

A Case of Acute Glomerulo Nephritis with Encephalopathy

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ABSTRACT

Encephalopathy describes a clinical syndrome of altered mental status, manifesting as reduced consciousness/altered behavior. It can be caused by many diverse causes, including systemic infections, metabolic derangement, inherited metabolic disorders, toxins, hypoxia, trauma, vasculitis, and CNS infections. We are here presenting an atypical case of Acute Glomerulo Nephritis (AGN) admitted with features of encephalopathy.

Keywords. Encephalopathy, Glomerulo Nephritis, vasculitis.

CASE REPORT

An 8 yr old girl with no significant past history was brought to casualty with c/o fever, headache and vomiting of 1 day duration. She had 3 episodes of generalised tonic posturing (lasting 2 min) over 12hr period, and following which, she developed blurring of vision also. Had h/o pyoderma , 3wks back. No history of decreased urine output, hematuria or oedema. On examination, she was drowsy with a GCS of 13, with tachypnea (febrile) and high BP 136/88 mmHg (99th centile was 122/84Hg) CNS examination had normal cranial nerves, normal tone, normal DTR, had no meningeal signs. Other systems were WNL. Because of pyoderma and high BP, a provisional diagnosis of Hypertensive encephalopathy in the setting of AGN was kept. But the odd thing was that she had no hematuria/ reduced urine output/oedema

INVESTIGATION

Blood investigations had normocytic normochromic anemia, normal RFT, LFT, S.electrolytes, calcium. URE had numerous RBCs.CSF analysis was deferred since the child had cerebral edema. Her S.C3 was low (0.08g/L) and ASO was high (300 IU/ml), strengthening other evidences. Lupus workup was also done and was negative. Imaging was also done in which CT showed features of cerebral edema. MRI brain showed abnormal increased T2 and FLAIR signal intensity involving grey matter and subcortical white matter of bilateral posterior parietal and occipital lobe, bilateral cerebellar hemispheres in a nearly symmetrical manner, and left pre and postcentral gyri with mildgyral swelling and mass effect, no evidence of contrast enhancement, diffuse restrictions. MRI features are suggestive of Posterior Reversible Encephalopathy Syndrome.

TREATMENT

Child was managed with antiedema measures (3% saline and mannitol), antiepileptics (IV Phenytoin) and empirical antibiotics (ceftriaxone). Labetolol infusion was also started following which BP became normal and child regained vision and consciousness within 24hrs. Later BP was maintained on oral labetolol and Nifedipine. Antihypertensives were tapered and stopped, and child was discharged with a normal BP within 2 weeks. She was asymptomatic on follow up. S.C3 was repeated after 3months and was normal.

DISCUSSION

Posterior reversible encephalopathy syndrome (PRES), initially delineated by Hinchey *et al.* in 1996, is a clinical condition presenting with neurological symptoms together with headache, seizures, altered sensorium, and loss of vision, and in characteristic resonance imaging (MRI) findings that are potentially reversible (Hinchey *et al.*, 1996). Over the years, this condition has been delineated by numerous names together with reversible posterior leukoencephalopathy, reversible posterior cerebral oedema, reversible occipitoparietal encephalopathy, and hypertensive encephalopathy (Girişgen *et al.*, 2010). Postulated underlying causes include a sudden increase in blood pressure, immunosuppression, lymphoma, and leukaemia chemotherapy agents, extreme hypercalcemia, thrombocytopenic syndromes, Henoch-Schönlein purpura, vasculitis, and renal failure, the most common of which tend to be sudden rises in blood pressure and renal failure (McKinney *et al.*, 2007). Vasogenic oedema is seen in neuroimaging affecting the white matter of the posterior bilateral cerebral hemispheres, most commonly the parietal and occipital lobes (Hinchey *et al.*, 1996; Girişgen *et al.*, 2010). The prevalence of PRES in children is not well known, but PRES reports have been reported in children following chemotherapy and tumor lysis syndrome and hypertension in children (Prasad *et al.*, 2007).

Although it is not often possible to show a substantial increase in blood pressure, PRES is known to be a type of hypertensive encephalopathy, with a sudden increase in blood pressure being the most often associated characteristic, although other causative factors have also been involved (Mirza, 2006).

Pathophysiology of PRES includes: sudden spike in blood pressure triggering vasospasm and the other being autoregulatory mechanism malfunction (Girişgen *et al.*, 2010). The

