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Posterior Reversible Encephalopathy Syndrome in a Child with Chronic Kidney Disease

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Abstract

A 13-year-old boy, a diagnosed case of stage V CKD due to obstructive uropathy secondary to renal stones. He was having severe uremia and Grade 1- 2 hypertension. After left sided pyelolithotomy, Patient received 6 sessions of hemodialysis, he developed headache, vomiting, depressed conscious level and seizures with cortical blindness. Condition recovered completely clinically as well as radiologically. Clinical and CT brain findings confirmed diagnosis of posterior reversible encephalopathy syndrome (PRES syndrome.)

This condition is relatively under diagnosed in pediatric population. With newer neuroimaging diagnostic modalities increasing reports on PRES syndrome have been described in pediatric and adult population. High index of suspicion need to be kept in mind as early diagnosis and management can lead to complete neurological recovery.

Keywords: Posterior reversible encephalopathy syndrome; Hypertension; Renal failure

Abbreviation: PRES: Posterior Reversible Encephalopathy Syndrome.

Introduction

Posterior reversible encephalopathy syndrome(PRES) is a clinical syndrome, was described in literature first time by Hinchey et al in 1996 [1], Although previously this condition has been described in literature with various other names including reversible posterior leukoencephalopathy, reversible occipitoparietal encephalopathy, hypertensive encephalopathy and reversible posterior cerebral edema [2]. It is a neurotoxic state characterized by neurological symptoms like headache, seizures, and altered sensorium, loss of vision coupled with characteristic potentially reversible findings on clinical examination and on neuroimaging [3]. This condition has been commonly described in adult population especially in association with eclampsia and organ transplantation [4]. It is relatively rare in pediatric population, probably under reported and an under diagnosed condition [5].

Various contributory etiologies for this condition include sudden rise in blood pressure, renal failure, chemotherapeutic agents, Vasculitis, thrombocytopenia, malignancies, immunosuppression, severe hypercalcemia, drugs like cyclosporine, tacrolimus, erythropoietin, interferon, and Henoch–Schönlein purpura. Probably renal failure and sudden rise in blood pressure are the most likely causes contributing towards PRES [2,6].

Hyper perfusion leads to disruption of blood brain barrier mainly affecting posterior circulation, resulting in vasogenic edema but not infarction. This is primarily because of failure of autoregulation and endothelial dysfunction of posterior circulation. It commonly affects Parieto-occipital region in 95% cases, however, PRES can be found in non-posterior regions, mainly in watershed areas like inferior temporal lobe, frontal lobe, thalamus, basal ganglia, cerebellum,

medulla, internal capsule and splenium of corpus calosum [3,7]. Complete neurological recovery is possible in PRES syndrome provided early diagnoses is made and appropriate management is offered in time, otherwise it can lead to irreversible neurological damage, infarction and even death [3].

Case Summary

An eleven years old boy presented with six months history of pain in both flanks, which was intermittent, dull in nature, non-radiating. He had occasional complaints of hematuria, dysuria and fever. His parents noticed that he remained lethargic most of the time, was losing weight and had a poor appetite. Boy was brought through E/R in a gasping state. His pulse volume was normal, rate was 109 beats/min, and BP was 140/90 mm Hg, with moderate pallor. He was having pedal edema, his neck veins were prominent. Liver was enlarged 2.5 cm below costal margin, along with borderline clinical cardiomegaly and bilateral crepitation's on chest auscultation, mainly basal. His growth parameters were below 2SD for his age. His blood glucose was 70mg/dl, Arterial blood gases revealed severe metabolic acidosis with bicarbonate of 8 meq/L, serum creatinine was 9.0mg/dl, blood urea 188mg/dl, serum sodium 133meq/L. His hemoglobin was 6.5gm/dl. His (KUB) Ultrasound of kidney and urinary bladder revealed bilateral severe hydronephrosis, right kidney measured 11cm with cortical thickness of 5 mm, left kidney measured 12 .5cm with cortical thickness of 4mm. Both kidneys had multiple stones ranging in size from 5 mm to 2.5 cm. A 2.5 cm stone was found at left renal pelvis and 1.7 cm stone at right renal pelvis. Ureters and urinary bladder was normal

After initial stabilization, double lumen catheter was passed through internal jugular and hemodialysis was started aiming at 30% urea reduction on day 1, which was increased in duration on day 2, 3 and 4 ultimately achieving 90 % reduction in urea levels. He underwent percutaneous nephrostomy on left side after which His breathing became normal, urine output was 1.3ml/kg/hour, BP was ranging from 125-145 systolic and 80-95 diastolic, for which he was started on oral amlodipine. His serum calcium was 7.0mg/dl, serum phosphate of 6.5mg/dl and intact parathormone was 860ng/ml. serum iron, and ferritin and transferrin saturations were low. His serum uric acid was 4.2mg/dl, serum magnesium was 2.1mg/dl, Urine R/E showed PH of 6.8, pus cells were 8-10/hpf, RBCs were 12-15 and proteins +1. Mid-stream collection of urine was sent for culture which grew E coli >10^5 and he was treated with appropriate antibiotics with renal dose adjustment. Appropriate treatment for anemia, bone disease and acidosis was started.

