischemic infarction can cause substantial morbidity and mortality [4].

Early recognition of PRES is important for timely institution of therapy, which typically consists of gradual blood pressure control and withdrawal of potentially offending agents. This diagnosis should therefore be kept in mind in the differential diagnosis of seizures or coma [5].

Several authors reported PRES in children with nephrotic syndrome but most of them were steroid resistant, were maintained on immunosuppressants or had severe hypertension [6, 7]. PRES was also reported with steroid sensitive nephrotic syndrome in a 21-years old patient [8]. Of note, however, are two patients with nephrotic syndrome who developed PRES with no steroid or immunosuppressant use, and had mild hypertension, reported by Aksoy *et al.* The authors postulate that an increase in vascular permeability related to severe hypoalbuminaemia may be the pathogenesis of PRES in nephrotic syndrome [9].

The report we have here highlights also a potentially important association of PRES with nephrotic syndrome even without the use of immunosuppressant medications.

Conclusion

Nephrotic syndrome in children should be considered a risk factor for developing PRES even without the use of immunosuppressants or high doses of steroids.

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