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POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME SECONDARY TO ACUTE POST-STREPTOCOCCAL GLOMERULONEPHRITIS IN A CHILD: A CASE REPORT

Paediatric Medicine

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ABSTRACT

Background: Posterior reversible encephalopathy syndrome is a clinico-radiographic syndrome of diverse etiologies. Developing hypertensive encephalopathy following post-streptococcal glomerulonephritis is a known but rare manifestation and developing posterior reversible encephalopathy syndrome following it is infrequent. We report a case with magnetic resonance imaging findings of posterior reversible encephalopathy syndrome in the setting of hypertensive encephalopathy and acute post-streptococcal glomerulonephritis. **Case presentation:** A fifteen year old boy presented with giddiness, vomiting and headache for one day and one episode of generalized tonic clonic convulsions at home resulting in loss of consciousness lasting about 10 minutes and developed one similar seizure episode subsequently following admission. He later developed severe hypertension with glomerulonephritis features, which was diagnosed as a manifestation of acute post-streptococcal glomerulonephritis. A computed tomography was done which revealed multifocal hypodensities involving sub cortical white matter of bilateral parietal lobe and occipital lobes. A T2/FLAIR weighted film of magnetic resonance imaging showed bilateral fairly symmetrical homogenous hyperdensity involving cortical and sub cortical regions of frontal,parietal and occipital lobes suggestive of posterior reversible encephalopathy syndrome(PRES). **Conclusion:** Post-streptococcal glomerulonephritis is an important cause of acute nephritic syndrome. This case report illustrates a rare association of posterior reversible encephalopathy syndrome in a patient with post-streptococcal glomerulonephritis.

KEYWORDS

Posterior reversible encephalopathy syndrome, magnetic resonance imaging, Post-streptococcal glomerulonephritis, Hypertensive encephalopathy

BACKGROUND

Posterior reversible encephalopathy syndrome (PRES) is a clinical radiographic syndrome of diverse etiologies which is increasingly identified now. Several medical conditions are delineated to be correlated with PRES and some of these include hypertensive encephalopathy, acute or chronic renal diseases, thrombotic thrombocytopenic purpura, hemolytic and uremic syndrome, vasculitis and porphyrias. Developing hypertensive encephalopathy following post streptococcal glomerulo-nephritis (PSGN) is a known but not a common manifestation in children [1], and developing PRES in such a circumstance is very rare [2].

CASE PRESENTATION

A 15-year-old boy presented to our emergency department with history of giddiness, vomiting and headache for one day and one episode of generalized tonic clonic convulsions at home resulting in loss of consciousness lasting about 10 minutes. On arrival blood pressure was 190/110mmHg ,and AVPU score-V. He developed one episode of similar seizures after admission and developed high grade fever with chills. His sensorium deteriorated from AVPU-P. On further inquiry from the parents, it was noted that he had sore throat and fever 2 weeks prior to admission and were resolved with treatment . He was afebrile for about one week but he had recurrent headaches. He did not had any gross hematuria, dysuria or decreased urine output prior to admission. On the day of admission the patient had developed repeated episodes of vomiting, one episode of generalized tonic clonic convulsions at home and, increased sleepiness. He had transient visual disturbances which he explained as double vision and distorted images. He had mild periorbital swelling, without clinically detectable dependant edema. His throat was examined and there was no signs of inflammation . He didn't had any signs of meningeal irritation. He was admitted to the intensive care unit (ICU) for close monitoring and for further management. In ICU intravenous antihypertensive ,labetalol was used to control the blood pressure and anti seizure medications, leviteracetam were given to control seizures. A computed tomography was done which revealed multifocal hypodensities involving sub cortical white matter of bilateral parietal lobe and occipital lobes. A T2/FLAIR weighted film of magnetic resonance imaging showed bilateral fairly symmetrical homogenous hyperdensity involving cortical and sub cortical regions of frontal,parietal and occipital lobes suggestive of posterior reversible encephalopathy syndrome (PRES) (fig 1,fig 2). His cerebrospinal fluid analysis revealed no inflammatory signs. His urinalysis reported protein-trace and dysmorphic red cells suggestive of hematuria of glomerular orgin. His anti streptolysin O titre (ASOT) reported as more than 200 U/mL which was significant,and urine and the throat swab cultures were sterile. He had normal serum creatinine and electrolytes level. His serum C3 complement levels were low. Diagnosis of acute PSGN was made and he was started on intra venous (IV) penicillin. During the second day of ICU stay his urine output started declining to less than 0.5 mL/kg/h. He was started on IV furosemide and strict fluid balance was maintained. His urine output improved after 48hrs. As cerebral vasculitis ,ischaemic stroke, cerebral venous thrombosis, infectious encephalitis and autoimmune encephalitis were the other possibilities, they were ruled out. Erythrocyte sedimentation rate (ESR), anti nuclear antibody (ANA) and anti neutrophil cytoplasmic antibody (ANCA) were done and found to be negative. After 3 days of ICU stay followed by 2 days of inward stay, patient's blood pressure was well controlled with oral enalapril.His vision and sensorium returned normal. His urine output became normal without diuretics and no clinical or biochemical evidence of acute kidney injury was noted. He was discharged with oral antihypertensive,enalapril and anti-epileptic leviteracetam and was planned to be reviewed weekly in the outpatient clinic. After 5 weeks, his hematuria had resolved on repeat urinalysis and antihypertensive drug was gradually tapered and stopped. He underwent a repeat MRI scan after 4 weeks and was reported as normal.

