LUMEN ET VIRTUS

Posterior Reversible Encephalopathy Syndrome in a dialysis patient – A case report

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ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is an acute clinical-radiological syndrome that occurs due to several causes such as hypertensive encephalopathy, eclampsia, neurotoxicity, drugs, and autoimmune diseases. The patient presents with elevated blood pressure and neurological symptoms such as headache, epileptic seizures, and visual disturbances. The syndrome is confirmed by imaging tests such as computed tomography and, especially, magnetic resonance imaging with findings of vasogenic edema, especially in the white matter in parieto-occipital regions. The symptoms are progressive, but the syndrome regresses completely once treatment is properly and early, otherwise, the patient can face sequelae and even death. Treatment is based on blood pressure control and resolution of the underlying cause. In view of the above, we present the case of a young patient with chronic kidney disease on dialysis who presented clinical and radiological symptoms and whose investigation resulted in the diagnosis of PRES.

Keywords: Posterior Reversible Encephalopathy Syndrome, Chronic Kidney Disease, Systemic Arterial Hypertension, Radiology, Neurology.

INTRODUCTION

Posterior Reversible Encephalopathy Syndrome (PRES) is a clinical-radiological syndrome characterized by a brain disorder caused by vasogenic edema and altered permeability and loss of cerebral vascular autoregulation. The patient usually presents acute symptoms such as seizures, headache and visual alterations and Magnetic Resonance Imaging (MRI) demonstrates changes seen

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in the white matter mostly bilaterally in the posterior region (parietal and occipital lobes) and the most common underlying causes include arterial hypertension, renal failure, preeclampsia, eclampsia, use of cytotoxic drugs and autoimmune diseases. (1) (2) The syndrome can affect any age group, but it often affects more middle-aged adults with an average age of 45 years and is more common in women, even after excluding patients with eclampsia. (2) (3) The prognosis in most cases is favorable, the lesions reversible, and imaging normalized when the underlying cause is effectively treated. The patient's clinic guides the neuroradiological diagnosis of PRES, which should be between the hypotheses of the clinician and the radiologist, because in the case of patients in whom other conditions overlap, the measures and medications used may be additional risk factors for PRES. We present a case of PRES confirmed by MRI in a young patient, emphasizing the importance of early diagnosis, its findings and imaging tests.

CLINICAL CASE

A 24-year-old Caucasian male patient was admitted to the care of the Medical Clinic on 07/25/2023, due to muscle spasms for one day, MSE paresthesia and tonic-clonic seizures. She had a history of systemic arterial hypertension, hyperuricemia, and dialysis CKD (peritoneal dialysis) by a single polycystic kidney.

On the date of admission, a Computed Tomography (CT) scan of the head was performed, demonstrating mucosal thickening in several sinuses, indicating inflammatory sinusopathy with other parameters within the normal range. Ionic calcium: 0.88, with improvement of symptoms after calcium replacement.

Patient returns on 10/05/2023 referred from the Emergency Room (ER), in IOT (orotracheal intubation) + MV (mechanical ventilation) due to seizures, presenting re-entrant crises. Episode of associated agitation and episodes of vomiting with bronchial aspiration and lowering of the level of consciousness, opted for OTI in the ER. A new CT scan of the head was performed at admission, which revealed bilateral parieto-occipital subcortical hypoattenuation, especially on the right, associated with erasure of the sulci between the regional cerebral gyri, suggestive of vasogenic edema, suggesting PRES Syndrome as a diagnostic hypothesis. Treatment was based on blood pressure management with antihypertensive drugs, management of seizures and prevention of new seizures with valproic acid 500mg every 12/12 hours, in addition to the management of patient discomfort, hydration and correction of hydroelectrolyte disorders. The condition improved, and the patient was extubated 10 days after admission, and antiplatelet therapy with ASA was also initiated 10 days after the stroke due to the risk of hemorrhagic transformation based on head CT.

The patient underwent an MRI of the head on 10/27/2023, which demonstrated a small ill-defined area of signal alteration compromising the cortico-subcortical region of the right parietal lobe

in posterior and median topography, suggesting a small area of cystic encephalomalacia with foci of gliosis and with a hemorrhagic component at a late chronic stage (IMAGE) – confirming the diagnostic hypothesis of PRES.

