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Clinical case

Posterior reversible post-partum encephalopathy syndrome (PRES): a case report and review of the literature

Le Syndrome d'encéphalopathie postérieure réversible du post-partum à propos d'un cas et revue de la littérature

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Résumé

L'encéphalopathie postérieure réversible (EPR) est un syndrome neurologique aigu ou subaigu réversible. Il s'agit d'une pathologie rare dont le diagnostic est radio clinique associant une atteinte réversible du système nerveux central à une imagerie encéphalique typique. Compte tenu de l'intérêt diagnostic, thérapeutique et évolutif de ce syndrome, rare dans la pratique courante, nous rapportons l'observation d'une patiente âgée de 31 ans, sans aucun antécédent pathologique connu, admise au service des soins intensifs de la Maternité Issaka Gazobi de Niamey pour altération de la conscience de survenue subaiguë au lendemain du post-partum, dans un contexte d'éclampsie, chez qui l'examen clinique initial retrouve une altération de la conscience avec un score de Glasgow à 8 (E : 2, M :2, V : 4), une hémiparésie droite, une apyrexie. L'examen cardio-pulmonaire retrouve une hypertension artérielle avec tachycardie régulière. Le scanner cérébral d'admission retrouve un œdème cérébral diffus avec des plages hypodenses d'allure ischémique à localisation occipitale bilatérale. La prise en charge à consister initialement à une mise en condition, protection des voies ariennes supérieures, oxygénothérapie, le contrôle de la pression artérielle, une réanimation hydro-électrolytique avec protection gastrique, une anticoagulation faite d'énoxaparine en sous cutanée, et l'acétyle salicylate de lysine. L'évolution au sixième jour s'est faite vers un réveil complet de la patiente. Le diagnostic de PRES a été retenu sur la base des données cliniques, radiologiques couplées à l'évolution clinique spontanément réversible.

Mots-clés : Encéphalopathie postérieure réversible ; syndrome clinico-radiologique, œdème cérébral.

Abstract

Posterior reversible encephalopathy (PRE) is an acute or subacute reversible neurological syndrome. It is a rare pathology whose diagnosis is radio clinical associating reversible damage to the central nervous system with typical brain imaging. Given

the diagnostic, therapeutic and evolutionary interest of this syndrome, which is rare in current practice, we report the observation of a 31-year- old patient, with no known medical history, admitted to the intensive care unit of the Maternity Issaka Gazobi from Niamey for impaired consciousness of subacute onset the day after postpartum, in a context of eclampsia, in whom the initial clinical examination found impaired consciousness with a Glasgow score of 8 (E : 2 , M : 2, V : 4), right hemiparesis, afebrile. The cardiopulmonary examination finds arterial hypertension with regular tachycardia. The first cerebral CT scan found diffuse cerebral edema with hypodense areas of ischemic appearance with bilateral occipital location. Management consists of conditioning, protection of the airways, oxygen therapy, control of blood pressure, hydro-electrolytic correction with gastric protection, anticoagulation made of subcutaneous enoxaparin, and lysine acetyl salicylate. The evolution on the sixth day was made towards a complete awakening of the patient. The diagnosis of PRES was retained on the basis of clinical and radiological data coupled with the spontaneously reversible clinical course.

Keywords: Posterior reversible encephalopathy, clinico-radiological syndrome, cerebral edema.

Introduction

Posterior reversible encephalopathy (RPE) is an acute or subacute reversible neurological syndrome. This is a rare pathology whose diagnosis is radio clinical associating reversible involvement of the central nervous system with typical brain imaging (hypodense areas of ischemic appearance with bilateral occipital location on brain CT scan). RPE has various pathological origins, such as severe hypertension, eclampsia, organ transplantation, renal dysfunction, autoimmunity, sepsis, and chemotherapy. Although the pathophysiology is unclear, the same pathogenic mechanisms are shared. There is great variability in the clinical presentation of this syndrome and sometimes atypical imaging aspects. It combines several neurological signs such as headaches, visual disturbances, impaired consciousness, seizures, focal neurological deficits and bilateral cerebral radiological abnormalities predominating in the posterior regions which are classically reversible [1-4].

Considering the diagnostic, therapeutic and evolutionary interest of this visibly rare syndrome in current practice, we report the observation of a patient admitted to the intensive care unit of the Issaka Gazobi Maternity for consciousness disorder occurring the day after the postpartum period preceding an attack of eclampsia.

