International Journal of Health Sciences and Research
www.ijhsr.org ISSN: 2249-9571

Case Report

Central Variant Posterior Reversible Encephalopathy Syndrome: A Masquerader with Brainstem and Basal Ganglia Involvement Lacking Cortical or Subcortical Edema

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ABSTRACT

Aim: To present a rare case of central variant of posterior reversible encephalopathy syndrome.
Clinical presentation: A 50-year-old male hypertensive patient with electrolyte imbalance presented with acute onset of headache, decreased vision, slurring of speech and ataxia with hypersomnolence. MRI brain showed diffuse T2/FLAIR hyperintensity of bilateral basal ganglia, brainstem and bilateral cerebellar peduncles without cortical or subcortical involvement.
Result: The isolated involvement of brain stem and basal ganglia without cerebral hemispheric involvement is rare and can be called 'central variant' Posterior Reversible Encephalopathy Syndrome, which can result in diagnostic confusion with osmotic demyelination, especially when the patient has associated chronic kidney disease and electrolyte imbalance.
Conclusion: When the MRI shows isolated involvement of brain stem and basal ganglia the diagnosis of central variant PRES should be considered in patients with hypertension even in the presence of electrolyte imbalance.
Key words: posterior, reversible, encephalopathy, central, brainstem

INTRODUCTION

Hinchey et al. in 1996 first described Posterior Reversible Encephalopathy Syndrome (PRES), which is also known as Reversible Posterior Leukoencephalopathy Syndrome (RPLS). [1] Hypertensive encephalopathy (HE) is reported in about 16% of patients with hypertensive crisis. [2] Various other etiologies like cytotoxic drugs, sepsis, autoimmune diseases and eclampsia also have been described. The term "central variant" Posterior Reversible Encephalopathy Syndrome was coined by McKinney et al to refer to a rare variant of Posterior Reversible Encephalopathy Syndrome, with brain stem and basal ganglia involvement without cortical or subcortical edema. [3] The usual presentation of these patients is with acute onset headache, decreased vision, confusions or sometimes seizures.

CASE REPORT

History: A 50-year-old male patient who was a known case of systemic hypertension on alternate medicine treatment presented with headache, giddiness, speech difficulties, hypersomnolence and difficulty in walking for the past two days. He also had gradually progressive decrease in vision over a period of one week, which resulted in near total loss of vision of right eye. There was no history of fever.

Examination: The patient was hypersomnolent and had slurring of speech at presentation. His blood pressure at presentation was 240/140 mm Hg. Pupils showed bilateral equal and sluggish reaction. Power in bilateral upper limbs was 3/5 and that in bilateral lower limbs was 5/5. He had flexor plantar reaction bilaterally. His serum electrolytes levels and renal function tests were abnormal.

He had hyponatremia (118 mEq/L), hypokalemia (2.1mEq/L), hypocalcemia (7.8 mg/dL), hypomagnesemia (1.5 mEq/L) and high serum creatinine(2.6 mg/dL) and blood urea nitrogen level (60 mg/dL). His hemoglobin level was 8.5 g/dL and RBS was 150 mg/dL.

CSF study showed a normal opening pressure of 130 mm Hg and was within normal limits.

Bacterial, fungal and mycobacterial stains and cultures were negative.

Imaging findings: MRI brain was done on the day of admission, which showed diffuse T2/FLAIR hyperintensity with engorgement involving the pons and bilateral inferior, middle and superior cerebellar peduncles and T2/FLAIR hyperintensities involving medulla, midbrain, bilateral lentiform nuclei, thalami and internal and external capsules (Figure 1).