Figure 1 - Axial CT slices of the skull showing hypodensity compatible with vasogenic edema, affecting the occipital regions bilaterally.

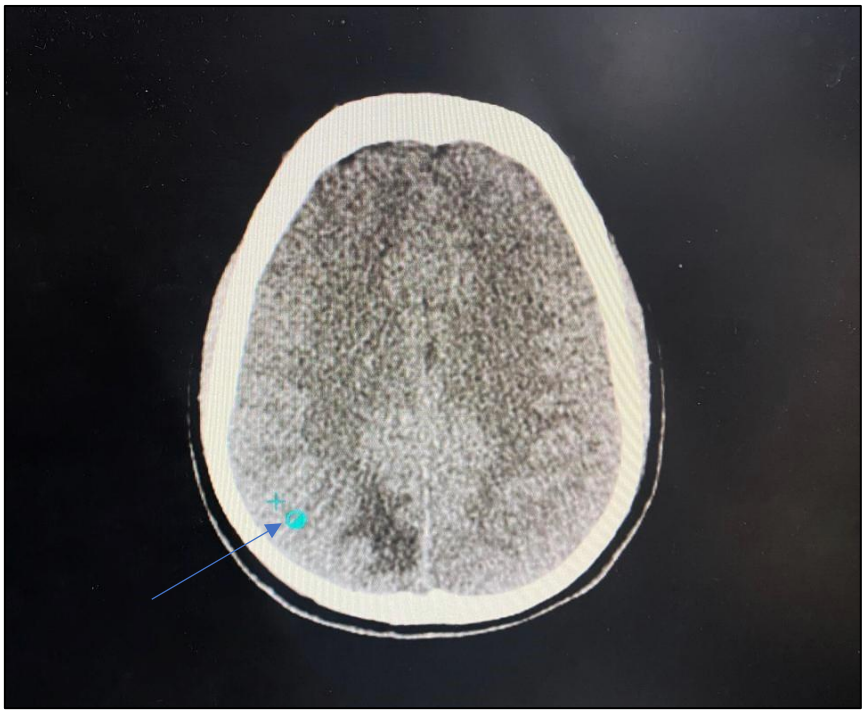
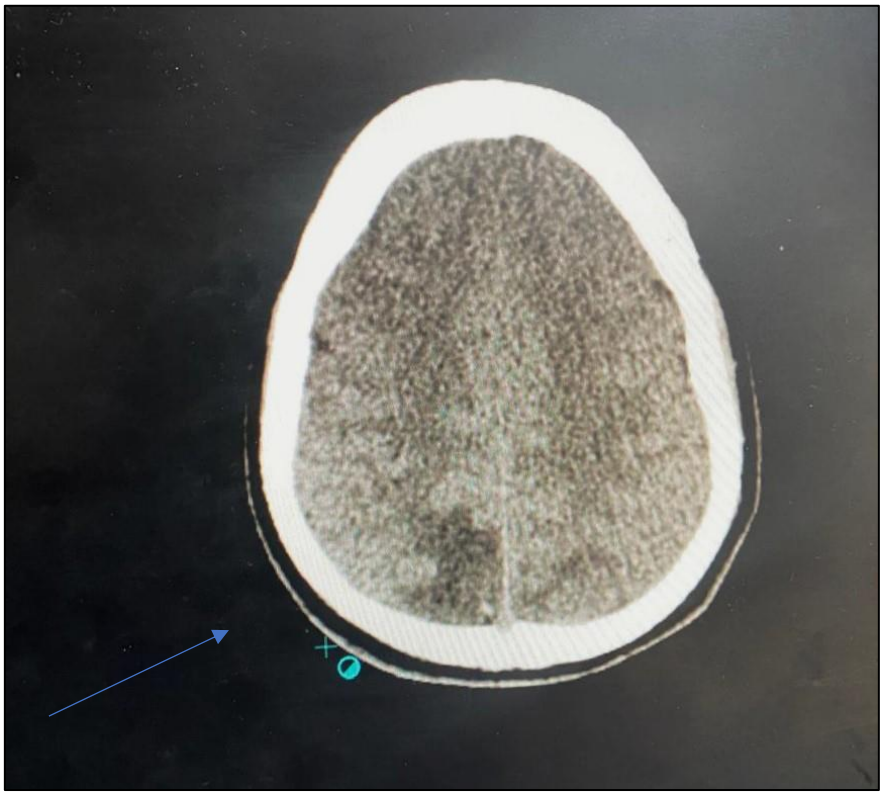


Figure 2 – CT scan of the skull showing hypodensity compatible with vasogenic edema; especially the right of the patient.



