

Posterior reversible encephalopathy syndrome in association with fibromuscular dysplasia: a case report

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Abstract

Posterior reversible encephalopathy syndrome is commonly associated with hypertension and hypertensive crisis. Clinical characteristics are headache, visual changes, altered mental status and seizures. There is a strong association with cytotoxic edema, which in most cases involve posterior areas of the brain. This syndrome

usually resolves with prompt recognition and standard treatment of the triggering condition. We present the case of a 17-year-old man who developed posterior reversible encephalopathy syndrome associated with hypertensive crisis due to fibromuscular dysplasia of renal arteries.

Key words: Posterior reversible encephalopathy syndrome, hypertensive crisis, fibromuscular dysplasia.

Introduction

Posterior reversible encephalopathy syndrome (PRES) was described in 1996 by Hinchey as a clinical and radiological disorder characterized by multiple neurologic manifestations such as headache, paresis, hemianopsia, blurred vision, nausea, change in mental status, generalized seizures, and in some cases coma may develop. (1)

The most recognized hypothesis about the pathogenesis of PRES is the auto-regulatory failure. When upper limit of cerebral auto regulation is exceeded, arterioles dilate and cerebral blood flow increases in a pressure-passive manner, this is accompanied by an increase in systemic blood

pressure. The resulting brain hyperperfusion, may lead to a breakdown of the blood brain barrier, allowing extravasation of fluid and blood products into the brain parenchyma with the subsequent development of vasogenic edema. The auto-regulatory failure can cause a local reactive vasoconstriction, leading to hypoperfusion and cytotoxic edema. (2,3)

This syndrome is strongly associated with hypertension and hypertensive crisis. The largest case series described in the literature found hypertension in 65%. (4) Although the recognition of this disorder has increased lately, the real incidence is still unknown and diagnosis is still a challenge. We describe a case of a patient with the association of PRES and hypertensive crisis due to fibromuscular dysplasia (FMD) of renal arteries.

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Case presentation

A 17-year-old man with irrelevant medical history presented to the emergency department of the General Hospital of Culiacan with a previous history of poor appetite, vomiting and altered consciousness for one week. After that, the patient suddenly developed intense headache and seizures. The initial blood pressure was 220/128 mmHg. Serum

electrolytes, renal function test and other routine laboratory tests were normal. Electrocardiogram, chest radiograph and transthoracic echocardiography were normal. The patient was treated with intravenous nitroglycerin and a combination of two oral antihypertensive medications, including a beta-blocker and a calcium channel blocker. Blood pressure was not controlled, seizure activity persisted and anticonvulsant therapy with phenytoin was implemented.

Magnetic resonance imaging (MRI) of the brain demonstrated the presence of edema primarily involving the white matter of the posterior cerebral hemispheres that was isointense or hypointense on T1 images and hyperintense on T2-weighted images. There was hyperintensity on the apparent diffusion coefficient (ADC) map and no enhancement was noted after intravenous contrast administration.

The approach for secondary hypertension was initiated, including a selective renal angiography (SRA) which revealed a left renal artery stenosis with a normal right artery. SRA showed the typical fibromuscular disease pattern of “string of beads” appearance, which is characterized by alternating areas of narrowing and dilatation, located in the middle portion of the left renal artery, associated with a reduction in luminal diameter greater than 80% (**Figure 1**).

Balloon dilatation of the stenotic renal artery was successfully performed. Headache and seizure control were achieved. Blood pressure was normalized over a period of 2 weeks with a single oral antihypertensive medication. One month and a half later a MRI showed resolution of the initial lesions (**Figure 2**).

Discussion

The clinical presentation of a patient with FMD may vary widely, and it is related in part with the arterial segment involved and the severity of the disease. The most common manifestation is hypertension. Neurologic disorders such as transient ischemic attack and stroke may also be present, but the association with PRES has not been described previously. The work-up for suspected PRES should include serial blood pressure measurements, basic metabolic panel, complete blood count and urine toxicology screening in order to differentiate PRES from other possible diagnoses. (2,5)

This report also highlights the value of cerebral imaging studies in the early recognition and diagnosis of this syndrome. The value of computed tomography (CT) scan is too low, and is useless to distinguish between PRES and acute stroke, making MRI the image study of choice. (6) The typical findings are symmetrical white matter edema in the posterior cerebral hemispheres, particularly the parieto-occipital regions due to the lack of sympathetic innervations of the vasculature in this zone. The calcarine and paramedian regions of the occipital lobe are usually spared, supporting the differentiation between PRES from bilateral posterior cerebral infarctions. (7) Conventional MRI usually demonstrate hyperintensity on T2WI. Both cytotoxic and vasogenic edema show an increased signal in DWI. The T2 ADC map serves to remove the underlying T2 signal contribution, and normally appears bright in PRES but dark in infarction; it is a very useful technique for a more accurate and specific diagnosis of this disorder. (8)

