POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME: A DISEASE RARELY DESCRIBED IN OLDER ADULTS

Síndrome da encefalopatia posterior reversível: uma patologia pouco descrita no idoso

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ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is a disease rarely described in older adults. It is characterized by subacute onset of a set of clinical and radiological signs and a variety of neurological symptoms, such as headaches, seizures, and cognitive disorders. In the vast majority of patients, clinical presentation includes high blood pressure and hypertensive emergency. Magnetic resonance imaging (MRI) is the gold standard for diagnosing this condition using imaging findings. When the underlying cause is promptly recognized and treated, symptoms and imaging abnormalities may be completely reversible. The authors report the clinical case of an 87-year-old woman first admitted for treatment of community-acquired pneumonia. She returned to the emergency department 24 hours after discharge presenting with complex visual and neurological symptoms. An MRI scan showed lesions of bilateral occipital hypodensities, suggestive of vasogenic edema and compatible with PRES. Complete regression of brain lesions was observed after tight control of hypertension.

KEYWORDS: posterior leukoencephalopathy syndrome; hypertension; magnetic resonance imaging.

RESUMO

A síndrome da encefalopatia posterior reversível, conhecida como PRES, é rara e pouco descrita em idosos e é caracterizada pelo início subagudo de um conjunto de sinais clínicos e radiológicos e uma variedade de sintomas neurológicos, como cefaleia, crises convulsivas e transtornos da cognição. Na grande maioria dos pacientes, a apresentação clínica inclui pressão arterial elevada e emergência hipertensiva. A ressonância magnética (RM) é o exame padrão-ouro para o diagnóstico imaginológico dessa entidade. O quadro clínico e as alterações de imagens podem se tornar reversíveis caso seja detectada precocemente e tratada a causa base da síndrome. Os autores apresentam o caso clínico de uma idosa de 87 anos, internada para tratamento de pneumonia comunitária retornando ao setor de emergência 24 horas após a alta hospitalar apresentando sintomas neurológicos visuais complexos. Ao exame de RM, observaram-se lesões de hipodensidades occipitais bilaterais, sugestivas de edema vasogênico, compatível com PRES. Após o rigoroso controle da pressão arterial, verificou-se a reversibilidade total dessas lesões cerebrais.

PALAVRAS-CHAVE: síndrome da encefalopatia posterior reversível; hipertensão arterial; imagem por ressonância magnética.

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INTRODUCTION

The neurological syndrome first known as reversible posterior encephalopathy is characterized, according to Hinchey et al.,[1] by headache, seizures, transient visual disturbance, and cognitive disorders. It is associated with the presence of vasogenic edema in the white matter of the occipital and parietal lobes.[1]

The term posterior reversible encephalopathy syndrome (PRES) was proposed by Casey et al. in the 2000s, who defined it as acute and subacute onset of headache and manifestation of focal neurological signs.[2]

The main causes of PRES include hypertension, eclampsia, thrombotic thrombocytopenic purpura, hemolytic-uremic syndrome, and use of immunosuppressive drugs, such as tacrolimus and cyclosporin A, and chemotherapy agents, such as cisplatin, methotrexate, and interferon α.[3]

PRES can also be diagnosed by complementary imaging studies, particularly computed tomography (CT) and magnetic resonance imaging (MRI). A typical finding is a bilateral edema surrounding the white matter and reaching the posterior vessels of the parietal and occipital lobes. MRI is fundamental to distinguishing a cytotoxic edema, which is associated with ischemia and infarction, from a vasogenic edema, which is typical of the syndrome.[4,5]

Some mechanisms have been suggested regarding the pathophysiology of PRES. Abrupt onset of hypertension is believed to cause a breakdown in cerebral autoregulation, especially in the occipital region. This leads to hyperperfusion, which is directly associated with leakage of proteins and fluids, forming a focal vasogenic edema. Other proposed mechanisms are related to endothelial dysfunction due to sepsis and eclampsia, despite its association with vasospasm and ischemia.[4,6]

Typical PRES lesions tend to be completely reversible, especially when the primary cause is treated, thus providing a good prognosis. When the primary cause is not addressed, the situation may become irreversible, with a poor prognosis involving cortical blindness and death.[3]

CASE REPORT

The authors report the clinical case of an 87-year-old Caucasian woman who presented with hypertension, obesity, and chronic obstructive pulmonary disease as comorbidities.

The patient had been admitted 10 days before the onset of neurological symptoms for treating a community-acquired pneumonia. She was clinically stable until the seventh day of admission, when she noticed her vision was blurred. In that occasion, she decided to omit the symptom from the medical team in order to avoid the risk of not being discharged, which was expected to occur at the end of antibiotic treatment.