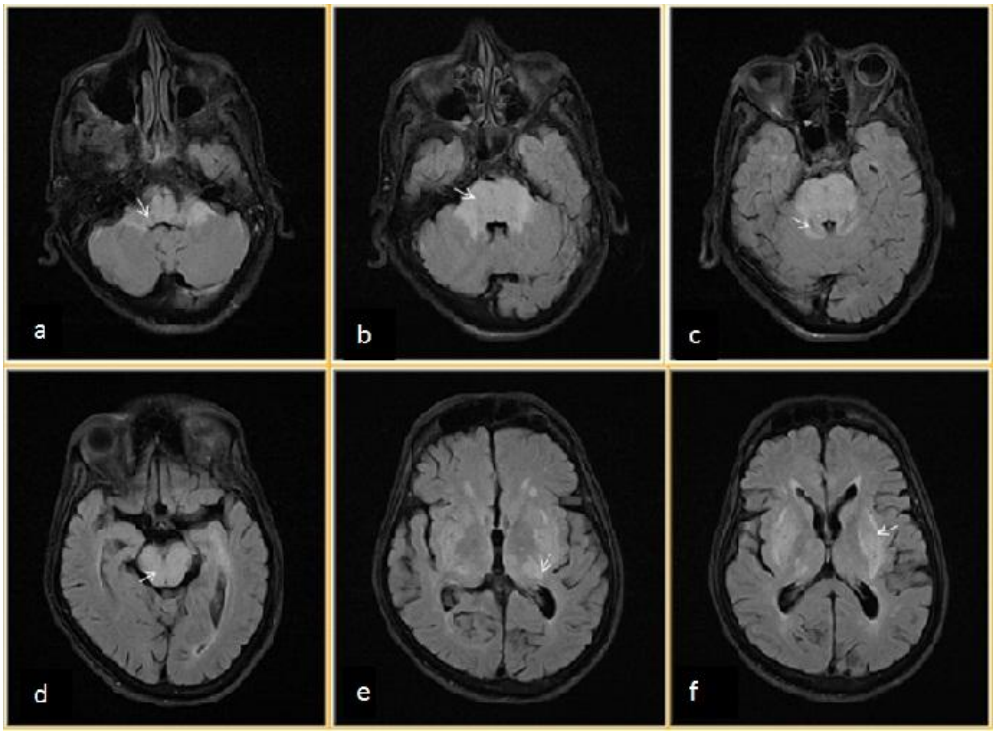

Figure 1: Axial FLAIR images on the day of admission show diffuse hyperintensity involving the medulla and inferior cerebellar peduncles (a), pons and middle cerebellar peduncles (b), superior cerebellar peduncles (c), midbrain (d), thalami (e) and lentiform nuclei (f).

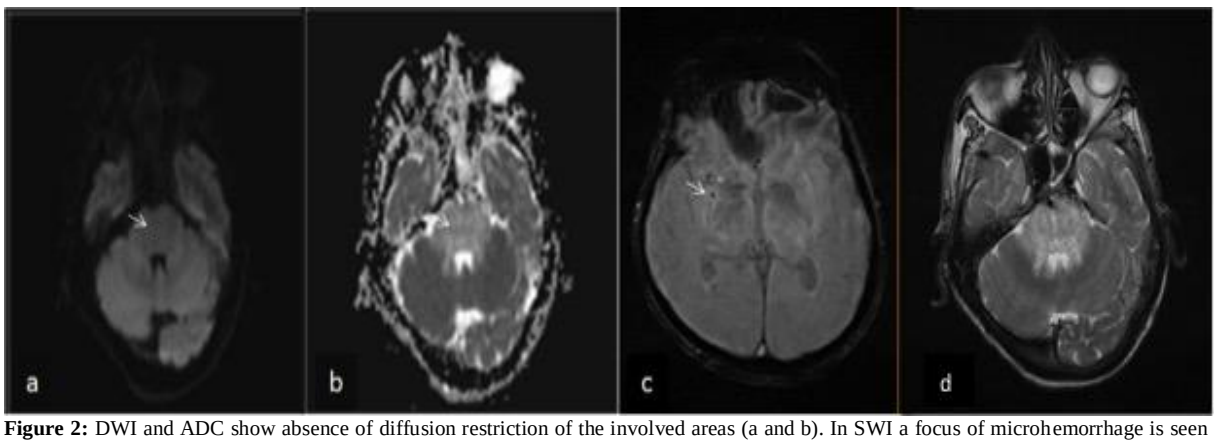

Figure 2: DWI and ADC show absence of diffusion restriction of the involved areas (a and b). In SWI a focus of microhemorrhage is seen in right putamen (c). Axial T2WI shows diffuse engorgement with hyperintensity of pons.

