Reversible posterior leukencephalopathy

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The patient has acute severe neurological symptoms associated to high blood pressure. Posterior reversible encephalopathy syndrome (RPLS) must be suspected and the cerebral tomodensitometry showed bilateral low density areas corresponding to the frontal, parieto-occipital white matter and to the internal capsula, thus the diagnosis of RPLS was confirmed (fig1). Treatment consisted in oxygen therapy, hydrosodic restriction, reducing the temperature and high blood pressure control using Fuosémide and Nicardipine.

Blood pressure was stabilized in four days. Neurological manifestations improved gradually with recovery in 5 days. Arterial hypertension was caused by renal scars of bilateral vesico-renal reflux. Hepatic and renal functions returned also to normal values. Two months later cerebral IRM was normal.

RPLS syndrome is a rare clinico-radiological disorder that occurs generally in the fourth decade of age (1). Clinical manifestations are similar to those of hypertensive encephalopathy. Symptoms are acute and appear in few hours or days (2, 3). Patients can complain of headaches, vomiting, visual disturbance, deterioration of consciousness and seizures. Disturbance of pupillary reflexes, papilledema and motor deficits are described as uncommon. Like our patient, seizure can herald the RPLS. In children, altered mental status and strokes are more encountered (4, 5).

Radiological findings of RPLS at cerebral TDM are images of hypodensity and at cerebral IRM areas of isointensity or hypointensity on T1 weighted images and of hyper intensity on T2 weighted images. Lesions are generally bilateral, relatively symmetrical and typically localized in the parieto-occipital white matter. Other regions like pons, cerebellum, thalamus, basal ganglia and medulla can be affected (1, 3). Calcarine and paramedian occipital lobes are always spared (5). With the radioimaging progress and the advent of the FLAIR (fluid attenuation inversion recovery) technique, the affection of the cortical and sub cortical areas can be proved (1, 6).

The main characteristic of this syndrome is its reversibility which is not systematic and depends on prompt and adequate management. Clinical recovery can occur as soon as 3 to 15 days (1, 2, 5). Radiological normalization can be observed in a delay of 4 to 15 days [1, 3,5).

Many circumstances can give rise to RPLS: high blood pressure (essential and secondary), toxemia, drugs (cyclosporine, interferon alpha, cisplatin...), HIV... (5).

Little is known about the histological course and the pathogenesis of RPLS. Vasogenic and cytotoxic oedema are implicated. RPLS is a disorder of cerebrovascular autoregulation with multiple underlying etiologies and it is commonly associated with increases in blood pressure. In the pediatric population, RPLS has been associated with chronic renal disease , the administration of chemotherapeutic agents , adrenocortical disease and Cushing's syndrome. It is thought that the sudden elevation in blood pressure leads to disruption of the autoregulatory mechanisms in the central nervous system, vasodilatation and vasoconstriction resulting in a breakdown of the blood-brain barrier. However, it is documented in some cases, particularly in the pediatric population, that BP may be only minimally elevated or fluctuant during the development of PRES(7)

The sympathetic innervations seems to be protective; which explains the frequency of lesions in the posterior region of the brain, where this innervations is rare (1, 6).

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Treatment of RPLS implies the treatment of the underlying aetiology (the control of the blood pressure, the withdrawal of the offending drug....) and a supportive management of complications (1,7, 9).

Conclusion:

PRES is a rare disorder in children. Early recognition of characteristic radiological features is key to the diagnosis as clinical symptoms may be non-specific or mimic other neurological illnesses.

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