Clinical case

Mrs. K.H, 31 years old, right-handed, primpara with no known medical history, was referred for an eclampsia crisis the day after postpartum. The delivery was by basal route with a male newborn in apparent good health. The patient presented two convulsive crises including one during transport. The family reports a monitored pregnancy, the complaints frequently encountered during the last trimester of pregnancy were headaches, dizziness and visual disturbances and the transfer sheet notifies the occurrence at 8 hours postpartum of a convulsive crisis without aura and without biting of the tongue, then a calm post-critical coma.

The patient was admitted to the intensive care unit of the Issaka Gazobi Maternity Hospital in Niamey, the general examination found an unconscious patient, Glasgow at 8/15, Blood pressure: 184 /105 mmHg, tachycardia at 112 pulses/min, polypnea at 22 cycles/min, pulsed oxygen saturation (SPO2) at 94% on ambient air, and a temperature at 36.7°C. On physical examination, the pupils were of normal size, the patient presented with right hemiparesis. Cardiac auscultation found regular tachycardia and pleuropulmonary auscultation, the vesicular murmur was perceived by crackles. The abdomen was flexible with a uterine globe of good involution; the lochia was considered normal the day after postpartum. The

urine was clear with an hourly diuresis of 1.3ml/kg/h. The initial biological assessment (NFS, glycaemia, urea, creatinaemia, AST, ALT, sodium, potassium, thick drop) was normal. +++ proteinuria (see Table I). The cerebral CT scanner found areas of hypodense ischemic appearance with bilateral occipital localization with cerebral edema producing an aspect compatible with a syndrome of posterior reversible encephalopathy (PRES syndrome in its typical form). (Figure 1)

The initial management consisted of a release of the upper airways, oxygen therapy, control of blood pressure by calcium channel blocker of the Nicardipine type with an electric syringe and hydro-electrolytic resuscitation with gastric protection by proton pump inhibitor. The treatment was completed by an anticoagulation made of 40mg of enoxaparin per 24 hours subcutaneously, and an acetyl salicylate of lysine 100mg per day.

The evolution on the sixth day was made towards a complete awakening (Glasgow score at 15/15). The patient was discharged from the intensive care unit on the 9th day, under oral treatment: (lysine acetyl salicylate 100mg sachet: 1 sachet/day, Alpha methyl dopa 500mg: 1 tablet twice a day, Carbamazepine LP 200mg: 1 tablet per day. A control brain scan was performed on day 15 and returned to normal. (Figure 2)

In total, it was a picture of postpartum neurological disorder preceding an eclampsia crisis in which a first cerebral CT scan had found hypodense areas of ischemic appearance with bilateral occipital localization with cerebral edema and whose control carried out 15 days later had returned to normal (see Figure 2).

In view of the above, the diagnosis of posterior reversible encephalopathy syndrome was retained.

Table 1: Biological assessments

Balance sheets	**Results**	**Normal values**
Hemoglobin	13g/dl 9.38,000 / µL	11 – 16 g/dl 9.38,000 / µL
White blood cells Platelets count	283,000 / µL	150,000 - 450,000 / µL
Hematocrit	37.7%	37 – 46%
Thick drop	Negative	Negative
Urea	1.2 mmol/l	1.2 - 6.2 mmol/l
Serum creatinine	56 µmol /l	40 - 110 µmol /l
AST (SGOT)	21 IU/l	5 - 35 IU/l
ALT(SGPT)	25 IU/l	5 - 40 IU/l
Natremia	139 mmol/l	135 - 145 mmol/l
Serum potassium	4.5 mmol/l	3 - 6 mmol/l
Blood sugar	5.2mmol/l	l 3.9 - 7 mmol/l
Proteinuria	+++	Trace

