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Case Report

A Rare Association of Posterior Reversible Encephalopathy Syndrome (PRES) with Glomerulonephritis: 2 Case Reports

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Abstract

Posterior reversible encephalopathy syndrome (PRES) is a disorder of reversible sub cortical vasogenic brain edema predominantly affecting the posterior occipital and Parietal lobe in the setting of acute changes in blood pressure. PRES is often associated with underlying medical conditions such as hypertension, renal disease, autoimmune disorders, and immunosuppressive therapies. On the other hand, Glomerulonephritis, an inflammatory condition affecting the glomeruli of the kidneys, has been suggested as a potential risk factor for PRES. The mechanism underlying PRES is not certain, but endothelial dysfunction is implicated. Treatment is supportive and involves correcting the underlying cause and managing associated complications, such as seizures.

In this case series, we presented two cases of glomerulonephritis with PRES.

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Introduction

Posterior reversible encephalopathy syndrome (PRES) is a clinico-radiological syndrome characterized by symptoms including headache, seizures, visual disturbance, altered consciousness, paresis, hemianopsia, generalized seizures and coma[1]. A variety of conditions have been implicated as causes of PRES, including severe arterial hypertension, acute and chronic kidney diseases eclampsia, pre-eclampsia, immunosuppressive medications such as cyclosporine, various antineoplastic agents, severe hypercalcemia, thrombocytopenic syndromes, Henoch Schonlein purpura, hemolytic uremic syndrome, amyloid angiopathy, Systemic Lupus Erythomatosus (SLE), post-transplantation, infection, sepsis (gram positive organisms predominate) and shock[1,2].

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The mechanism underlying the development of PRES is poorly understood. It is thought that, it is a disorder of dysregulated perfusion of the brain, due to acute rise of blood pressure, typically resulting in reversible vasogenic oedema. There are several theories regarding the cerebral vasculature dysregulation in PRES, but no single mechanism explains the development of PRES in all cases[7].

On other side, Glomerulonephritis is inflammatory condition of glomeruli, where renal parenchymal hypertension develops. Hypertension occurs in the setting of acute glomerulonephritis (GN), chronic GN, diabetic nephropathy, polycystic kidney disease, hypertensive nephrosclerosis, and renal microvascular disorders[3]. Both Glomerulonephritis and hypertension can predispose to PRES which is a medical emergency[4].

PRES is diagnosed by Neuroimaging studies that demonstrate a characteristically reversible vasogenic edema generally involving the subcortical white matter in the posterior cerebral regions[5].

As a result of the low index of suspicion, early diagnosis and treatment of the syndrome are essential for the resolution of the symptoms and radiological features as well as to prevent complications like long-term neurological deficit.

Here we explored 2 cases of rare association of PRES with Glomerulonephritis.

Case 1

Case 1, 20 years old boy, was a diagnosed case of IgA nephropathy with Hypertension evidenced by renal biopsy with histopathology and DIF and was on 3 antihypertensives (Bisoprolol, Nifedipine, Prazosin) and maintenance hemodialysis since then. 6 months after the initiation of treatment, Patient came to us with several episodes of generalized tonic clonic convulsion followed by headache, vomiting and unconsciousness. This event was not preceded by fever, photophobia, neck stiffness, head injury, limb or facial weakness.

On examination, he was moderately anemic, bipedal edema was present, blood pressure 190/120 mmHg. Urine dip stick test showed protein 2+, GCS was 8/15. Fundoscopy Showed multiple blot and flame shaped hemorrhages in both eyes. Planter was bilaterally extensor.

Investigation profile shows Hb 8.1 g/dl, TC-WBC 12450/mm^3, Total platelet count 200000/mm^3, S. creatinine 10.8mg/dl, serum Na+ 135mmol/L, K+ 5.87 mmol/L, Urine PCR 4.47(<o.21 normal). Immunological profile including ANA, complement were negative.

Brain MRI showed (Fig. 1) hyperintense signal change involving part of both frontoparietal, periventricular white matter. Antihypertensive and antiepileptic medications were continued, and with this he showed symptomatic improvement. He is presently on maintenance hemodialysis with 5 antihypertensive drugs.

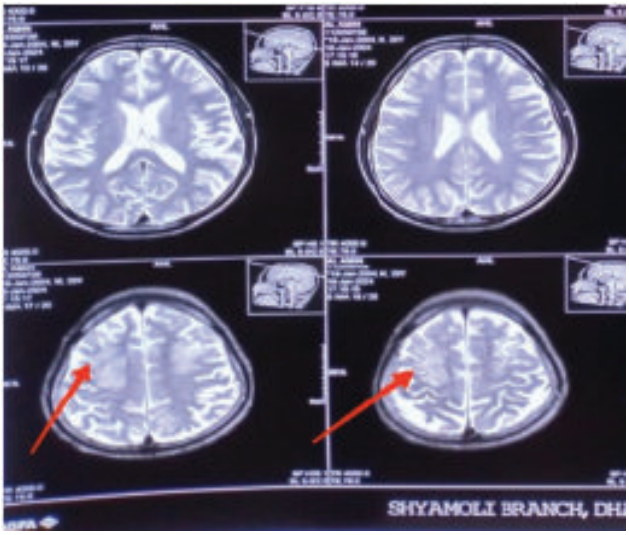

Figure 1. *MRI Brain T2 flair showing hyperintensity in parietal lobe suggesting PRES*