Normal signal intensities were seen in the involved areas in DWI with increased values of apparent diffusion coefficient (Figure 2, a and b). The same areas showed isointense signal in T1WI. Absence of diffusion restriction of the involved areas, suggest lack of cytotoxic edema.

Bilateral mild periventricular leucoaraiosis was present. One punctate foci of blooming was noted in right putamen in susceptibility-weighted images, suggestive of microhemorrhage (Figure 2, c). Intracranial MRA was normal. Since the patient had high serum creatinine value, gadolinium contrast was not given.

Ultrasound abdomen showed features of chronic kidney disease.

With the above clinical history and radiological findings, the primary diagnosis of central / extra pontine myelinolysis was made. Hypertensive encephalopathy was also considered in differential diagnosis taking into account of the high blood pressure.

Management: High blood pressure was treated using intravenous nitroglycerine and was lowered to 170/100 mm Hg by the second day. His blood pressure was normalized in 2 days, which was associated with dramatic improvement in his neurological status, with improvement in sensorium, speech, visual acuity and pupillary reaction to light.

Serum electrolyte imbalance was corrected gradually using intravenous normal saline, injection KCl and syrup potchlor, thereby raising serum sodium level to 134 mEq/L and serum potassium level to 3.4 mEq/L.

Nephrology consultation was sent and the deterioration in renal function was controlled. Ophthalmological evaluation revealed grade IV hypertensive retinopathy with resolving inferior exudative retinal detachment, which was managed accordingly (Figure 3).

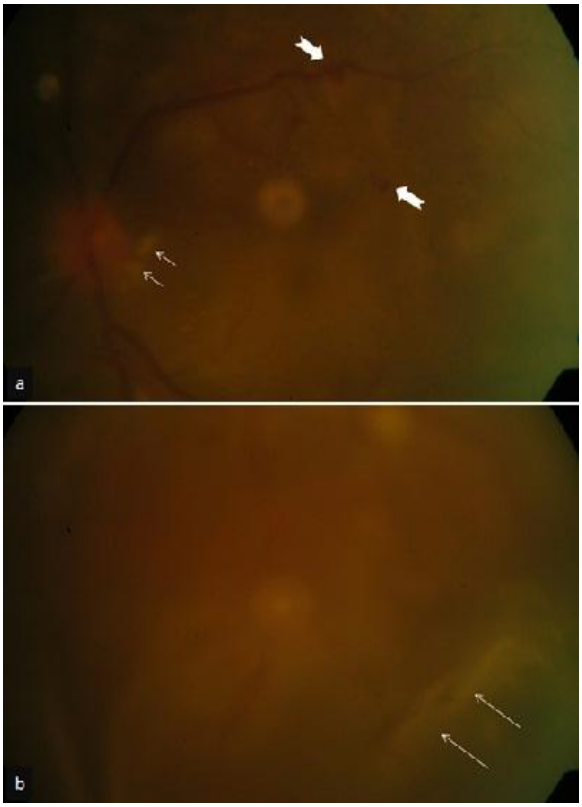

Figure 3: Fundoscopic image (a) of the patient shows multiple microhemorrhages (notched thick arrow) and cotton wool spots (small arrow). Fundoscopic image (b) shows exudative retinal detachment (long arrow).

Repeat MRI brain was done after 4 days which showed dramatic reduction in T2/FLAIR hyperintensity involving pons, medulla, midbrain, bilateral inferior, middle and superior cerebellar peduncles, bilateral thalami and bilateral gangliocapsular regions, which confirms the diagnosis of central variant posterior reversible encephalopathy syndrome, as central /extra pontine myelinolysis does not show rapid resolution in MRI findings (Figure 4). The microhemorrhage, which was seen in the previous MRI, was present in the repeat scan also.

