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Case Report

CASE REPORT: SUCCESSFUL MANAGEMENT OF PRES IN PATIENT OF CKD

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ABSTRACT
Posterior reversible encephalopathy syndrome (PRES) clinico-radiological syndrome characterized by a headache, seizures, altered mental status. It is also characterized by white matter vasogenic edema affecting the posterior occipital and parietal lobes of the brain predominantly. It is common in hypertensive chronic kidney disease patient. We present a case of 30-year-old young non-hypertensive female suffering from chronic kidney disease who developed PRES.

Keywords: *CKD, Chronic Kidney Disease, PRES, Posterior Reversible Encephalopathy Syndrome, Reversible Posterior leukoencephalopathy Syndrome, PRES in Non-Hypertensive Patient*

INTRODUCTION
Posterior reversible encephalopathy syndrome (PRES) or reversible posterior leukoencephalopathy syndrome is a clinico-radiological syndrome first described in 1996 by Hinchey et al. & is characterized by a headache, seizures, altered mental status, visual loss and white matter vasogenic edema affecting the posterior occipital and parietal lobes of the brain predominantly (Sudulagunta *et al.,* 2017; McKinney *et al.,* 2007; Hinchey *et al.,* 1996). It is almost always but not necessarily associated with acute hypertension (McKinney *et al.,* 2007, Roth and Ferbert, 2011). PRES may be commonly seen in chronic kidney disease (CKD) and acute kidney injury (AKI) patients (Hobson *et al.,* 2012, Fugate et al., 2010). These patients are especially vulnerable to this syndrome, because they are frequently exposed to several of its possible causes, including uremia, hypertension, vascular and autoimmune diseases, exposure to immunosuppressive drugs, and organ transplantation (Hobson *et al.,* 2012; Ergin *et al.,* 2008). It is therefore important to consider PRES in the differential diagnosis of patients with renal disease and rapidly progressive neurologic symptoms. Posterior reversible encephalopathy syndrome is an increasingly recognized disorder, with a wide clinical spectrum of both symptoms and triggers, and yet it remains poorly understood.

CASE
A 30-year-old female admitted from casualty with shortness of breath since 1 month (NYHA class I which progress to NYHA class III); decrease urine output & generalized weakness since 3 days. She was a known case of chronic kidney disease & on conservative management since last 1 year.
There was no past history of hypertension, diabetes mellitus, bronchial asthma & tuberculosis. There was no other significant history.
No history of recent travel to any new place, cardiac disease, acute or chronic blood loss.
On examination – pulse - 90/min regular, normal volume; BP - 130/90 mmHg; pallor +++; anasarca +. No signs of meningeal irritation.
On systemic examination – Her cardiovascular system, respiratory system & neurological system were normal except for bibasal crepts (suggesting pulmonary edema).
She was admitted to MICU (Medical Intensive Care Unit) & all routine investigations were done which was s/o (suggestive of) severe iron deficiency anemia with deranged kidney function with hyperkalemia.

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Hemoglobin – 2.6 gm/dL ; Total Leucocyte Count – 6700/cmm ; Platelet – 98000/cmm ; Mean Corpuscular Volume – 67 fL ; Urea – 365 mg/dL ; Creatinine – 7.65 ; Na (sodium) – 139 mEq/L ; K (potassium) – 6.0 mEq/L.

Ultrasonography of abdomen revealed – small size kidneys with raised cortical Echotexture & loss of corticomedullary junction s/o grade III renal parenchymal disease with moderate ascites.

Patient was managed with 3 unit of packed red cell transfusion, 3 session of hemodialysis, appropriate antibiotics, diuretics, iron & calcium supplement over a period of three days.

During third day of hospital stay, patient had 3 episodes of generalized tonic clonic convulsions, for which CT brain was done, which showed Ill-defined hypo density in bilateral (B/L) occipital, parietal & centrum semiovale in subcortical white matter. Linear blood densities along sulco-gyral space in right high paritel region are also noted. For further information MRI brain was done, s/o – B/L symmetrical hyperintense areas in both occipital & parieto-occital lobe s/o posterior reversible encephalopathy syndrome (PRES).

