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CASE REPORT

Refractory epilepsy associated with ventriculoperitoneal shunt over-drainage: case report

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Abstract

Epilepsy and intracranial pressure (ICP) can be interrelated. While shunt malfunction is recognized as a cause of seizures, shunt over-drainage is seldom reported as such. We report a child who had undergone ventriculoperitoneal shunt insertion at the age of 6 months following an excision of a left ventricle choroid plexus papilloma, who developed refractory epilepsy since the age of 3 years. An MRI showed small ventricles. The child presented with acute hydrocephalus due to proximal shunt malfunction at the age of 11 years and was treated with an endoscopic third ventriculostomy. Following the procedure, the seizures abated. Our case suggests that intractable epilepsy may be related to intracranial hypotension. Potential treatments for shunt over-drainage may be indicated even in the absence of classic over-drainage symptoms, in the presence of refractory epilepsy.

Keywords Hydrocephalus · Refractory epilepsy · Ventriculoperitoneal shunt · Over-drainage · Endoscopic third ventriculostomy

Introduction

Epilepsy and hydrocephalus may share common etiologies, such as intraventricular hemorrhage (IVH) of prematurity, traumatic brain injuries, subarachnoid hemorrhage, meningitis, and prior neurosurgical procedures for various indications [1–3]. Additionally, a ventricular shunt poses a risk for future epilepsy, with an incidence of 20% [4].

Shunt malfunction is another cause of epilepsy, caused by increased intracranial pressure [5]. Shunt over-drainage is seldom reported as a cause of epilepsy [6], possibly due to decreased awareness of the treating physicians. We describe a child with refractory focal epilepsy associated with shunt related over-drainage who recovered after shunt malfunction and an endoscopic third ventriculostomy (ETV).

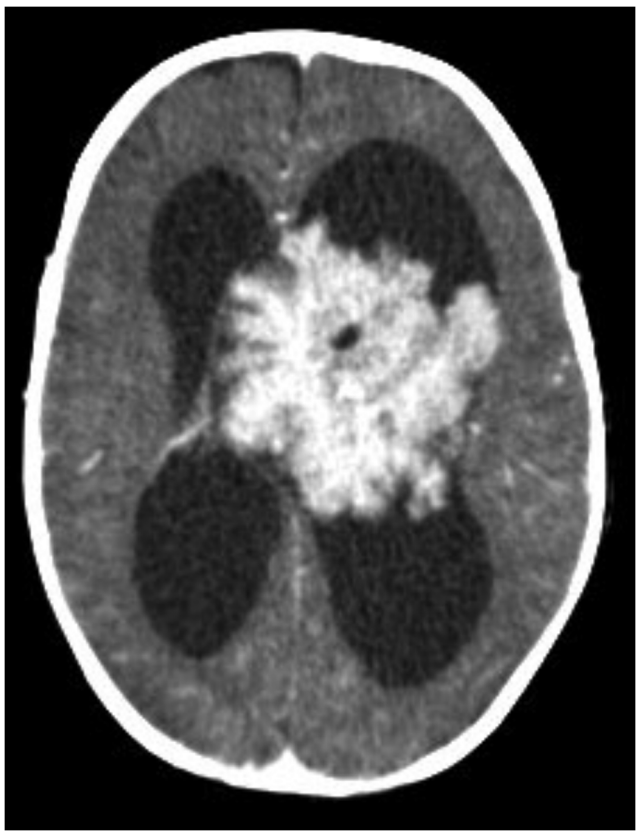

Fig. 1 Axial CT scan showing the large left ventricular tumor and hydrocephalus

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Case history

The patient was born at term following a spontaneous uneventful pregnancy and delivery, APGAR scores were 9/10, and birth weight was 2660 g.

At 6 months of age, he presented with a rapidly increasing head circumference, vomiting, irritability, somnolence, and mild right hemiparesis. Imaging showed a large intraventricular tumor (Fig. 1). The child underwent an uneventful left trans-cortical resection of the tumor, which was diagnosed as a choroid plexus papilloma (CPP). Six weeks later, a left ventriculoperitoneal shunt (VPS) was placed due to persistent hydrocephalus.

Following the procedure, he developed focal seizures, mostly during sleep. The seizures were initially well-controlled under phenobarbital.

He walked at the age of 1.4 years and talked fluently at 1.8 years. At the age of 3 years, seizures recurred and were focal with or without secondary generalization. The seizures occurred multiple times nightly.

