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CASE REPORT

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Posterior Reversible Encephalopathy Syndrome. A case report.

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Abstract

We report a case of posterior reversible encephalopathy syndrome (PRES) in a 52-year-old patient with a history of chronic unmonitored hypertension who was brought to the emergency department for sudden onset of seizures, headache and confusion while driving, leading to a violent traffic accident. Magnetic resonance imaging (MRI) Flair sequence showed hypersignals of the posterior cerebral fossa corresponding to vasogenic edema. The radio-clinical evolution was favorable under symptomatic treatments with a disappearance of the hypersignals three months after the beginning of the symptoms. It seems important to think of a RPRS in front of a picture of confusion, headaches and seizures in a context of malignant hypertension badly followed.

Keywords: PRES, MRI, hypertension, vasogenic edema

1 | INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) is a rare neurological disorder (1, 2). Described for the first time in 1996 by Hinchey et al, this syndrome is characterized by seizures, confusion, headache, visual disturbances and cerebellar ataxia (3, 4). The etiologies are multiple and varied such as malignant hypertension or eclampsia, severe renal failure, malignant tumor under chemotherapy or immunosuppressive treatment for organ transplantation (1). The diagnosis is most often made by magnetic resonance imaging (MRI) Flair sequence (fluid-attenuated inversion recovery), which reveals hypersignal lesions corresponding to

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vasogenic oedema that may involve mainly the cerebellum, brainstem, occipital and temporal lobes, and sometimes the basal ganglia and white matter. Treatment is based on the treatment of the causative condition. The prognosis of RPRS is generally satisfactory with control of hypertension and treatment of the cause (2).

We report a case of reversible posterior encephalopathy syndrome in a chronic severe hypertensive patient with hypersignals on brain MRI Flair sequence.

Clinical case

A 52-year-old right-handed patient with a history of known chronic hypertension and alcoholism was brought to the emergency room after a sudden onset of horizontal binocular diplopia, headaches and seizures while driving, resulting in a violent traffic accident. The physical examination in the emergency room revealed an increase in blood pressure to 250/132mmHg in the left arm, 243/148mmHg in the right arm, a confused but conscious patient, nausea, horizontal binocular diplopia, absence of sensory-motor deficit and nystagmus. The National Institute of Health Stroke Scale (NIHSS) was estimated to be 3. Emergency brain magnetic resonance imaging (MRI) diffusion sequence (Fig 1A) was without abnormality, Flair sequences showed diffuse hypersignals related to cerebral edema lesions of the cerebellum (Fig 1B), brainstem, and vermis (Fig C and D).

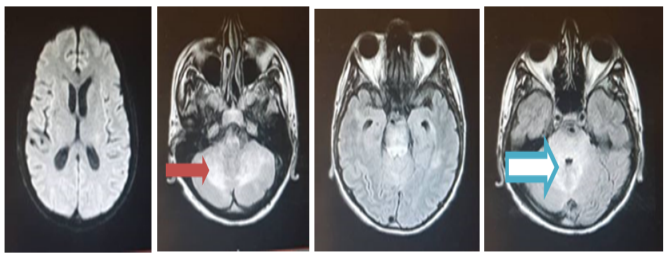

FIGURE 1: Braindiffusion MRI (A) showed no visible lesions, in MRI Flair sequence diffuseedema lesions of the cerebellar posterior fossa (B), brainstem and vermis (Cand D).

The electrocardiogram (ECG) revealed significant left ventricular hypertrophy (LVH) with ventricular overload (T wave in V4, V6 and D1 in AVL), fragmented QRS. Transthoracic echocardiography (TTE) showed concentric LVH at 15mm with no valvulopathy, preserved left ventricular ejection fraction (LVEF) with possible hypokinesis of the lateral wall rather poorly cleared. The echodoppler of the renal arteries was normal. Biological tests revealed chronic renal failure with a creatinine clearance according to the Cockcroft and Gault formula of 34ml/min, troponin was elevated to 87 μ/l. A fundus showed a Kirkendall stage III hypertensive retinopathy. The patient was treated with benzodiazepines (Valium injection), Nicardipine (Loxen) injection, analgesics (Paracetamol), and cerebral antiedematous drugs (Mannitol). The relay had been made by Perindopril (Coversyl), Amlodipine (Amlor) and Indapamide (Fludex), sodium valproate (Depakine) and acetyl salicylic acid (kardégic 75mg). The radio-clinical evolution was favorable. Brain MRI Flair sequence performed three months after the onset of symptoms did not show obvious lesions of vasogenic edema of the posterior cerebral fossa (Fig 2A and 2B). We retained the diagnosis of posterior reversible encephalopathy syndrome of malignant hypertensive origin (PRES).

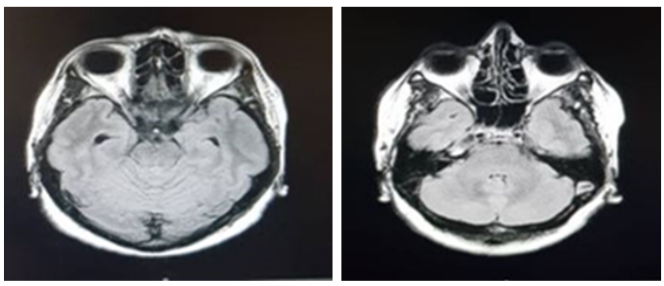

FIGURE 2: ControlFlair sequence brain MRI three months after the onset of symptoms showed novisible lesions (A and B).

