ABSTRACT Posterior reversible encephalopathy syndrome (PRES) is a fairly rare complication, especially in children. Therefore, cerebral magnetic resonance imaging (MRI) is considered the reference examination to evoke this diagnosis. We report the case of a 7-year-old boy presenting with a reversible posterior encephalopathy syndrome secondary to malignant arterial hypertension complicating CKD on congenital nephropathy, which presented with a tonic-clonic seizure and loss of consciousness, with a BP of 190/130mmHg in an apyretic context. Imaging revealed characteristic white and grey matter oedema in the form of multiple areas of T2 hyper signal and cortico-subcortical temporal-occipital and frontal, of the caudate nucleus and bilateral lenticulars, of the brain stem and cerebellum in relation to the PRES given the clinico radiological context. The course of action was hospitalization in intensive care with stabilization and conditioning, and he was put on antihypertensive and anticonvulsant drugs. Rigorous monitoring was required in our patient, and the evolution was very favourable.

KEYWORDS Posterior reversible encephalopathy syndrome, malignant hypertension, CKD, congenital nephropathy, imaging.

Introduction

Posterior-reversible leukoencephalopathy, known as PRES (posterior-reversible encephalopathy syndrome), is a rare clinicopathological entity and even more so in children [1]. Clinical features include headache, nausea, visual disturbances, seizures, and disturbances of consciousness, associated with neuroimaging abnormalities predominantly in the parieto-occipital lobes [2]. Cerebral magnetic resonance imaging (MRI) is considered the gold standard for this diagnosis.

The causes are dominated by hypertension, eclampsia, chronic renal failure, treatment with immunosuppressive drugs, systemic and haematological diseases, and sepsis. One of the major characteristics of this syndrome is that it is reversible clinically and radiologically.

We report a 7-year-old boy with reversible posterior encephalopathy syndrome caused by malignant hypertension complicating chronic renal failure secondary to congenital nephropathy.

Case report

This is a 7-year-old patient, followed for congenital nephrotic syndrome, diagnosed by a renal biopsy at the stage of chronic end-stage renal failure. He is on peritoneal dialysis.

He presented 2 hours prior to admission with a sudden onset headache with vomiting, followed by generalised tonic-clonic seizures with post-critical coma. The patient was obtunded, with no sensory-motor deficits, meningeal stiffness or fever. Blood pressure measurement showed malignant hypertension at 190/130mmHg.
He was apnoeic with a correct capillary blood glucose level of 1.2g/l and oxygen saturation of 92% in free air.

He was then referred to our paediatric radiology department for brain imaging.

The brain scan revealed patchy, hypodense cortico-subcortical lesions, poorly limited to the temporo-occipital level, the basal ganglia, and to a lesser degree to the posterior cerebral fossa.

We completed the radiological work-up with a brain MRI. The images show multiple areas of T2 hyper signal and cortico-subcortical temporo occipital and frontal, bilateral caudate and lenticular nuclei, brainstem and cerebellum. Some of these lesions are of high signal in diffusion with low ADC in the brainstem. There are haemorrhagic petechiae in the right cerebellar, left frontal and right temporo-occipital hemispheres. (Figure 1, 2)

Faced with all these radiological signs in favour as well as the clinic, in particular, the antecedents of the patients of ESRD and the hypertensive peak, we concluded to a syndrome of reversible posterior leukoencephalopathy.

The patient was then admitted to the intensive care unit for stabilisation and conditioning with oxygen therapy and received anti-hypertensive and anti-seizure treatment.

The patient was admitted to the intensive care unit. He received oxygen therapy, correction of fluid and electrolyte disorders, and anti-hypertensive and anti-convulsant treatment.

The patient’s clinical evolution was favourable with normalisation of blood pressure and neurological function after 10 days of treatment.

The patient is currently in a good state of consciousness with a Glasgow score of 15/15 with no sensory-motor deficits or cognitive disorders. He is scheduled for a renal transplant.

Discussion

The syndrome of posterior reversible encephalopathy was first individualized in 1996 by Hinchey et al. from a series of 15 patients with common clinical and neuroradiological features [2].

The clinical picture is variable, ranging from headache with vomiting to a convulsive state requiring intensive care as in our patient [3].

The evolution is generally favourable and reversible, as in our patient’s case.

