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POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME(PRES) IN CHILDREN

R.V.Dhakshayani MD Asst. Professor of Paediatrics From the department of Paediatric Neurology, Institute of Child Health and Hospital for Children, Madras Medical College, Chennai, Tamilnadu, India.

S. Shobhna MD Senior Resident, Paediatrics Asst. Professor of Paediatrics From the department of Paediatric Neurology, Institute of Child Health and Hospital for Children, Madras Medical College, Chennai, Tamilnadu, India.

C. Leema Pauline MD DM* MD Professor of Paediatric Neurology From the department of Paediatric Neurology, Institute of Child Health and Hospital for Children, Madras Medical College, Chennai, Tamilnadu, India.*Corresponding Author

ABSTRACT

Objectives: To describe the presentation, etiology and outcome of children presenting with PRES in an urban tertiary care centre. **Design:** Descriptive study. **Setting:** Tertiary care teaching hospital. **Method:** This descriptive study was done in all children who had radiologically confirmed PRES recruited between Jan 2016 to Dec 2017 at Institute of Child Health, Chennai. Children.were studied regarding age, sex, symptoms and signs, imaging findings, aetiology and outcome. Results: There were 17 children with radiologically confirmed PRES. Males (65%) predominated the study. Seizures (59%) were the most common symptom followed by headache (47%), altered sensorium (29%), vomiting (24%), and visual disturbances(12%). Parieto occipital region was the common site of involvement. Hypertension was the commonest triggering factor.

INTRODUCTION:

Posterior reversible encephalopathy syndrome (PRES)is a clinico radiological syndrome presenting with neurological symptoms including headache, seizures, altered sensorium, and loss of vision. It was first described by Hinchey *et al*. in 1996.[1] It is thought to represent a disorder of cerebral auto regulation. The disease has been more commonly described in adult population,[2] especially in the setting of eclampsia and organ transplantation. Of late it has been described in children also. Conditions associated with PRES in children include hypertension, renal diseases, hematologic disorders, bone marrow transplantation, drugs like immuno suppressants, chemotherapeutic drugs, monoclonal antibodies etc. Symptoms typically resolve spontaneously or after correction of blood pressure. Herewith we present the clinical symptomatology, neuroimaging findings in a cohort of 17 children presenting with PRES.

SUBJECTS AND METHODS:

The study group comprised of all children who had radiologically confirmed PRES recruited between Jan 2015 to Dec 2016 at Institute of Child Health, Chennai. The data regarding age and sex distribution, clinical symptoms and signs, prior history of PRES, neuroimaging findings, possible cause and outcome were collected and analyzed. Relevant investigations such as renal function tests, 24 hour urinary protein, spot urine protein creatinine ratio, X ray chest, Echocardiogram, ultrasonogram abdomen, C reactive protein, ASO titre, C3 levels, renal doppler, plasma renin activity, plasma aldosterone levels, urine vanyl mandelic acid, were done wherever required.

RESULTS:

During the study period, we came across 17 children with radiologically confirmed PRES. There were 11 (65%) males and 6 (35%) females. Of the 17 children with PRES, 5 (29%) were between one and five years and 12(71%) belonged to more than five years age group.

Seizures 10(59%) were the most common symptom followed by headache 8 (47%), altered sensorium 5(29%), vomiting 4(24%), visual disturbances in 2(12%).Of the ten children presented with seizures, 6 presented with generalised status epilepticus. Acute loss of vision in both eyes was noticed in two children. There were 7 children with a prior history of renal disease, 3with acute lymphatic leukemia. There were no recurrences.

The site of involvement in neuro imaging was parieto occipital in 9, occipital in 4, frontal in 2, parietal in 2. Altogether 15(88%) had findings suggestive of PRES in either parieto occipital or parietal or occipital region. In addition, cerebellum, brainstem, spinal cord were involved in one child each respectively. Intracranial haemorrhage was seen in one child.