2 | DISCUSSION

Posterior reversible encephalopathy syndrome (PRES) is a rarely reported neuroradiological condition (1, 2). It is characterized by diverse neurological clinical manifestations and vasogenic edema lesions on brain neuroimaging (3, 5). The diagnosis of PRES syndrome is based on clinical and radiological arguments. Clinical symptoms are varied associating headache (26-63%), confusion (67-90%), seizures (70-74%), visual disturbances (20-67%) and focal neurological deficits (5-15%) reported by the authors (1, 3, 6). However, all these

clinical manifestations are rarely found in the same patient, as was the case in our observation where our patient did not present focal neurological deficits. The absence of focal deficit in our observation could be explained by the absence of obvious ischemic or hemorrhagic lesions associated with vasogenic edema. The seizures are related to the cortical and diffuse localization of the cerebral edemas. The confusional syndrome would be related to the damage of the reticular activating system of the brain stem. The diagnosis of PRES syndrome in our observation was made by brain MRI Flair sequence. On the other hand, brain computed tomography (CT) allows the diagnosis by demonstrating patches of hypodensity in the posterior brain region reported by Fugate et al in 2015 (5). However, the same authors believe that the lesions observed on cerebral CT could therefore pose the problem of differential diagnosis with ischemia lesions. This said, brain MRI is the most powerful diagnostic tool of PRES syndrome in particular the Flair sequence due to its high specificity and sensitivity recognized by all authors (5, 6).

PRES syndrome is under-diagnosed, described at all ages from childhood to old age but with a peak in young adults and a female predominance due to the etiological aspects (6). Our patient was 52 years old, thus a young adult, which corresponds perfectly to the data reported in the literature. Vasogenic edema is predominantly localized in 98% of cases in the occipital and temporal regions according to the authors (3, 5). This localization in the posterior cerebral circulation is found in our observation. This can be explained by the low density of sympathetic innervation in the posterior cerebral region compared to the anterior region which is very dense in cervical ganglionic innervation. This innervation prevents excessive vasodilatation and consequently reduces the risk of cerebral hyperperfusion as described in the literature (6, 7). However, the authors believe that this hypothesis is true only in 30% of patients with PRES. On the other hand, vasogenic edema lesions are due to the rupture of the blood-brain barrier (BBB) by chronic hypertension or toxic endothelial damage (7, 8). Indeed, hypertension leads to a disruption of cerebral autoregulation, resulting in cerebral hyperperfusion, which would be responsible for vasogenic edema, but this is only observed in 70% of PRES cases. The second hypothesis would be related to endothelial damage of toxic origin which successively leads to vasoconstriction, vasospasm, ischemia with hypoperfusion and rupture of the BBB and then cerebral edema. However, all these hypotheses remain controversial according to the authors (7, 8).

There are several causative conditions of PRES syndrome. Malignant and chronic HAT remains by far the main cause in 70%. Autoimmune diseases such as systemic lupus erythematosus, glomerular nephropathy, hematological and renal transplants, infectious pathologies, eclampsia and preeclampsia, cytotoxic agents (3, 6, 9). In front of a non-specific neurological clinical picture in a patient under immunosuppressive treatment, it is necessary to know how to evoke a PRES syndrome and to carry out a cerebral imaging in particular an MRI for an urgent management (9). In our observation, chronic malignant hypertension remains the cause. This shows the importance of controlling malignant hypertension in order to avoid cerebral complications, namely PRES syndrome, but also and above all strokes with their morbidity and mortality consequences.

Therapeutically, PRES syndrome requires a symptomatic treatment shared by the authors (3, 4, 6). The diagnosis of PRES syndrome must be rapidly established and the treatment must not suffer any delay. The authors all agree that the therapeutic strategy depends on the cause and the clinical picture of PRES syndrome (8, 9). Indeed, preventive therapy would consist in avoiding or stopping all triggering or aggravating factors. Thus, patients under immunosuppressive drugs or chemotherapy should stop their treatment. The control of malignant hypertension is the essential element of the treatment of PRES syndrome (4, 9). Anticonvulsants are therefore the second component of the symptomatic treatment reported in the literature (4, 6, 9). This involves emergency antiepileptic drugs of the benzodiazepine type (Clonazepam or Rivotril, Diazepam or Valium). Valproic acid is a therapeutic option although there are no specific recommendations related to antiepileptics. Magnesium sulfate is recommended for pregnant women because of its vasodilatory effect. Cerebral anti-oedematous drugs (mannitol) and corticos-

teroids are sometimes administered depending on the clinical picture according to the authors (9).

The prognosis of PRES syndrome is generally favorable and all authors are unanimous (3, 9). According to these authors, neurological manifestations regress on the seventh day of antihypertensive and/or etiological treatment in 90% of cases, whereas an improvement in imaging is only possible in 15 days, but sometimes beyond a year, and even persistence of sequelae, especially in the case of cytotoxic edema, according to the authors (9). In our observation, regression of neurological symptoms was obtained on the 7th day and cerebral lesions after three months.

3 | CONCLUSION

The posterior reversible encephalopathy syndrome or PRES syndrome is a rare radio-clinical entity under-diagnosed. Several causes or factors are responsible for it, with malignant hypertension at the head. The clinical symptoms and radiological lesions are reversible if the diagnosis and treatment are not delayed. It is therefore important to think about it quickly and to perform a brain imaging, especially a Flair sequence MRI, in front of a polymorphic clinical picture associating a confusional syndrome, seizures and visual disorders in a context of chronic hypertension.

4 | CONFLICTS OF INTEREST

The authors declare no conflict of interest in relation to this article.

Authors' contributions

All authors contributed to the writing of this article

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