The most important to implement an effective treatment is early recognition since this syndrome may be reversible. It is important to identify patients with severe hypertension because the initial aim in this setting is blood pressure control. The use of an intravenous short half-life antihypertensive drug is the preferred approach. Drugs such as nicardipine, labetalol or nitroprusside are the drugs of choice in this setting because they permit easy titrating. (9) Additive administration of eplerenone, especially in patients with bilateral renovascular hypertension has been described recently. (10) The present case also shows the importance of antiepileptic medications in these patients. Phenytoin is indicated for seizures associated with acute episodes of PRES, except in eclampsia setting. The progression to chronic epilepsy has not been reported previously, making long-term antiepileptic treatment not necessary. (4)

The identification of a comorbid condition in patients with PRES is very important because some pathologies such as systemic lupus erythematosus, (11) cryoglobulinemia, hemolytic uremic syndrome and patients under chemotherapy with drugs such as cyclosporine (12) have their specific treatment implications; further the majority of these comorbid conditions share a high prevalence of renal impairment which is also associated with fluid overload, electrolyte disturbances or uremia. (2,13)

Finally it is worth mentioning that this syndrome deserves

special attention by the clinician, in part because of early identification and correct diagnosis may be a clinical challenge. Proper use of brain imaging studies, the search for comorbidities and timely and efficient treatment of hypertension are of vital importance, allowing the opportunity of avoiding long-term sequelae. The association of PRES and FMD of renal arteries should be considered,

particularly in young patients with severe hypertension.

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Figure 1. Beaded appearance of the mid-left renal artery (red arrows) with focal areas of stenosis

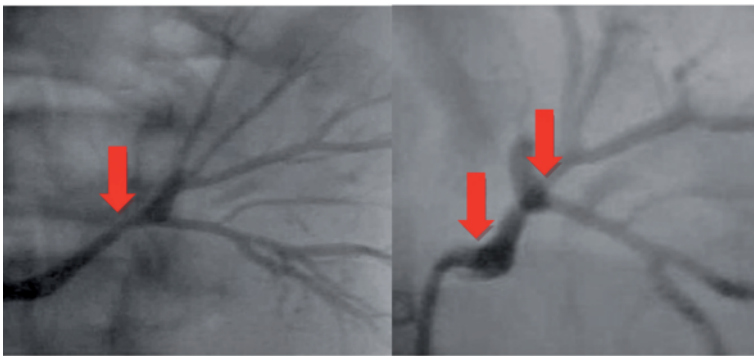
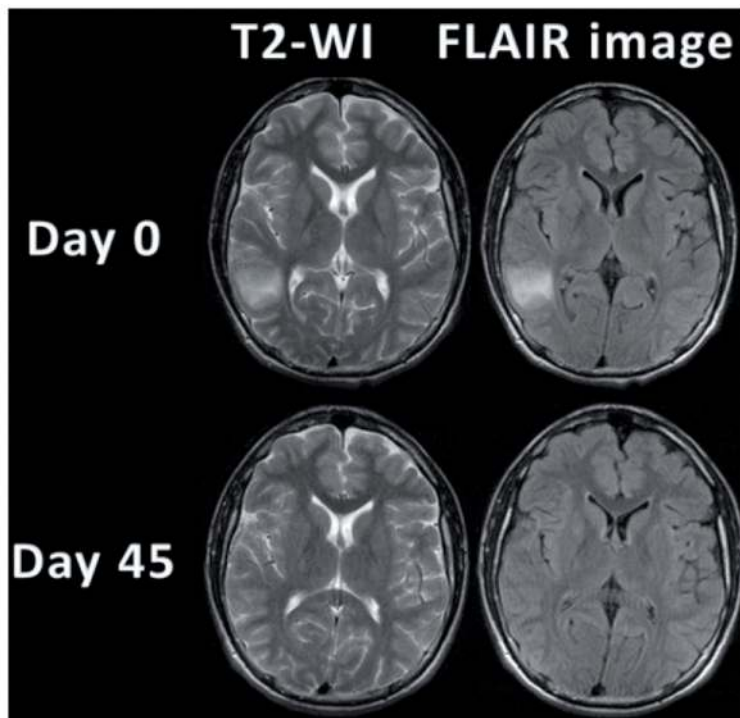


Figure 2. T2-WI and FLAIR image at day 0 and day 45



Legend: T2-WI=T2-weighted image; FLAIR=fluid attenuation inversion recovery

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