Hypertension was noticed in 14(82%) children. The etiological conditions included Chronic Kidney Disease(CKD) in 7(41%), nephrotic syndrome in one. Of the remaining 6 children who were found to be hypertensive, the cause was identified as acute nephritis in 5 and in one child probably idiopathic as all the investigations were negative. Chemotherapy associated with the treatment of Acute Lymphatic Leukemia(ALL) was the offending factor in 3(18%).

Children were managed with supportive therapy including anticonvulsants and antihypertensives. Withdrawal of the offending drug was carried out as indicated. Repeat imaging was done in 10 children which showed resolution of the lesions. All patients had complete resolution of their symptoms.

DISCUSSION:

The mean age of our patients was 7.2 years with a range of 4years to 12 years. Males(65%) predominated the study which is in accordance with other studies.

We noticed that in our patients seizures 10(59%) were the most common symptom followed by headache 8 (47%), altered sensorium 5(29%), vomiting 4(24%), visual disturbances in 2(12%). Gera et al in their study of 11 children with PRES in renal diseases found seizures in ten cases (91%), headache in eight cases (72%), altered mental status in five cases (45%), and dimness of vision or cortical blindness in three cases (27%) in different combinations [3]. Seizure as a presenting symptom was most common in Ishikura's study (85%) also[4]. Status epilepticus frequently presenting with PRES has been described in. patients.[5]. Of the 10 patients with seizures, 6 presented as status epilepticus in our cohort. Only occasionally do patients with PRES show focal neurologic signs, including hemiparesis and aphasia. [6]. We did not find any child with focal neurological deficit in our study population..

Parieto occipital region was the commonest site of involvement in neuroimaging. 88% of the patients had findings suggestive of PRES in either parieto occipital or parietal or occipital region. Predominantly parieto-occipital involvement is usually observed (from 50% to 99% of cases), whereas the frontal and temporal

lobes are affected in half of cases. The cerebellum, basal ganglia, and brainstem are involved in approximately one-third of cases. [7,8]. Cerebellum, brainstem, spinal cord were involved in one child each respectively in our study. CT brain depicting hypodensities in both parieto occipital regions. Repeat imaging after 2 weeks shows complete resolution of the lesions.(Fig 1)

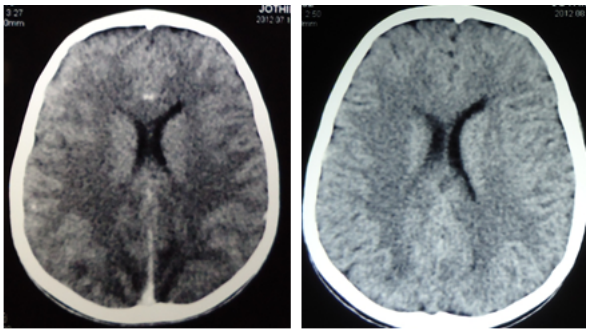

Fig .1. CT brain showing bilateral hypodensities in both parieto occipital regions

Many of the reported articles cite that hypertension, renal diseases, immunosuppression, chemotherapeutic agents for lymphoma and leukemia, severe hypercalcemia, thrombocytopenic syndromes, Henoch–Schönlein purpura, vasculitis, and renal failure as causes for PRES.[9] Chen et al in their study of 14 children with PRES described that their underlying disorders included nephrologic disorders (6), systemic lupus erythematosus(4), malignancies(3),and kidney transplant (1). Precipitating

factors were identified in all patients, including hypertension (100%), immunosuppressant agents (71%), antineoplastic agents(21%), and hemodialysis (14%).[10] We found that renal diseases(76%) as the major cause followed by chemotherapy for leukemia(18%). We also 82% of our patients presented with hypertension. Resolution of lesions was noticed in all cases in accordance with other studies.

CONCLUSION:

Posterior reversible encephalopathy syndrome is not uncommon in children. Hypertension associated with renal diseases was the major triggering factor. Complete recovery was noticed in our cohort.

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