Various clinical contexts can precipitate the syndrome, hypertension being the most frequent cause (diastolic blood pressure above 120mmHg is usually observed). Other main conditions associated with PRES are haematological and autoimmune diseases, hemolytic-uremic syndrome, chronic renal failure, HIV infection, hepatitis C, immunosuppressive drug treatments, blood transfusions, and sepsis [4,5].

The mechanism of PRES is not well understood [7]. However, it is thought to be related to disruption of the blood-brain barrier with extravasation of intravascular fluid due to elevated blood pressure [8].

This is explained by an imbalance between vasoconstrictor and vasodilator substances, resulting in cerebral vasodilation with cerebral hyperperfusion, endothelial dysfunction, blood-brain barrier disruption and capillary leakage leading to cerebral oedema [3].

It should be noted that the deficit of autoregulation is predominant in the posterior regions because the perivascular sympathetic innervation of the posterior cerebral and vertebrobasilar circulation is less important than that of the anterior cerebral and Sylvian circulation [3].

Indeed, the syndrome’s cause in our patient appears to be multifactorial [6], combining chronic end-stage renal disease complicating congenital nephropathy and malignant hypertension.

From an imaging point of view, leukoencephalopathy lesions are detected by magnetic resonance imaging (MRI), which is considered the reference examination [3].

Cortico-subcortical parieto-occipital vasogenic oedema is found in the majority of cases [9], bilateral but often asymmetric. Best detected on T2 and FLAIR sequences as a hypersignal, with usually a hypo- or iso-signal on diffusion (DWI) and hyper signal on the corresponding apparent diffusion coefficient (ADC) maps. Restricted diffusion is seen in 11-26% of cases. It reflects cytotoxic oedema and may indicate progression to infarction and irreversibility, usually associated with poor outcomes [10]. Injected sequences are not mandatory and may show gyriform
or leptomeningeal contrast [11]. Sometimes the abnormalities go beyond the parietal and occipital lobes to involve the frontal and temporal lobes, and even the brainstem, the basal ganglia and even the cerebellum, as was the case in our observation [12]. The realization of ADC diffusion sequences could have a prognostic interest by differentiating vasogenic oedema usually encountered during the Pres syndrome from cytotoxic oedema, a marker of potentially irreversible lesions [2].

Recent studies have demonstrated vascular irregularities with focal and diffuse vasoconstriction and focal vasodilation, often producing a cord-like appearance on angiography.[13]. Other imaging techniques can be used, particularly Magnetic resonance spectroscopy (MRS) with a prognostic value in the acute phase, and brain scintigraphy most often shows images of hyperperfusion in the acute phase and hypoperfusion in the late phase [6].

Hemorrhagic complications may occur in the area of the brain parenchyma affected by oedema, such as microhemorrhage, hematoma with mass effect or meningeal haemorrhage [10].

In addition, children are considered more vulnerable to cerebrovascular dysfunction than adults because they have a narrower range of autoregulation of cerebral blood flow [14].

Prompt diagnosis and treatment of pres syndrome can prevent the onset of irreversible neurological signs and permanent sequelae [3]. As in our case, the evolution is usually favourable under appropriate treatment, with the disappearance of neurological signs in less than 15 days [2].

Nephrectomy is the best treatment for nephrogenic hyperten-

sion in children with a non-functioning unilateral kidney and a normal contralateral kidney [15], but unfortunately, our patient is in CKD and is scheduled for renal transplantation.

Conclusion

Pres is a radio-clinical syndrome characterized by the association of variable neurological signs and white matter and grey matter abnormalities preferentially affecting the posterior regions. The suggestive clinical context and the rapidly reversible nature of the clinical and radiological abnormalities suggest the existence of vasogenic cerebral oedema related to a vasculopathy. Imaging, particularly MRI, plays an essential role in diagnosing this condition. The knowledge of this syndrome should avoid unnecessary repetition of imaging controls when the clinical evolution is favourable [14].

Abbreviations

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<tr>
<th>Abbreviation</th>
<th>Definition</th>
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<tr>
<td>PRES</td>
<td>Posterior reversible encephalopathy syndrome</td>
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<td>MRI</td>
<td>magnetic resonance imaging</td>
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<td>CKD</td>
<td>Chronic kidney disease</td>
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<td>ESRD</td>
<td>End-stage renal disease</td>
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<td>HIV</td>
<td>human immunodeficiency virus</td>
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Conflict of interest

There are no conflicts of interest to declare by any of the authors of this study.

References