Neurological examination at 4 years revealed left dominance, axial hypotonia, weak shoulder girdle, and joint hypermobility. An electroencephalogram (EEG) showed focal epileptic activity with infrequent left temporal spike-and-waves.

Over the years, the seizures were drug-resistant with a partial response to phenobarbital, primidone, topiramate, levetiracetam, clobazam, oxcarbazepine, valproic acid, and lacosamide.

The patients' cognitive ability was average, yet he had difficulty with executive functions and learning. On magnetic resonance imaging (MRI), there was no tumor recurrence and no signs of intracranial hypotension (such as subdural collections, venous engorgement, or dural enhancement), but the ventricles were small.

At the age of 7 years, a video-EEG revealed focal electrical status epilepticus during sleep (ESES) with constant epileptic activity over the left fronto-parieto-temporal area at electrode positions T3-T5 during seizures (Fig. 2). MRI showed a decrease in ventricular width (Fig. 3). There were no associated headaches, nausea, or vomiting, and following a multidisciplinary discussion, conservative treatment was continued despite the continuous epileptic activity.

The child presented at the age of 11 years with acute headaches and nausea. MRI showed obstructive hydrocephalus

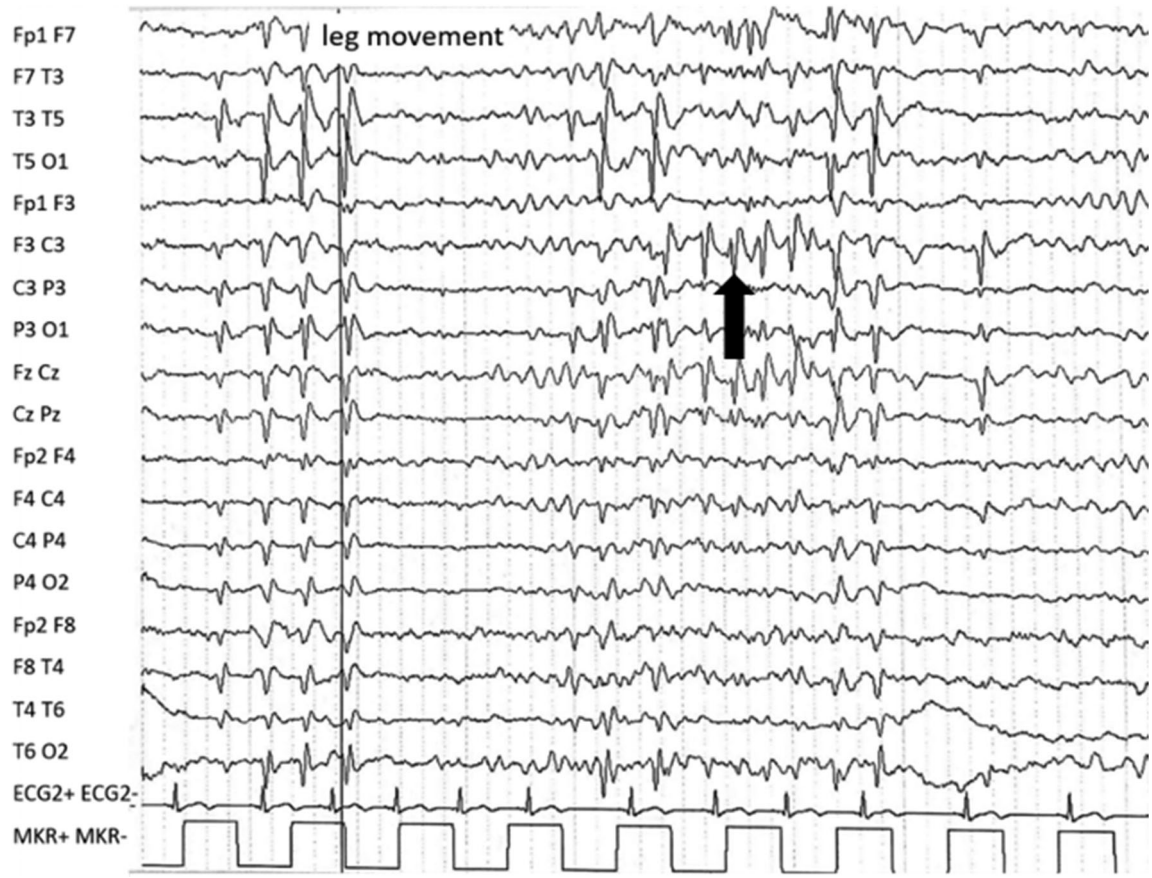

Fig. 2 Representative example of sleep EEG before shunt malfunction. Continuous spike wave activity present 70% of the time, most prominent over the left fronto-parietal area

