Case Report

An Unusual Complication following Ventriculoperitoneal Shunting

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INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) is a rare syndrome with an unknown exact aetiology. It is a radiological diagnosis. In this report we describe it’s occurrence following the insertion of a ventriculo-peritoneal shunt. We discuss the plausible aetiology of this unusual mechanism for PRES.

CASE REPORT

A very active 10-year-old male presented with a 2-week history of daily headaches. His background included asthma, atopy, and a proven allergy to cat hair. He was born full-term with no puerperal complications. Immunizations are up to date and he had achieved developmental milestones.

He described the headaches as pressure-like bilaterally, worse mid-morning, and again in the evening. There were no obvious triggers and he did not experience aura. The headaches did not wake him in the night and there was no associated vision or speech change. He began to have associated vomiting and an optician noted bilateral papilledema with splinter hemorrhages resulting in urgent referral to the local hospital.

On examination, there were no dysmorphic features or neurocutaneous stigmata. Head circumference was above the 99th centile for his age. Fundoscopy confirmed bilateral papilledema with splinter hemorrhages. Full neurological, cardiovascular, respiratory, and abdominal examination were otherwise unremarkable.

Computer tomography (CT) of the brain revealed panventriculomegaly and crowding at the craniocervical junction secondary to cerebellar tonsillar descent. Magnetic resonance imaging (MRI) of the neural axis excluded space-occupying lesions, venous sinus thrombosis, or spinal cord pathology. Dilated ventricles were seen with no evidence of transependymal edema. Intracranial pressure (ICP) monitoring confirmed pathological ICP (multiple B waves and mean ICP of 22 mmHg in a 24 h period). A right parietal ventriculoperitoneal (VP) shunt was inserted with a Codman-Hakim medium-low pressure valve. No immediate complications were encountered.

In the following days, the patient developed persistent low-pressure headache. This progressed to unresponsive episodes with rapid alternating eye movements associated with significant desaturations that were short-lived. CT scan of the brain showed smaller ventricular size and no change to the craniocervical junction. Following this, there was an episode of transient loss of vision followed by diplopia although detailed clinical examination of the eyes was normal including fundoscopy on this occasion. It was noted that the patient was tending to be hypertensive.

A repeat MRI scan of the brain shown in Figure 1 revealed a decompressed right lateral ventricle. There were new signal changes in the occipital lobes and cerebellar hemispheres. The diagnosis of posterior reversible encephalopathy syndrome (PRES) was made on clinical and radiological grounds. The patient was

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started on dexamethasone since the high pressures in the posterior fossa were thought to contribute.

Soon after, a quick recovery was observed with return to normal vision, no focal neurology, and well enough for discharge home. A CT scan just before discharge showed improved sulcal pattern with resolution of the cerebellar tonsil descent. At 3-month follow-up, the patient remains neurologically stable with a functioning VP shunt.

**Discussion**

PRES is a rare clinicoradiological entity whose exact incidence is not known. There is a trend toward the female population. MRI is crucial for the diagnosis and classically will display bilateral symmetrical hyperintense signal change on fluid-attenuated inversion recovery and T2-weighted imaging in the posterior parietal and/or occipital lobes. Lesions can appear hypointense on T1-weighted imaging with usually no contrast enhancement.

The true etiology underlying the condition is not well understood but is thought to relate to cerebral autoregulatory dysfunction with subsequent cerebral edema. Four theories have been postulated in literature:

- The “vasogenic” theory suggests that high systemic mean arterial pressures overcome cerebral autoregulation resulting in hyperperfusion of cerebral vessels. This leads to raised capillary hydrostatic pressures and subsequent breakdown of the blood–brain barrier resulting in edema. It is known from previous studies that the posterior circulation is more prone to hypertension-related damage since it carries less sympathetic neural innervation than vessels in the anterior circulation. However, this does not explain PRES in disease processes not associated with hypertension, such as autoimmune disease or septic shock.

- The “cytotoxic” theory hypothesizes that toxins or chemokines will cause endothelial damage resulting in cerebral edema. The “immunogenic” theory highlights the plausible role of the immune system. It suggests that mediators such as arachidonic acid, bradykinin, or nitric oxide activate T-lymphocytes which in turn result in vascular instability and downstream hypoperfusion.

- Similarly, the “neuropeptide” theory hypothesizes that potent vasoconstrictors such as thromboxane A2 cause vasospasm resulting in hypoperfusion of areas supplied in the arterial territory.

In literature, PRES is most commonly not only described in association with hypertension but also seen with infection (sepsis), pregnancy and neurosurgical pathology. In our patient, we believe that the development of PRES was directly related to the shunt insertion. It is plausible that brain shift following cerebrospinal fluid (CSF) diversion leads to a compromise of the posterior circulation. With the benefit of hindsight, the CT scan in Figure 1b showed less CSF space in the posterior fossa (quadrigeminal cistern and fourth ventricle increasingly effaced) suggesting an upward herniation of posterior fossa contents following decompression of the supratentorial compartment. This would be in keeping with the patient’s complaint of low-pressure headaches. Compression of the vertebrobasilar system or posterior cerebral arteries against tentorium cerebelli could have led to hypoperfusion of the areas of supply.

Alternatively, it is also plausible that the reduction of pressure within the supratentorial compartment improved circulation in the posterior circulation vessels, which may have been compressed against the tentorium. This could lead to hyperperfusion injury as suggested in the vasogenic theory described above.

This case is presented to educate and increase awareness of a rare and possibly under-reported entity and may have future management implications (e.g., care not to release high volumes of CSF during insertion of VP shunt). To our knowledge, this would be the first case report of PRES described in a pediatric patient following VP shunt insertion.

**Conclusions**

PRES is rare and a likely under-reported entity. Early diagnosis and management will result in better short-term outcomes.
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Conflicts of interest
There are no conflicts of interest.

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