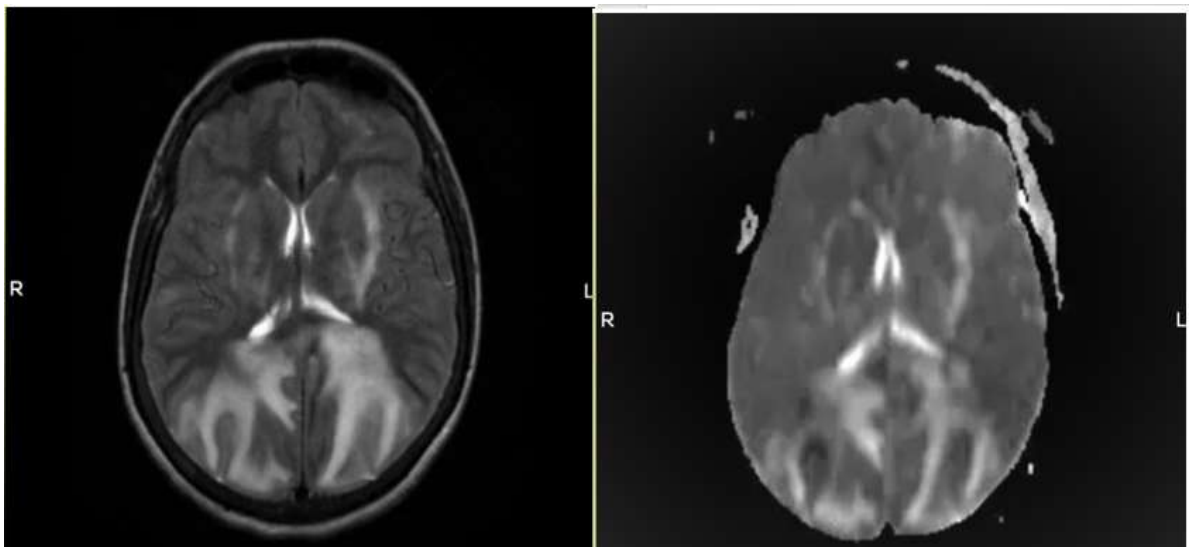

Figure 1: MRI brain showing symmetrical hyperintense areas in both occipital & parieto-occital lobe s/o posterior reversible encephalopathy syndrome (PRES)

After seizure episode patient developed severe hypertension with fluctuations for which intra venous antihypertensives (nitroglycerine & labetolol) & anti-convulsants were started. Though her urea & creatinine were on lower side after dialysis, her neurological status & pulmonary edema worsened, leading to development of respiratory failure for which she was being put on mechanical ventilator with Fractional inspired Oxygen concentration (FiO2)-80%, Positive end expiratory pressure (PEEP)-7, Respiratory rate (RR)-21/min, I:E ratio-1:2, Tidal volume (TV)-360 ml (6 ml/kg). Patient improved after 3 days of ventilation. Ventilation weaned off, & patient being shifted gradually on conservative management. Patient discharged on advice of maintenance hemodialysis, antihypertensives, anticonvulsants, hematinics & antibiotics.

DISCUSSION

PRES is a clinico-radiological diagnosis generally associated with acute hypertension. Till date 3 hypotheses have been proposed (i) Cerebral vasoconstriction causing subsequent infarcts in the brain, (ii) Failure of cerebral autoregulation with vasogenic edema, and (iii) Endothelial damage with blood–brain barrier disruption further leading to fluid and protein transudation in the brain (Sudulagunta *et al.*,, 2017; Bartynski and Boardman, 2007; Bhatia *et al.,* 2019).

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Cerebral blood flow is usually regulated by dilatation and constriction of vessels to maintain adequate tissue perfusion and to simultaneously avoid excessive intracerebral hypertension (Hobson, 2012; Bartynski, 2008) Breakdown in autoregulation occurs above a mean arterial blood pressure of 150-160 mmHg; in chronic hypertension, it occurs at higher pressures (Hobson *et al.*, 2012; Bartynski, 2008) Uncontrolled hypertension leads to hyper perfusion and cerebral vessel damage, resulting in interstitial extravasation of proteins and fluids, causing vasogenic edema (Hobson *et al.*, 2012, Bartynski, 2008). Irreversible damage is seen at mean arterial pressures above 200 mmHg (Bartynski, 2008).

