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Review article

Childhood posterior reversible encephalopathy syndrome

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ABSTRACT

Posterior Reversible Encephalopathy Syndrome (PRES) is a clinicoradiologic syndrome characterised clinically by headaches, altered consciousness, visual disturbances and seizures and radiological changes which can resolve. However left untreated it can be fatal and not all cases are reversible. It can occur in many settings, the most common being hypertensive crisis. We discuss the clinical and radiological features of this increasingly diagnosed condition among children and current thinking on its pathogenesis. A brief case is used to highlight the variable presentation of PRES. PRES is often unsuspected by the clinician and radiologists may be first to suggest the diagnosis. Accurate assessment including blood pressure measurement, appropriate imaging and rapid treatment is required to avoid a devastating outcome.

Contents

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1. Introduction

Posterior reversible encephalopathy syndrome (PRES) has become increasingly recognized in recent years. Initially termed reversible posterior leuko-encephalopathy syndrome (RPLS), it was first reported by Hinchey in 1996 following his study of a group of pregnant women with pre-eclampsia who developed acute neurological deterioration due to hypertension and had acute bilateral parieto-occipital signal abnormalities on imaging.[1] The imaging changes were thought to represent ischaemia, however were found to be reversible when the hypertension was treated.

Advances in imaging have enabled radiologists to distinguish the cytotoxic oedema of ischaemia from the vasogenic oedema associated with PRES.[2] The imaging findings involve both white and grey matter and for this reason the original term was changed to PRES.[3]As reports of this condition in adults and children increase, a wide spectrum of clinical and radiological patterns has emerged. The more we learn of this syndrome the more we are led to question its title as it is often not exclusively posterior in its location nor is it always reversible.[4–8]

Currently, the definition of PRES is of a clinico-radiological syndrome which is characterised clinically by headaches, altered consciousness, visual disturbances and seizures and radiologically by posterior (parieto-occipital) imaging changes in the sub-cortical white matter predominantly.[7]

We present the clinical and radiological features of PRES, including an illustrative paediatric case with atypical imaging findings, and discuss current thinking regarding its aetiology.

2. Case study

A nine-year-old boy was referred for a neurology review with an 18 month history of frequent frontal headache, associated with vomiting and dizziness and relieved by lying in a dark room. Physical examination revealed an anxious boy with a blood pressure of 110/70 mmHg and a normal neurological examination including fundoscopy.

In his past history he had a urinary tract infection at the age of one. Significant renal impairment was detected on investigation at the time. Because of adverse social circumstances he had failed to attend follow-up appointments. He was subsequently placed in a foster home.

An MRI of brain was requested to outrule underlying pathology in view of his headaches and this was performed 6 weeks later (Fig. 1). This revealed abnormal signal in the cerebellum bilaterally on T2 FLAIR (FLuid Attenuated Inversion Recovery) sequences. On review in the clinic urgently that day his headaches were persistent. His blood pressure was 215/160 and swelling of his right optic disc and AV spotting was seen on fundoscopy. He was transferred urgently to the intensive care unit for intravenous anti-hypertensive therapy. Renal investigation revealed bilateral grade IV vesicoureteric reflux. His blood pressure was rapidly controlled and he was discharged 5 days post-admission with renal follow-up. A follow-up MRI Brain was performed 9 months later and it showed complete resolution of the cerebellar changes (Fig. 2).

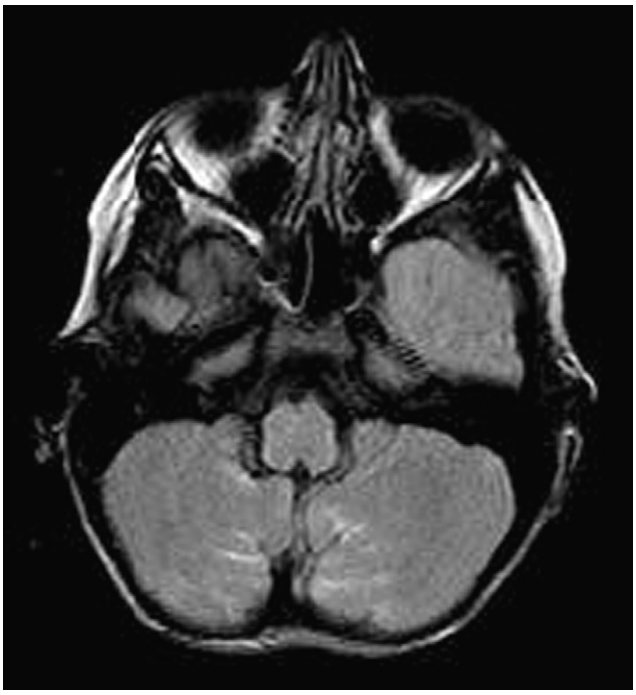

Fig. 1 – T2 FLAIR axial MRI demonstrates patchy increased signal intensity in the cerebellar hemispheres bilaterally. Diffusion imaging was normal as was the supratentorial brain.

3. Clinical features

PRES has been documented worldwide among a diverse patient population. Often not immediately suspected by the clinician, it may be first suggested by the radiologist. It has been described in

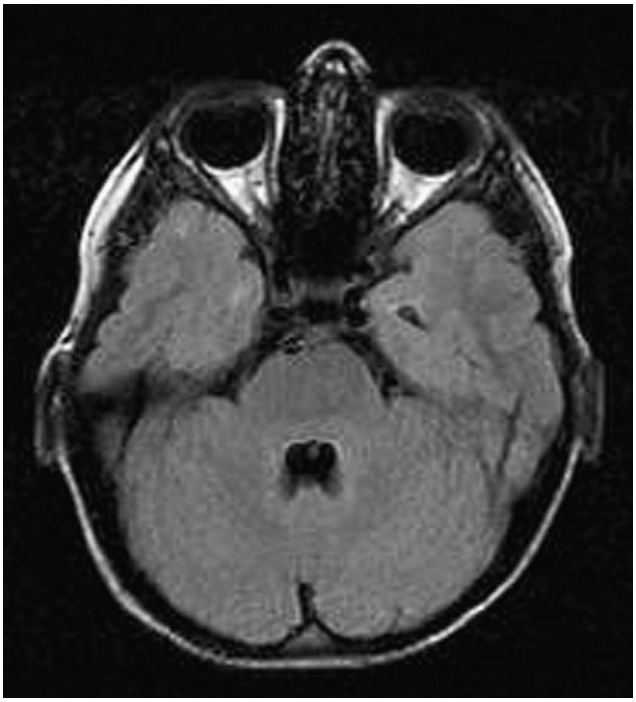

Fig. 2 – T2 FLAIR axial MRI on follow up is normal.