DISCUSSION

The syndrome was first described by Hinchey, possibly caused by altered brain permeability and the action of inflammatory cytokines, generating vasogenic edema due to endothelial dysfunction. Its pathophysiology is not well understood, but the prevailing theory is that the abrupt increase in blood pressure exceeds brain self-regulation and causes hyperperfusion and extravasation, with the posterior region of the brain being more susceptible due to less sympathetic innervation in the posterior fossa. Epidemiological data on the syndrome should be interpreted with caution due to the great potential underdiagnosis because its confirmation depends on imaging tests. The syndrome can present in infants up to senility, but is more present in middle-aged women, which may be associated with etiological aspects. (4) (2) (3) Among adults, PRES is mostly associated with hypertension or renal failure and its highest incidence is in patients with eclampsia, followed by patients after bone marrow and organ transplantation, with a lower incidence in patients with chronic kidney disease and autoimmune diseases such as SLE (Systemic Lupus Erythematosus). Even with a lower incidence, the case reported in the present study shows relevance in the case of a young patient with chronic kidney disease, a percentage of the pathology that lacks further evidence in diagnosis and management to expand current knowledge. (5) (2) (6)

The symptoms of the syndrome have a variable clinical spectrum, commonly patients present with headache and seizures, which can be reentrant, as in the case of the patient, until a status epilepticus is established. Motor, visual and state of consciousness changes, nausea and balance disorders can also be seen. The suspicion of PRES requires clinical history, neurological examination and radiological examination for confirmation, preferably Magnetic Resonance Imaging (MRI) – revealing white and gray matter edema mainly in the occipital and parietal lobes and to a lesser extent in the temporal frontal lobes, pons and cerebellum. (2) (4) (7)

The differential diagnosis should be made with the main pathologies involving severe hypertension and crises such as hypertensive, infectious, autoimmune or paraneoplastic encephalitis; manifestation of neoplasm (lymphoma, metastasis); subcortical leukoaraiosis; progressive multifocal leukoencephalopathy; vasculitis of the central nervous system; acute disseminated encephalomyelitis; toxic leukoencephalopathy; among others. (1)

In line with the case of the patient presented, one of the greatest predictors of PRES is renal failure having an association in 55% of the cases, other causes such as eclampsia, autoimmune diseases such as SLE and idiopathic thrombocytopenic purpura and the use of immunosuppressants such as calcineurin inhibitors (Tacrolimus and Cyclosporine) also show a great association with the syndrome (5) due to the possible immunological involvement that is compromised in these cases.

Treatment is based on the underlying cause with measures such as blood pressure control, medications such as sodium valproate to control seizures, management of immunosuppressants if

used and treatment of kidney injury since the syndrome does not have a specific treatment. The profile and symptomatology of the patient in the case raised the suspicion that it was managed correctly and the patient's blood pressure levels normalized without subsequent seizures. The prognosis is good and most patients have total remission, and the aforementioned patient evolved without other complications, resuming his baseline health status. It is worth mentioning that the syndrome can be severe and deadly, with injuries and complications such as cerebral hemorrhage, posterior fossa edema leading to intracranial hypertension and hydrocephalus, therefore, there is great importance in early diagnosis and correction of the causes, as this greatly impacts the patient's prognosis. (5)

CONCLUSION

In short, the case presents hypertensive and chronic renal patients, both conditions that can serve as a "trigger" for PRES and should be in the hall of hypotheses of clinicians if the patient presents symptoms such as headache, visual changes and seizures. This syndrome, presented by a clinical-radiological picture in which reversible vasogenic edema with varied clinical manifestations is presented, tends to regress when treatment is done correctly and is instituted quickly, with blood pressure control and correction of possible electrolyte disturbances. Otherwise, PRES syndrome can leave sequelae such as epilepsy and motor deficits, costing the patient's quality of life. Therefore, the great importance of early diagnosis with correct treatment based on scientific evidence is emphasized. Regarding the pathophysiology and conditions that may have led the episode to occur, it is up to future studies to elucidate.

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