autoregulatory ability of the brain vasculature is exceeded with sudden elevations in systolic blood pressure, resulting in a region of vasodilatation and vasoconstriction, especially in the arterial boundary zone. This triggers blood-brain barrier breakdown with subsequent fluid shift along with haemorrhage (Mirza, 2006). The preferential involvement of the posterior circulation was postulated to be comparatively lower in the arterioles supplied by the vertebrobasilar system than in the anterior circulation due to the sympathetic innervation shielding the brain from sudden increases in blood pressure (McKinney *et al.*, 2007). While PRES usually includes the parietal-occipital zone, the first was the holohemispheric watershed pattern with a linear involvement of the frontal, parietal, and occipital lobes mainly along a watershed distribution. Asymmetrical and partial manifestations of the primary patterns were also identified in addition to these patterns (Bartynski *et al.*, 2007). Brain stem, cerebellum, basal ganglia, thalami, internal capsule, and splenium of corpus callosum are atypical sites of participation that have been identified, with uremic encephalopathy often described as having a preference for central distribution (Casey and Truwitt, 2000). Clinical Manifestations of PRES: Typical PRES characteristics include impairment of consciousness, seizure activity, headaches, visual abnormalities, nausea / vomiting, and neurological focal deficiency (Casey and Truwitt, 2000). Consciousness disorder can vary in severity from encephalopathy or coma to depression, somnolence, and lethargy. In all paediatric PRES patients, all these clinical features were not identified. In a study of 25 children with PRES, 44% displayed all four clinical symptoms, 32% displayed three, 16% showed two, and 8% had only one symptom. 12 patients who had seizures (42 %), visual disturbances (33 %), headache (17 %), or altered mental state (8 %) were reported by Kwon *et al.* The most common clinical characteristics such as seizure (6/9), headache (6/9), and altered consciousness (4/9) were detected by Incecik *et al.* Nausea and vomiting were the other signs, and blurred vision (Incecik *et al.*, 2009; Legriél *et al.*, 2011).

DIAGNOSIS

PRES is a clinical-radiological entity. The intensity and severity of its clinical manifestations vary and may require ICU admission. Findings from imaging often differ in severity. For the diagnosis, complete familiarity with the imaging requirements is vital. The diagnosis of PRES was identified by suggestive clinical manifestations along with radiological criteria. In suspected cases, the diagnosis is verified by the clinical and radiological change after

adequate therapy. There are, however, no consensual criteria for PRES diagnosis validation (De Laat *et al.*, 2011)

Roles for computed tomography and MRI in diagnosing PRES: Findings from computed tomography (CT) are often normal or unspecific. Hypodensities indicate PRES in a suggestive topographic distribution. The essential study for the diagnosis of PRES is cerebral MRI. Proton-density and T2-weighted images display edema-indicating elevated signal areas. The lesions are often visualised by fluid-attenuated inversion recovery (FLAIR) sequences. Improvement in the diagnosis of PRES and the identification of subcortical and cortical lesions in PRES has been demonstrated by the application of FLAIR. T1-weighted images show low-intensity focal points. Diffusion-weighted imaging (DWI) is common, but it increases the apparent coefficient of diffusion. Finally, in around half the instances, change is seen. MRI is more appropriate for the diagnosis of PRES than CT (Siebert *et al.*, 2013).

The four radiological patterns of PRES are as below:

- a. Holohemispheric watershed trend (23%): A swath of confluent vasogenic edema spreads across the frontal, parietal, and occipital lobes. There is less apparent involvement of the temporal lobes. This topography matches the region of the watershed between, on the one hand, the anterior and posterior cerebral arteries and, on the other, the middle cerebral artery.
- b. Superior frontal sulcus pattern (27%): Patchy edema, around the superior frontal sulci, predominates in the frontal lobes. There is variable involvement of the parietal and occipital lobes.
- c. Dominant parietal-occipital pattern (22%): The posterior portion of the parietal and occipital lobes are primarily involved in this pattern that was previously considered to be characteristic of PRES. The edema changes from moderate to extensive in severity.
- d. Partial or asymmetric expression of the primary patterns (28%): The partial type is defined in either the parietal or the occipital lobes as a bilateral absence of edema. Sometimes, the frontal lobes are involved. The asymmetric form is distinguished in either the parietal or the occipital lobe by the unilateral absence of edema. Finally, there is both a lack of involvement of either the parietal or the occipital lobes in the partial and asymmetric form, and asymmetric abnormalities in the affected parietal or occipital lobes (Arzanian *et al.*, 2014).

DIFFERENTIAL DIAGNOSIS

Diagnostic problems are increased by non-specific clinical manifestations and the multiplicity of radiological trends. Various conditions can be similar to PRES, including ictal or post-ictal state (with or without epileptic status), infectious encephalitis, progressive multifocal leukoencephalopathy (PML), acute disseminated encephalomyelitis, mitochondrial encephalopathy, lactic acidosis and stroke-like episode syndrome (MELAS), Creutzfeldt-Jakob disease, vasculitis, cerebral sinus thrombosis, and stroke-like episode syndrome (MELAS). In diagnosis, the MRI features of these conditions are helpful (Won *et al.*, 2009)

TREATMENT

The treatment strategy involves general measures to correct the underlying cause of PRES. An early etiologic diagnosis helps the cause of PRES to be promptly corrected. Blood pressure management, removal of cancer chemotherapy or immunosuppressive agents, dialysis, or other treatments may be appropriate. In order to minimise the risk of ischemia or bleeding and thus to avoid permanent injury or death, prompt correction of the cause is necessary. Anticonvulsant therapy is prescribed in patients with seizures, often after a single seizure. As soon as the patient's condition stabilises, transfer of anti-seizure medication to non-hepatic microsomal enzyme-inducing drugs is advised. An anticonvulsant regimen was also prescribed for at least 12 months after the next seizure episode in patients with an irregular electroencephalogram or cranial MRI findings and recurrent seizures. Lucchini *et al.* (2008) advised anticonvulsant therapy for 12 months in patients with brain damage. No consensus has yet been achieved about which patients should receive antiepileptic drugs for seizures after PRES

Author contribution

Kannan N, Prabhu K, Jamunarani A and Thatiparthi Stephen encouraged and supervised the findings of this work. All authors discussed the results and contributed to the final manuscript.

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Study significance

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