The patient was discharged three days later, when the 10-day cycle of intravenous antibiotic therapy (tazobactam + azithromycin) was completed. She was prescribed oral antibiotic therapy (moxifloxacin) at home for four days, associated with usual medications for blood pressure control and long-term bronchodilator.

The patient returned to the emergency department 24 hours after discharge presenting with worsening visual symptoms, including blurriness and visualization of "yellow and blue confetti." On initial physical examination, the patient was alert and oriented. Muscle strength was preserved in the four limbs (grade 5), and overall reflexes were normal (grade 2). Bilateral flexor plantar responses were normal. Tactile sensitivity was preserved on double stimulation. Facial muscles had no changes. A confrontation visual field test showed quadrantanopsia to the upper right. Pupils were equal and reactive to light. Blood pressure was 183 × 85 mmHg, and heart rate was 98 bpm. She had no fever.

A non-contrast-enhanced CT scan of the brain showed mild cortical and subcortical hypodensity in the occipital lobes and calcified atherosclerotic plaques in the carotid siphon and vertebral arteries.

An MRI scan of the brain confirmed the presence of occipital and parietal lesions to the right (Figure 1), symmetrically

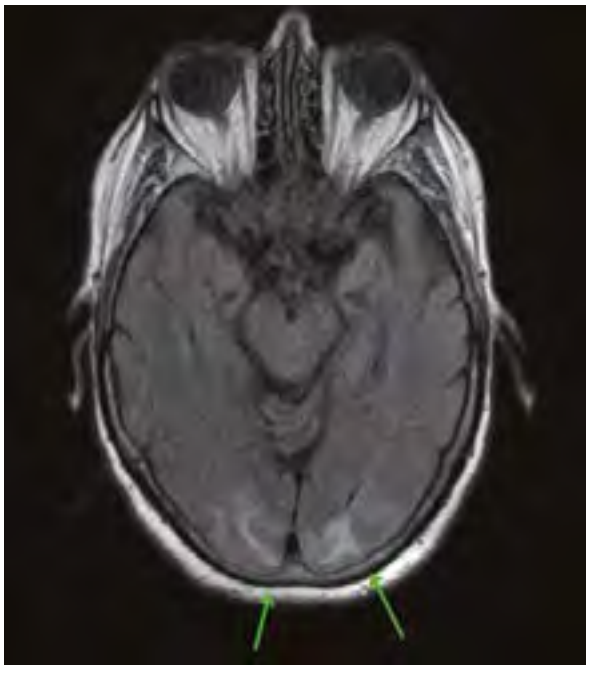

Figure 1 Magnetic resonance imaging with fluid-attenuated inversion recovery sequences showing cortical and subcortical focal areas of hyperintensity, corresponding to an edema.

distributed in both sides at the level of the white matter. There was no restricted water diffusion and no associated bleeding (Figure 2). Signal intensity of the optic nerve was normal (Figures 1 and 2). MRI findings suggested the presence of transudate, especially in the vessels of the vertebrobasilar system. Apparent diffusion coefficient (ADC) showed no changes, thus confirming the absence of cytotoxic edema and suggesting the presence of vasogenic edema (Figure 3).

The patient was admitted to the intensive care unit (ICU) for neurological monitoring and blood pressure control. She required intravenous administration of vasodilator for blood pressure control, which was interrupted on the third day of ICU admission, when oral antihypertensive therapy was started. On neurological examination, she showed nasal hemianopsia to the right and scintillating scotomas on her visual field, which she defined as "brilliant little red balls."

Complementary tests were performed for assessing the clinical consequences of hypertension. Renal function, ionogram, and lipid profile showed no changes. Echocardiogram revealed preserved left ventricular systolic function and normal segmental analysis, normal right ventricular contractile function, normal valve function, and absence of areas of fibrosis or hypokinesis. Chest radiography showed enlarged cardiac silhouette and signs of vascular congestion. Color Doppler ultrasonography of the carotid arteries revealed mild and diffuse medial-intimal thickening and calcified atherosclerotic plaques, eccentric and 30% obstructive, in both bulbar regions and where both internal branches emerge. Blood flow velocity was normal in both carotid arteries and their respective internal and external branches. Ultrasonography of the lower extremities detected no signs of deep vein thrombosis.

The patient was discharged on the 13th day of admission with complete regression of symptoms and improved occipital lesions. Her blood pressure was 140 × 90 mmHg.