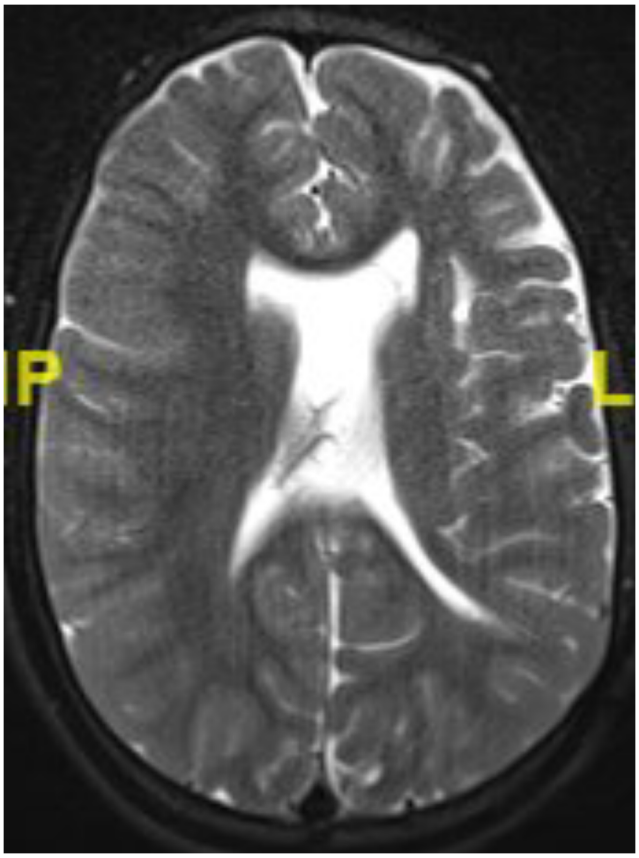

Fig. 3 MRI when shunt was functioning, axial T2 weighted image showing small lateral ventricles

(Fig. 4), associated with proximal shunt malfunction. The child underwent an uneventful ETV.

Over the course of 22 months since the surgery, the child has had no headaches and no seizures. Most anticonvulsants were discontinued, and he remains on monotherapy with low-dose primidone. EEG shows infrequent spike-and-wave epileptic activity over the left fronto-temporal area (Fig. 5). MRI shows a decrease in ventricular width following the ETV (Fig. 6), but they are considerably larger compared with the baseline, shunted ventricles. Currently, he continues to have difficulties related to his learning disabilities and attention deficit disorder.

Discussion

We report recovery of refractory epilepsy associated with over-drainage of hydrocephalus following shunt malfunction and ETV. This case supports the notion that changes in ICP, over-drainage, and epilepsy are interrelated. Following ETV for acute hydrocephalus, our patient recovered from refractory epilepsy that occurred while the ventricles were over shunted, implying the influence of intracranial hypotension on epilepsy control.

Epilepsy following low-grade tumor resections in children is rare [7]. In presence of a massive lateral ventricle tumor, and in presence of significant hydrocephalus in a baby with open sutures and fontanelle, a transcortical approach is preferred on