Clinically, the patient improved significantly in 4-5 days. He was discharged with advice on drugs. He needed follow up with nephrology and ophthalmology departments.

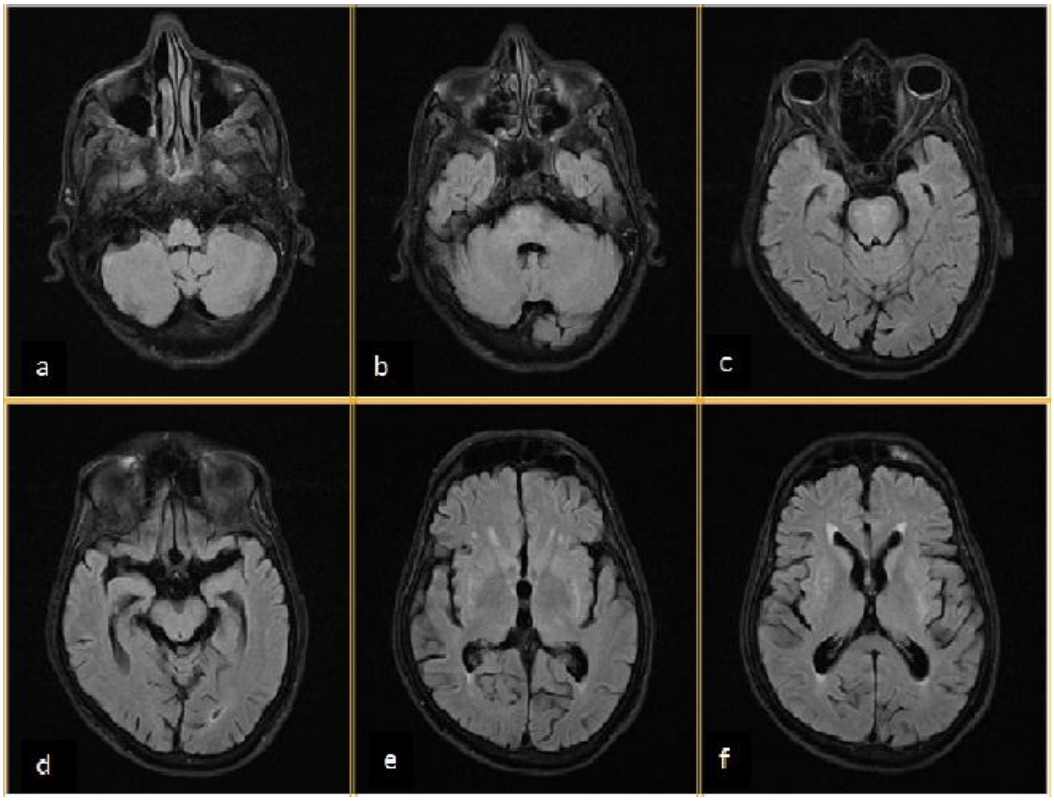

Figure 4: Axial FLAIR images after 4 days showsignificant reduction in the hyperintensity involving the medulla (a), pons and middle cerebellar peduncles (b), superior pons (c), midbrain (d), thalami (e) and lentiform nuclei (f).