Case 2

Case 2, a 15-year-old boy, developed facial puffiness and pedal edema. Investigations revealed nephrotic syndrome with mild renal failure (serum creatinine, 1.6 mg/dl) with hypertension and hypothyroidism. Renal biopsy findings revealed membranous glomerulonephritis. He was on oral corticosteroids, completed the cycle. 1 year after this illness, he developed accelerated hypertension and seizures. Blood pressure 170/130mmHg, Pulse 85beats/min. Temperature was normal, Respiratory rate 18 breaths/min, bipedal edema was present, bed side urine dip stick test protein 2+, Sugar nil. GCS: E4V4M5=13/15, Cranial nerves examination including fundoscopy revealed normal. Ascites was present as evidenced by shifting dullness.

Investigation profile shows, Hb 11.8 mg/dl, Total WBC 11,300 mm^3, Platelet count 348000/mm^3. Serum electrolytes including Ca, Mg Are within normal limit. Brain MRI (Fig. 2) shows Signal Change lesion in both temporal and parietal region (T1W1- Isointense, T2W1 & FLAIR- Hyper intensity) Suggestive of PRES. Cerebrospinal fluid analysis including viral panel results were normal.

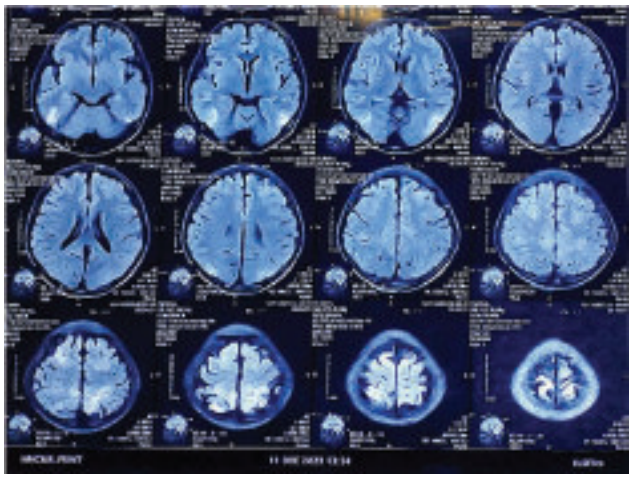

Figure 1. MRI Brain T2 Flair shows hyperintensity in Temporo-parietal region suggestive of PRES

He was treated symptomatically with blood pressure (BP) control. However, he needs 2 antihypertensive agents now for adequate BP control. No further convulsion has occurred.

Discussion

Hypertension risk is a common complication of chronic glomerulonephritis (GN), which includes focal segmental glomerulosclerosis and proliferative forms of GN such as IgA nephropathy. Causes of chronic GN with hypertension include IgA nephropathy (IgAN), membranous nephropathy, membranoproliferative GN, and focal segmental

glomerulosclerosis[3]. There are three main factors working as trigger for development of hypertension in patients with chronic GN, which are similar to those in essential hypertension, but more accentuated. Sodium retention is the prime influencer. Increased Renin-Angiotensin-Aldosterone System (RAAS) activity is responsible for the hypertension. Renal ischemia induced by microvascular damage is a potent stimulus of RAAS. Hypertension also results from overactivity of the sympathetic nervous system. Much evidence indicates increased sympathetic nervous activity in renal disease. Renal ischemia is probably a primary event leading to increased sympathetic nervous activity[4].

Posterior reversible encephalopathy syndrome (PRES) is frequently linked to renal disorders, particularly chronic kidney disease and hypertension[4].Besides GN, there are some reported cases of anti-GBM disease[6] and Stage 3 acute kidney injury[5] associated with PRES .

It has been suggested that severe acute-onset arterial hypertension accounts for most cases of PRES[8]. In our 2 cases, blood pressure was severely elevated in both of cases, thereby supporting the abovementioned link. The term hypertensive encephalopathy was traditionally used to denote this important form of PRES.

Regarding mechanism of PRES, Vasogenic oedema is the defining symptom. There is an ongoing debate on the role of hypertension which is frequently cited as the primary cause of vasogenic oedema. There have been put forth two theories. According to the most widely accepted explanation, severe hypertension prevents the cerebral vasculature from regulating itself, causing hyperperfusion, arteriolar dilatation, and vasogenic oedema. A relative absence of sympathetic innervation (because the sympathetic system raises the threshold of autoregulation) has been proposed as an explanation for PRES's preference for the posterior regions of the brain[4] .

For diagnosis of PRES, Fugate et al. suggested the following criteria : i) acute onset of neurological symptoms; ii) (focal) angiogenic edema in neuroimaging and; iii) reversible findings clinically and/or radiologically[7].