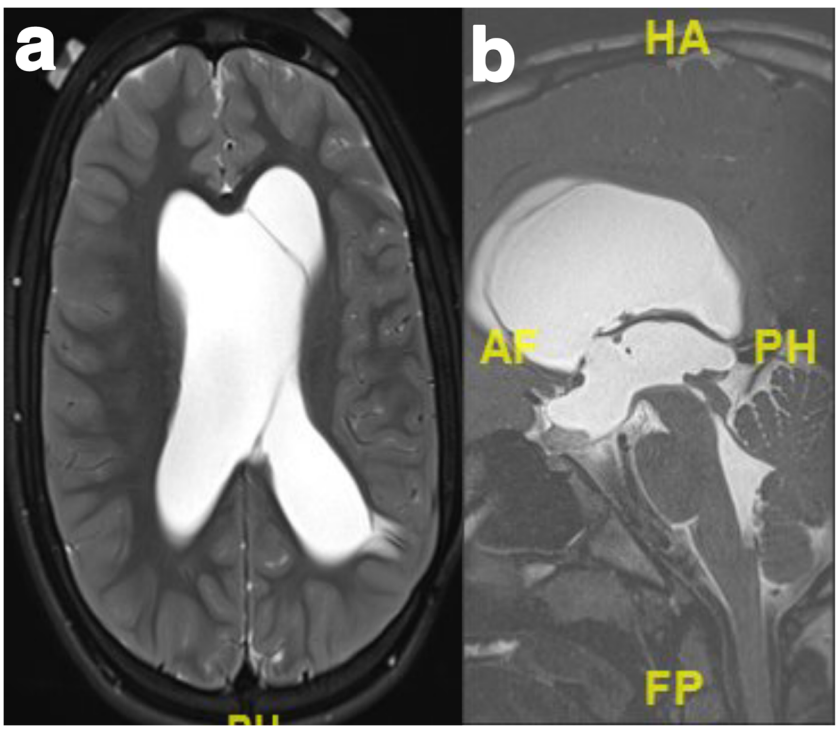

Fig. 4 MRI when shunt failed (before ETV), axial (**a**) and sagittal (**b**) T2 weighted images. Ventricles have enlarged, and there is an obstruction at the aqueduct

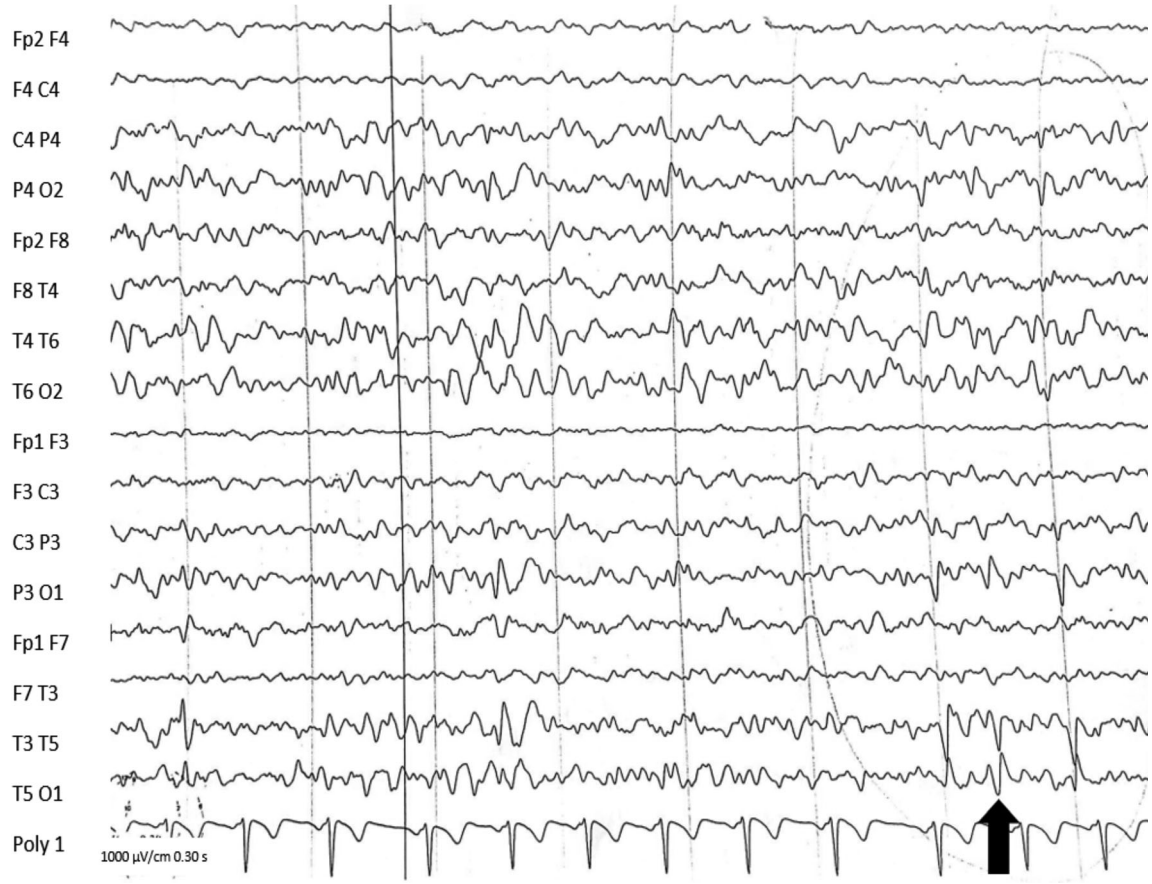

Fig. 5 Representative example of EEG following the ETV. EEG shows infrequent spike-and-wave epileptic activity over the left fronto-temporal area

the interhemispheric. The large overlap in etiologies causing both hydrocephalus and epilepsy may hinder a possible association between shunt-related problems and epilepsy [1].