His DTPA scan revealed obstructive pattern bilaterally with GFR of 15ml/min on right side and 13 ml/min on left side. He was operated for left sided pyelolithotomy, stone was sent for analysis and it was found to be mixed stone consisting of calcium oxalate and ammonium phosphate. His recovery was smooth and he was started oral feed on the first postoperative day. 48 hours after surgery he complained of headache, which was non localized, and dull. Half an hour later he started having blurred vision bilaterally, his conscious level was depressed and he threw a colonic fit of 6 minute duration and became unconscious. His BP was 150/95 mmHg, signs of meningeal irritation were negative; he was afebrile .Fundus examination showed papilledema. He was given IV midazolam and later started on phenytoin, his blood glucose and serum calcium was found to be normal. BP control was optimized by increasing dose of amlodipine, 2 hour later he regained full consciousness level but his vision was lost and could only perceive light.

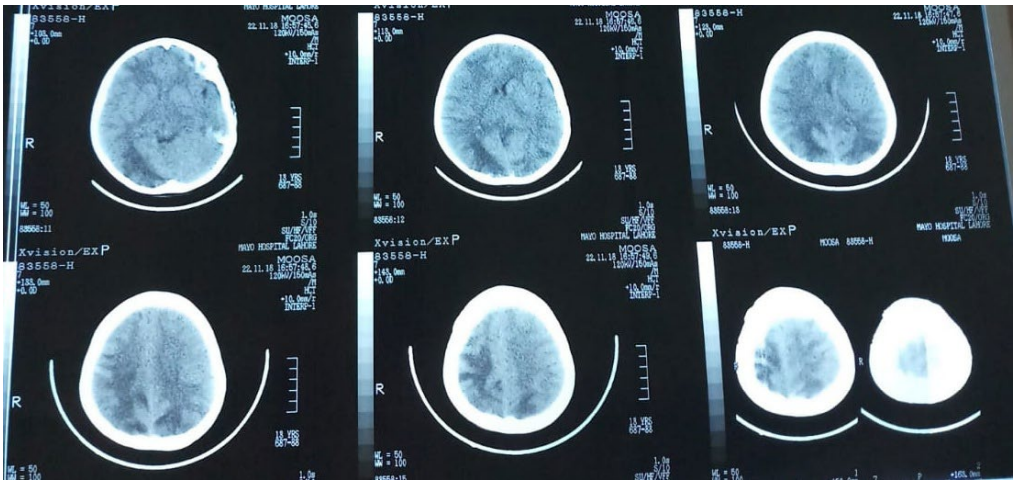

Figure 1: Plain brain computed tomography.

Plain brain computed tomography was performed which showed white matter hypo dense areas with finger like projections in right parieto occipital area and small hypo dense area in left occipital region suggesting PRES (figure 1). Fits were controlled with antiepileptic (phenytoin) and BP was controlled with amlodipine. There were no further episodes of fits, or any deterioration of conscious level. He needed another session of hemodialysis as blood urea was 100mg/dl and serum creatinine was 5.2 mg/dl. Complete recovery of his vision was witnessed within 72 hours. Repeat CT scan at day 10 was normal. Radiological and neurological opinion was sought and diagnosis of PRES syndrome was made.

Discussion

Various factors have been blamed to play a role in PRES syndrome, most likely factors found are hypertension and renal dysfunction, either acute kidney injury or chronic kidney disease[2]. Study by Fugate JE, et al. suggested 38% contribution of CKD and little more than 50% contribution of hypertension [8] Studies have documented irreversible brain damage when mean arterial BP rise of up to >200 mmHg. Uncontrolled hypertension leads to cerebral vessel damage which results in extravasation of proteins and fluid and causes vasogenic edema [9] usually peak systolic BP between 170-190 mm Hg have been documented, however, normal or mild elevation of hypertension have been found in 10-30% of the patients [10].

We used fourth Task Force report on high BP in children and adolescents for staging of hypertension by comparing BP in relation to gender, height and age. BP between 95th till 99th percentile + 5 mmHg was defined as Grade 1 hypertension while BP > 99 percentile +5 mmHg was graded as Grade 2 HTN [11]. Our patient BP ranged from normotensive to grade 2 hypertension.

Symptoms of PRES syndrome reported in literature with varying percentages. Commonest symptom found was seizure 100%, headache 92%, , coma and hypertension 64% each, vision disturbance was found in 46% , severity may range from only blurred vision, to homonymous hemianopia till cortical blindness. Similarly conscious level derangement may vary from mild state of confusion to a deeply comatose state [10]. Conscious level deterioration in our patient began as a state of confusion and then progressed rapidly to a deeply comatose state but got recovered to normal conscious level in the next few hours. Our index case manifested colonic seizures of left half of body with loss of conscious level, however cases with convulsive and non-convulsive status epileptics has also been described in literature [12].

Plain brain computed tomography was performed which showed white matter hypo dense area with finger like projections in right parietoccipital area and small hypo dense area in left occipital region suggesting PRES[figure 1]. Varied pattern of neuroimaging findings have been described in literature which varies from with Holohemispheric water shed pattern, superior frontal sulcus pattern in 59% has been mentioned as “string-of- pearl” appearance. Dominant parietal or occipital in 53% , partial expression in 50%, asymmetric expression in 58%, and partial and asymmetric expression 60%. Focal areas of restricted diffusion or hemorrhage typically remained as residual areas of encephalomalacia [13].

Conclusion

PRES is not an uncommon condition in pediatric age group, a high index of suspicion needs to be kept in mind as timely diagnosis and management can lead to good prognosis.

Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of Interest

None

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