MRI of brain is more sensitive in comparison to CT, which shows high intensity lesions in T2-emphasized or FLAIR (Fluid Attenuated Inversion Recovery) sequences. Parieto-occipital distribution is seen in about 70% of all cases, but frontal sulcus or watershed patterns are usually seen. Lesions in other regions such as the cerebellum, basal ganglia, brain stem, and spinal cord are less common[7].

In our Both cases, MRI of brain T2 flair shows Hyperintensity in parieto-occipital and parieto-temporal distribution respectively.

The differential diagnosis of PRES includes various acute neurologic conditions including ischaemic stroke[11]. A study of 76 patients by Alexander M. McKinney et al, showed that the incidence of regions involvement was parieto-occipital 98.7%, temporal 68.4%, thalamus 30.3%, cerebellum 34.2%, brainstem 18.4%, and basal ganglia 11.8%. The incidence of less common manifestations was enhancement 37.7%, restricted diffusion 17.3%, haemorrhage 17.1% and a newly described unilateral variant 2.6%[10]. W. S. Three major patterns of PRES were noted: the holohemispheric watershed 23%, superior frontal sulcal 27%, parietal occipital 22%, with additional common partial or asymmetric expression of these primary PRES patterns 28%[9].

Here We have presented the case series of 2 cases of young males with Chronic Glomerulonephritis who developed recurrent seizures. Commonly we encounter this seizures as hypertensive encephalopathy. This case series highlights the need to explore neurological events as PRES. Awareness of the diverse clinical and radiographic presentation of acute PRES is essential to avoid misdiagnosis and treatment delay. It is imperative that the syndrome of PRES is correctly recognized on neuro-imaging, as the condition is reversible and potential complications can be avoided with appropriate therapy.

Conclusion

Early recognition and prompt management of PRES in individuals with underlying renal pathology are paramount to mitigate potential neurological complications and improve patient outcomes. Young patient diagnosed with glomerulonephritis with uncontrolled hypertension and Convulsion should put a suspicion of PRES besides hypertensive encephalopathy, although rare presentation.

References

1. Naqi R, Ahsan H, Azeemuddin M. Posterior reversible encephalopathy syndrome: A case series in patients with Eclampsia. J Pak Med Assoc. 2010;60(5).

2. Orlando C, Milani GP, Simonetti GD, Goeggel Simonetti B, Lava SAG, Wyttenbach R, et al. Posterior reversible leukoencephalopathy syndrome associated with acute postinfectious glomerulonephritis: systematic review. Pediatr Nephrol. 2022;37(4):833–41.

3. Ihm CG. Hypertension in Chronic Glomerulonephritis. Electrolyte Blood Press. 2015;13(2):41.

4. Khurana K, Acharya S, Shukla S, Kumar S, Mishra P. Chronic Glomerulonephritis and Malignant Hypertension With PRES (Posterior Reversible Encephalopathy

Syndrome) Presenting As Status Epilepticus: A Case Report. Cureus [Internet]. 2023 Aug 22 [cited 2024 May 3]; Available from: https://www.cureus.com/articles/179510-chronic-glomerulonephritis-and-malignant-hypertension-with-pres-posterior-reversible-encephalopathy-syndrome-presenting-as-status-epilepticus-a-case-report

5. Cha B, Kim DY, Jang H, Hwang SD, Choi HJ, Kim MJ. Unusual Case of Posterior Reversible Encephalopathy Syndrome in a Patient with Anti-glomerular Basement Membrane Antibody Glomerulonephritis: A Case Report and Review of the Literature. Electrolyte Blood Press. 2017;15(1):12.

6. Regmi AR, Bhatta U, Gurung A, Rijal B, Regmi A, Amatya S, et al. Posterior Reversible Encephalopathy Syndrome (PRES) and its relation with COPD. Annals of Medicine & Surgery [Internet]. 2022 Dec [cited 2024 May 2];84. Available from: https://journals.lww.com/10.1016/j.amsu.2022.104877

7. Triplett JD, Kutlubaev MA, Kermode AG, et al. Pract Neurol. 2022;22:183–189.

8. Fugate, Jennifer E et al. Posterior reversible encephalopathy syndrome: clinical and radiological manifestations, pathophysiology, and outstanding questions The Lancet Neurology. 2015;14(9):914-925.

9. Bartynski WS, Boardman JF. Distinct imaging patterns and lesion distribution in posterior reversible encephalopathy syndrome. AJNR Am J Neuroradiol. 2007;28:1320-7.

10. McKinney AM, Short J, Truwit CL, McKinney ZJ, Kozak OS, Santa Guz KS, et al. Posterior reversible encephalopathy syndrome: incidence of atypical regions of involvement and imaging findings. AJR Am J Roentgenol. 2007;189: 904-12.

11. Al-Ansari M, Todwal A. A 20-year-old man with status epilepticus and uncontrolled hypertension. Chest. 2007; 131: 309-12.