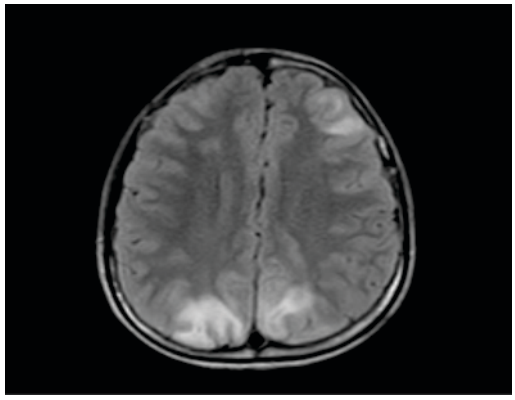

Fig 1 T2/FLAIR showing hyperintensity on the frontal and occipital area

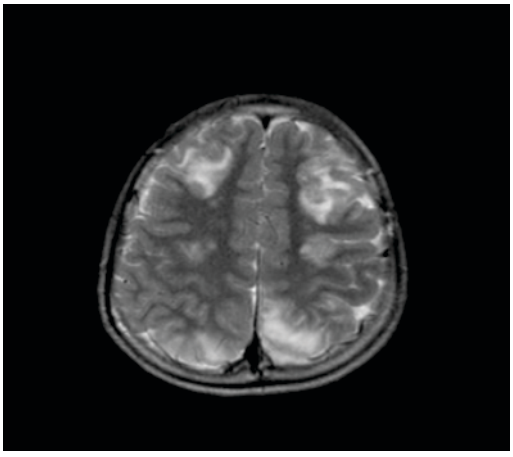

Fig 2- bilateral fairly symmetrical homogenous confluent T2/FLAIR hyperintensity involving cortical and subcortical regions of frontal,parietal and occipital lobes suggestive of PRES

DISCUSSION

The possible diagnosis of PRES subsequent to acute PSGN was arrived based on clinical presentation, laboratory and imaging data. The pathophysiology of PRES remains complex,but most suggested explanations focus on the role of hypertension. Hypertensive encephalopathy has consistently been quoted to explain the mechanism of PRES. Severe hypertension outstrip the limit of cerebro-vascular auto-regulation, leading to cerebral hyper-perfusion and leads to extravasation of fluid into the brain parenchyma leading to vasogenic oedema[3,4]. Children are more likely to suffer from PRES under hypertensive conditions because in children the cerebral blood flow (CBF) auto-regulation threshold is lower than in adults [5].The initial presentation of PRES such as headaches, altered mental status, seizures, and visual loss are correlated with white matter vasogenic edema affecting the posterior occipital and parietal lobes of the brain [6]. There are case reports which shows association of PRES with non-hypertensive patients such as eclampsia, severe infection, immunosuppressive medication, autoimmune diseases and cancer chemotherapy [7,8]. These PRES-related clinical conditions initiates the activation of the immune system, resulting in endothelial dysfunction. Brain biopsy of a PRES patient concluded the evidence of endothelial activation, T-cell trafficking, and VEGF expression[9]. Another study stated that activation of innate immunity is common rather than adaptive immunity in the pathophysiology of PRES[10]. These reports indicates that systemic immune system activation is involved in initiating PRES. In APSGN, the interactions of streptococcal antigens and antibodies induce immune system activation, including complement activation, leukocyte infiltration and cytokine/chemokine production[11]. The immune system, when activated in APSGN, produces tumour necrosis factor (TNF)-α and interleukin (IL)-1β, which upregulates the expression of vascular endothelial growth factor (VEGF), an importantregulator of vascular permeability [11].

Seizures are identified as the most common presenting symptom in the paediatric population, occurring in more than 90% of children [12,5]. Generalized tonic–clonic seizures occur in around 60–75% of patients [3].Retrospective studies suggests that few patients (10–15%) develop recurrent seizures in the first few years after PRES, most of which are because of triggering factors, including recurrence of PRES [13]. There are no prospective randomized studies to guide the anti-seizure medication duration in PRES. It might be appropriate to continue these drugs for several weeks after the initial presentation in patients without residual brain lesions [14].

In general, PRES with APSGN has a commending prognosis if it is identified early and treated.

Past studies showed a full resolution of clinical findings and neuroimaging from 3 to 16 weeks [15].Radiological imaging such as CT and MRI imaging are important in the diagnosis of PRES. The parietal and occipital lobes are commonly affected, succeeded by the frontal lobes, the inferior temporal-occipital junction, and the cerebellum [16,17]. On MRI brain, bilateral symmetrical edema in the parieto-occipital region is hyper-intense on T2-weighted and hypo-intense on T1-weighted sequences are seen [18]. These changers are usually reversible over time.

CONCLUSION

Acute PSGN is a known complication of streptococcal infections and one of the commonest causes of acute nephritic syndrome especially in children, which could be complicated with hypertensive emergencies [20].PRES is not a well described presentation in acute PSGN. The above case report illustrates a rare occurrence of PRES secondary to APSGN in a 15-year-old child. Delayed diagnosis or misdiagnosis may lead to irreversible damage or even death.

Consent

Written informed consent was obtained from the patient's father for publication of this case report.

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