PRES occurring in systemic inflammatory process such as sepsis, eclampsia, transplantation, and autoimmune disease support the endothelial dysfunction theory (Hobson *et al.*, 2012; Bartynski, 2008).

In case of chronic kidney disease (CKD), Severe anemia, deranged kidney functions, acute hypertension, acute blood transfusion, metabolic acidosis, hypoxemia & various inflammatory markers are more prevalent & these may cause functional loss or damage to the integrity of the blood–brain barrier or by other mechanism causes vasogenic edema or cerebral vasoconstriction (Sudulagunta *et al.*, 2017; Wada *et al*., 2013).

Acute blood transfusion may cause a rapid increase in total blood volume leading to cerebral blood flow overload & hyperperfusion (exceeding the capacity of autoregulation of cerebral capillary perfusion pressure) might result in vasogenic edema found in PRES (Sudulagunta *et al.*,, 2017;, (Sudulagunta *et al.*, 2017; Wada *et al*., 2013).

The possibility of severe anemia as the predisposing factor, due to the inadequate supply of oxygen to the brain may result in dysfunction of endothelial cells, further causing a functional loss or damage to the integrity of the blood–brain barrier in capillary circulation cannot be ruled out (Sudulagunta *et al.*,, 2017; Wada, 2013) So PRES can be a major problem in rapid and massive blood transfusion. A high index of suspicion and prompt treatment can reduce morbidity, mortality and pave the path for early recovery.

The differential diagnosis of PRES may include Venous sinus thrombosis, or subdural, intracerebral, or subarachnoid hemorrhage, all of them may have headache, seizures, reduced consciousness, and focal neurologic signs. Other differential diagnosis may include encephalitis (infective, autoimmune & metabolic) or meningitis, posterior circulation stroke, basilar artery thrombosis, Central nervous system vasculitis, or drug toxicity (for example cyclosporine)) (Hobson *et al.*, 2012).

The reversible nature of PRES has been challenged recently based on new reports of permanent neurological impairment and mortality reaching 15%, ranging from 16% to 37% (Sudulagunta *et al.*,, 2017; Burnett, 2010; Lee *et al.*, 2008). Most common complication reported by a study is impaired consciousness (94%) followed by seizures (81%) (Legriel *et al.*, 2012).

Diagnostic modality of choice, Magnetic resonance imaging brain show bilateral occipital, parietal, frontal cortex and subcortical white matter T2/Fluid-attenuated inversion recovery (FLAIR) hyperintensities (Sudulagunta *et al.*, 2017) .

No clinical studies are available till now regarding patients with PRES needing life-sustaining treatments. Symptomatic treatment in the form of immediate control of blood pressure for hypertension; withdrawal of offending agent for drug-induced PRES; anti-convulsive therapy for seizures; temporary renal replacement therapy (hemodialysis/peritoneal dialysis) for acute or chronic kidney disease are the only available therapies. Corticosteroids may improve vasogenic edema, but there is no solid evidence for usage in PRES. Early diagnosis & management results in favorable outcomes vice versa delayed diagnosis & treatment may lead to mortality or irreversible neurological deficit (Sudulagunta *et al.*,, 2017; Roth and Ferbert, 2011) .

CONCLUSION

Our is a rare case of a successful management of PRES occurring as accelerated severe hypertension with seizures in non-hypertensive CKD patient on conservative management. Early diagnosis & management results in favorable outcomes in PRES. That's why it is important & essential for the clinicians to keep the differential diagnosis of PRES in the worsening scenario of CKD, anemia, acute blood transfusion &

hypertensive emergencies. And evaluate the patient accordingly. Timely taken decisions can not only save the life but also give the positive outcomes.

Disclosure

The Financial or other competing interests: None.

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