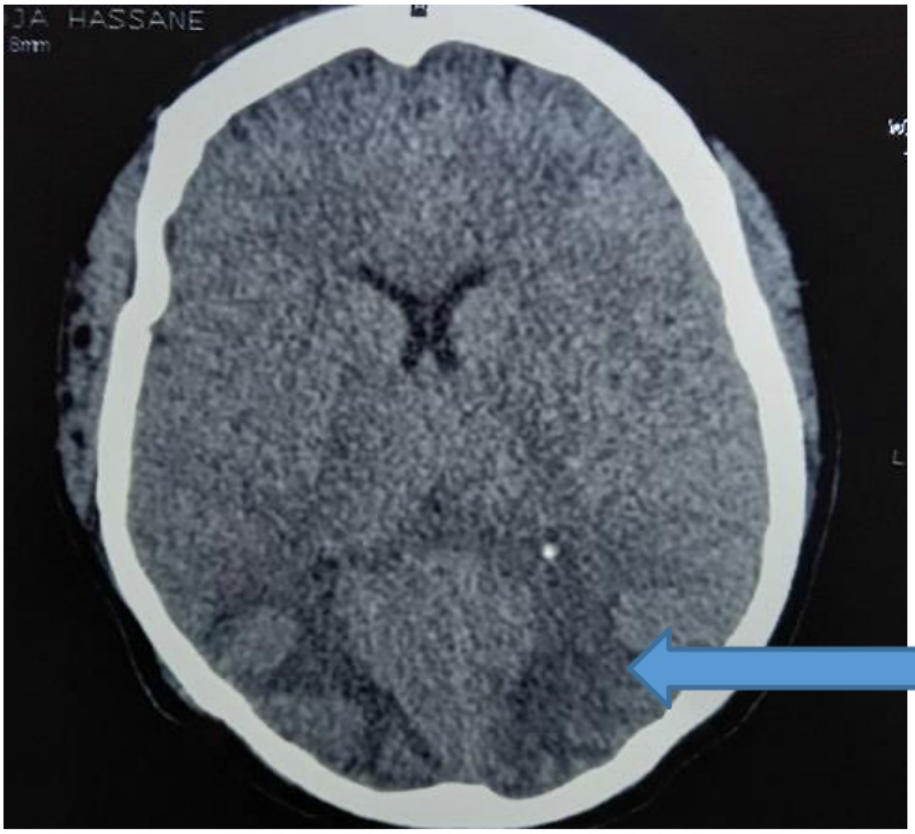

Figure 1: Cerebral CT showing bilateral occipital hypodense areas with cerebral edema

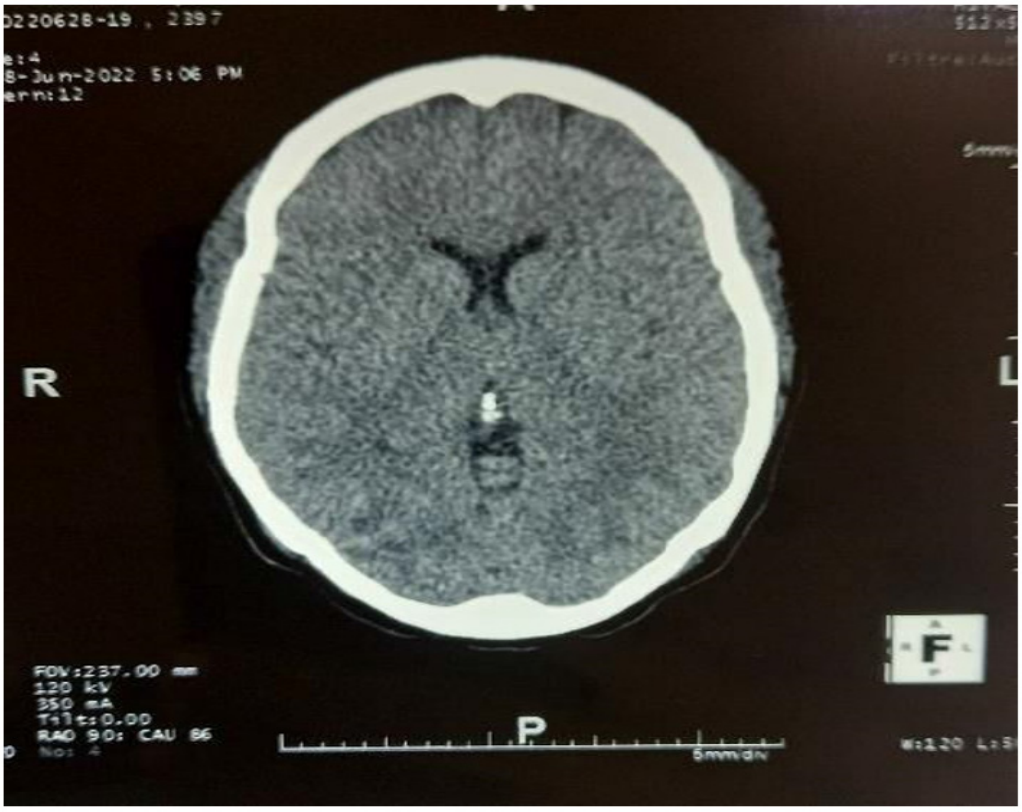

Figure 2: Control cerebral CT

Discussion

Posterior reversible encephalopathy is a rare disease, little known and probably remains underdiagnosed. The worldwide incidence is unknown. About 60% of cases of posterior reversible encephalopathy are secondary, most often postpartum. However, it can occur at the end of pregnancy. However, independently of pregnancy, certain hormonal treatments modifying estrogen and progesterone levels have been implicated in the occurrence of posterior reversible encephalopathy [5,6].