Shunt malfunction leading to increased intracranial pressure may cause epilepsy too and is appreciated as such a cause [5]. Diagnosis is based on clinical symptoms, as

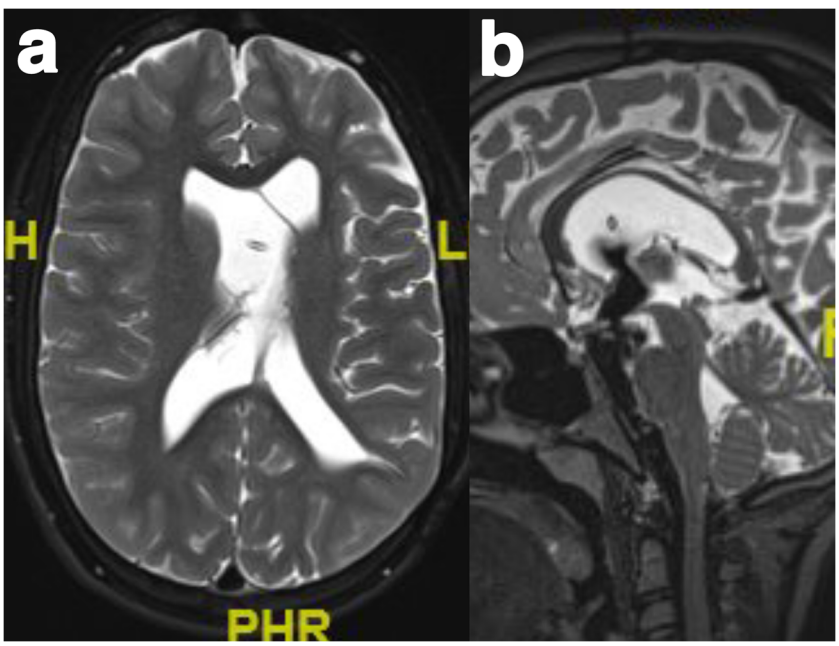

Fig. 6 MRI following the ETV, axial (**a**) and sagittal (**b**) T2 weighted images showing smaller ventricles, yet larger than when shunt was working, and a flow void through the ETV

well as ventricular enlargement [8, 9]. However, shunt over-drainage is more difficult to diagnose.

Saukkonen et al. [10] showed that children with VPS due to hydrocephalus who developed slit ventricles (SLV) had an increased rate of seizures compared with children in the non SLV group. EEG revealed a characteristic spike-and-wave activity, same as in our patient. Dramatic improvement of seizures occurred after shunt repair in the SLV-group. Since that study, only sparse information regarding over-drainage and epilepsy has been published. Agrawal et al. [11] described a 5-year-old girl with VPS and postural seizures, occurring only at upright position, implying a component of intracranial hypotension. Imaging showed decompressed ventricles, and seizures resolved following valve replacement. Uchida et al. [6] recently reported a 23-year-old patient who underwent shunt surgery during childhood who presented at the age of 20 years with slit ventricles and intractable seizures that decreased significantly after shunt replacement. They suggested that elevation in ICP could reduce certain seizures in shunted patients. Intracranial hypotension due to cerebrospinal fluid (CSF) leakage may also provoke seizures [12, 13].

Both obstructive hydrocephalus and shunting are known causes of brain microstructure changes, leading to increased brain stiffness [14, 15], possibly explaining the influence of minor changes in ICP on brain tissue function, and subsequent rise in seizures frequency as suggested formerly [6]. However, further studies are needed to confirm the mechanism behind this association.

Positional headaches, accompanied by radiological signs which may include small ventricles, pachymeningeal enhancement, and subdural collections, are suggestive of shunt over-drainage [16, 17]. However, often, the clinical symptoms are subtle and radiology may not show the typical findings [17]. Conversely, small ventricles might be misinterpreted as a well-functioning shunt [18].

Thus, in shunted patients with refractory epilepsy, we suggest a high level of suspicion that the epilepsy is related to over-drainage. If in doubt, we recommend using ICP monitoring for making the diagnosis of intracranial hypotension.

Potential treatments for shunt over-drainage include adding a shunt assist, upgrading the valve, or converting to ETV.

Conclusion

Although rare, shunt over-drainage may be associated with refractory seizures even in the absence of other typical clinical and radiological findings. These patients may have improved epilepsy control following surgical treatments aimed at reducing shunt over-drainage.

Compliance with ethical standards

Conflict of interest The authors declare they have no conflict of interest.

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