DISCUSSION

PRES is a syndrome that rarely affects older adults and whose typical features are clinical and imaging abnormalities. Because its main cause is blood pressure variation, the condition is underdiagnosed.[2,7]

Clinical and imaging abnormalities are usually reversible. The presence of bilateral vasogenic edema surrounding the white matter in the posterior vessels is more clearly shown on MRI, in which a high ADC confirms the absence of cytotoxic edema.[7-10]

Thus, the pathophysiology of PRES is believed to be associated with cerebral blood flow autoregulation, which allows the leakage of fluids into the interstitium, forming a vasogenic edema (Figure 1). The posterior vessels are reached because of decreased sympathetic innervation.[8,10]

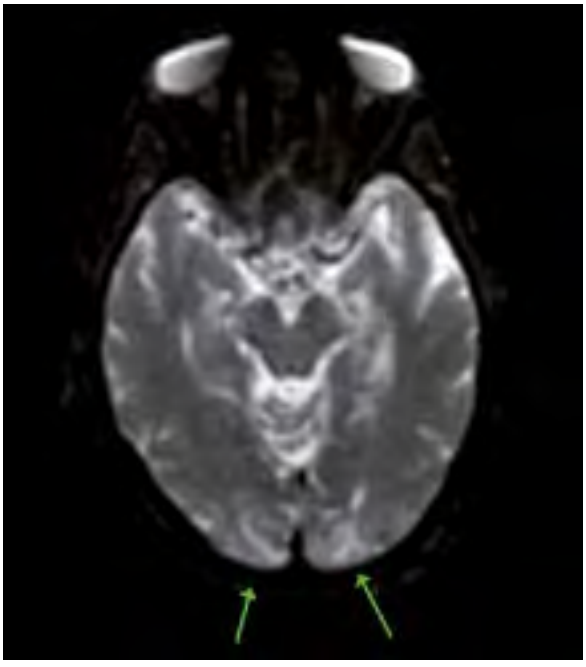

Figure 2 Diffusion-weighted magnetic resonance image showing cortical and subcortical hypersignal areas in the occipital lobes.

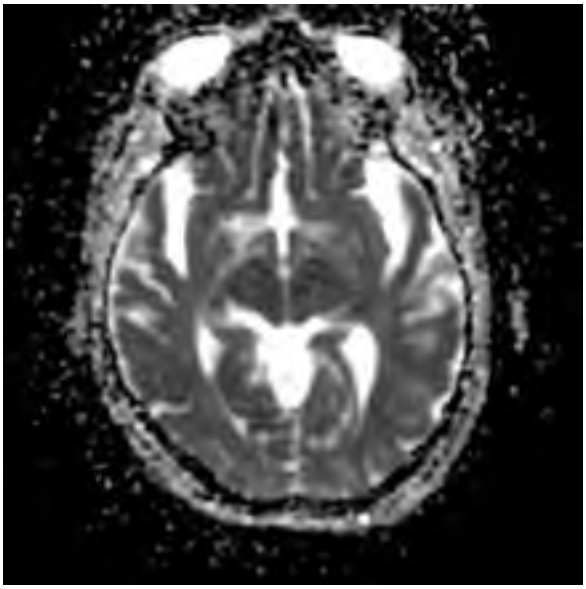

Figure 3 Apparent diffusion coefficient map showing no changes, thus confirming the absence of cytotoxic edema and suggesting the presence of vasogenic edema.

Differential diagnoses include cerebral infarction, vasculitis, sagittal sinus thrombosis, and encephalitis. The neurological symptoms associated with the syndrome are headache, seizures, aphasia, nausea, and cognitive and sensory disorders. Visual acuity is most commonly affected, progressing with hemianopsia and amaurosis or visual hallucinations.[10,11]

PRES is potentially reversible if the primary cause is promptly recognized; otherwise, the patient may have a poor prognosis, as the condition leads to ischemic damage and irreversible neurological sequelae.[5,8,9]

The reported patient did not present with seizures or headaches. Predominantly, she showed visual disturbances, including the visualization of colorful hats and brilliant clothes, which occurred in the ICU setting with full lighting. Acute mental confusion was observed by a geriatrician of the ICU team during anamnesis and physical examination, then confirmed by family members. The onset of symptoms during admission for treatment of an acute infection made possible a prompt recognition when the patient returned to the emergency department. The patient's clinical presentation combined with imaging findings provided by a radiologist allowed an early diagnosis. Prompt control of blood pressure resulted in favorable outcomes, with complete regression of symptoms and lesions.

The main purpose of this case report is to highlight the importance of both clinical and imaging investigation in early detection of PRES, a neurological syndrome that may lead to irreversible sequelae.

CONFLICT OF INTERESTS

The authors declare no conflict of interests.

AUTHORS' CONTRIBUTIONS

All authors were involved in the preparation of the manuscript and organization of data and images, and all of them read and approved the final manuscript.

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