The pathophysiology of the cerebral edema of PRES (vasogenic edema) is not completely elucidated to date, however two theories are evoked in the literature. The first is the theory of cerebral hyperperfusion: a transient rise in blood pressure is classically found with cerebral hyperperfusion by overcoming the autonomic mechanisms regulation of cerebral blood flow explaining vasogenic cerebral edema; hence the neurological manifestations of rapidly favorable evolution as soon as the disease is mastered blood pressure. The second theory is that of reflex cerebral hypoperfusion (vasoconstriction, spasm, ischemia) to any stimulus of systemic origin. This results in endothelial dysfunction with vascular hyperpermeability, hence the installation of vasogenic edema [7,8].

The circumstances favoring the occurrence of RPE are numerous. The most common are arterial hypertension, which is classic and was the first factor described (hypertensive encephalopathy). Moderate to severe hypertension is observed in 75% of patients. Eclampsia, the link has often been established, including with normal blood pressure. Late onsets up to several weeks after delivery have been reported, chemotherapies such as cyclophosphamide can induce RPE syndrome even in the absence of other known risk factors, chronic renal failure and dialysis, autoimmune diseases have also been implicated [9,10].

Arterial hypertension with eclampsia were the risk factors identified in our patient. The peripartum seems to be a period at risk for the occurrence of an RPE syndrome. The clinical picture consists of neurosensory disorders such as headaches, confusion, agitation, convulsions or even a coma. The pathognomonic triad of the RPE syndrome is the association of a neurological picture, evocative radiological lesions and a rapidly reversible course as soon as the factor in question is mastered. Although abnormalities are sometimes visible on the cerebral scanner without injection (bilateral posterior hypodensity), magnetic resonance imaging (MRI) is considered the reference radiological examination today. It demonstrates localized cerebral edema, typically bilateral and symmetrical at the level of the subcortical white matter of the posterior (parieto-occipital) regions of the cerebral hemispheres. Narbone et al. noted that the radiological abnormalities of the brain are not exclusively posterior and that the reversibility of the lesions depends on the severity of these and rapid and adequate therapeutic management [11-13].

The therapeutic strategy of RPE syndrome depends on its etiology and the clinical picture. Stopping the triggering or aggravating factor represents the first therapeutic measure. There is no standardized therapeutic management. Many treatments have been described with discordant results. However, the control of hypertension is the essential part of the treatment; it uses the usual antihypertensive agents: calcium channel blockers (Nicardipine or diltiazem), beta-blockers (labetolol) and diuretics. The therapeutic objective is to maintain a mean arterial pressure between 105 and 125 mmHg, without reducing this pressure by more than 25% during the first hour. Magnesium sulphate has a vasodilating effect, which increases cerebral blood flow, thus preventing the appearance of ischemic lesions which are the cause of convulsive attacks [14].

Corticosteroids are the most commonly used drugs to fight against vasospasm and headaches. The association with an anti-oedematous treatment, in this case mannitol, must be discussed on a case-

by-case basis and can only be beneficial in certain situations. However, the spontaneous regression of the RPE syndrome makes it difficult to assess the efficacy of these treatments. In addition, the treatment of arterial hypertension must be cautious because it is advisable not to induce hypotension when there is already cerebral vasospasm reducing cerebral output. In the event of a seizure, anticonvulsant treatment should be instituted urgently. Benzodiazepines should be administered first line intravenously [15]. The evolution under early treatment is remarkably favorable. In the classic form, the clinical and radiological symptoms are completely reversible if the treatment is initiated early, however this reassuring evolution is not always found and necrotic- hemorrhagic complications have been reported [15,16].

Death is linked to neurological complications on a background of delayed diagnosis and management. Early diagnostic and therapeutic management conditions the evolution and prognosis of the RPE syndrome. The search for differential diagnoses is crucial as much as neuroprotection by controlling the factors involved. Elimination of the underlying etiology and regulation of blood pressure are the first steps in RPE syndrome but the reversible nature of RPE syndrome has been challenged recently based on new reports of permanent neurological damage and mortality as high as 15%. This syndrome, well understood by neurologists and neuroradiologists, is still unknown to clinicians [16].

Conclusion

Posterior reversible encephalopathy is a rare pathology, little known and probably remains underdiagnosed. The diagnosis is retained in front of the association of a neurological picture and typical radiological abnormalities. There are many etiologies and predisposing factors. Eclampsia remains the most probable cause in our patient.

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