DISCUSSION

The vasogenic edema involving the cortical and subcortical regions of parietal, occipital and rarely the posterior frontal lobes as seen in Posterior Reversible encephalopathy Syndrome is thought to occur due to failure of posterior circulation autoregulation, usually, in response to high blood pressure. The causes of PRES include hypertension, cytotoxic drugs, sepsis and eclampsia. In central variant Posterior Reversible encephalopathy Syndrome, the edema involves the basal ganglia and brainstem without cortical or subcortical involvement. McKinney, et al. referred this variant as "central-variant PRES" or "central reversible encephalopathy syndrome", particularly because central structures other than the brainstem can also be involved in this variant. [3] T2/ FLAIR images show hyperintensity of the involved areas. Usually there is no evidence of diffusion restriction indicating the absence of cytotoxic edema of the involved regions. Multiple microhemorrhages may be noted in basal ganglia and brainstem in susceptibility-weighted images, which do not resolve on follow up. These microhemorrhages are thought to be due to underlying vasculopathy. Chronic kidney disease, in association with accelerated hypertension as seen in our patient, might have been a contributing factor for the development of central PRES. Cruz-Flores, et al. reported that two-thirds of the hypertensive encephalopathy patients with brainstem involvement, in addition to elevated blood pressure, had comorbidities such as renal failure. [4] A possible explanation to this would be that the vasoconstriction response fails to maintain constant blood perfusion during hypertension in the uremic state, as demonstrated in the animal model. [5]

Our patient also had serum electrolyte abnormalities like hyponatremia and hypokalemia, which also could have contributed for the development of vasogenic edema. Kastrup et al reported two cases of PRES due to electrolyte imbalance. [6] Hypokalemia has been found to be associated with a decreased concentration of

sodium- and potassium-activated adenosine triphosphatase (Na-K-ATPase) in endothelial or glial cell membranes, as shown in skeletal muscle. [7] A decrease in Na-K-ATPase activity during hypokalemia may limit the ability of a cell to preserve and/or regulate its volume. This may facilitate the development of vasogenic edema.

The alternate diagnoses that can be considered in case of atypical presentation include central or extra pontine myelinolysis, rhombencephalitis, basilar thrombosis with infarctions and pontine glioma.

In our case, the diagnosis of central or extra pontine myelinolysis was made after the first MRI, as the patient had electrolyte imbalance. However, follow up MRI showed dramatic improvement in the MRI findings which rules out central / extra pontine myelinolysis. Rhombencephalitis is an inflammatory process of the hindbrain involving the brainstem and cerebellum, which can be of infectious or autoimmune etiology. The patient usually presents with headache, vomiting, fever, cranial nerve palsies, ataxia, mental alteration or seizures. Listeria is the most common cause of rhombencephalitis. Jubelt et al reported cerebrospinal fluid (CSF) pleocytosis in 75% of patients with listeria rhombencephalitis and in 94% of patients with Behçet rhombencephalitis. [8] In various viral infections causing rhombencephalitis, CSF pleocytosis showed lymphocytic predominance. [8] In our patient, CSF study was normal and basal ganglia involvement was also present in addition to brainstem involvement and thus rhombencephalitis was ruled out.

Basilar thrombosis with acute infarction shows diffusion restriction of the involved areas, which was absent in our case.

Diffuse brainstem glioma in adults has a longer duration of symptoms. Gait disturbance, headache, limb weakness and cranial nerve palsies are the main presenting symptoms and signs in most of the cases. [9] MRI at presentation reveals a diffuse infiltration of the pons, often increasing the size of the brainstem considerably. The involved area shows T2-weighted high signal intensity and T1-weighted low signal intensity and usually there is no contrast enhancement. Our patient had an acute presentation and the rapid resolution of MRI findings in 4 days is against the diagnosis of brainstem glioma.

CONCLUSION

Central variant Posterior Reversible Encephalopathy Syndrome is a clinico radiological syndrome that results in vasogenic edema of the brainstem and basal ganglia without cortical or subcortical involvement, which can lead to diagnostic confusion. When the MRI shows isolated involvement of brain stem and basal ganglia, the diagnosis of central variant PRES should be considered in patients with hypertension even in the presence of electrolyte imbalance. The associated uremia and / or electrolyte imbalance could be the contributory factor for the development of PRES. PRES being a treatable condition, prompt diagnosis and treatment helps in rapid improvement of the patient's clinical status.

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How to cite this article: Mohanan S, Anand R, Nair MNR. Central variant posterior reversible encephalopathy syndrome: a masquerader with brainstem and basal ganglia involvement lacking cortical or subcortical edema. Int J Health Sci Res. 2017; 7(9):326-331.

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