Bilateral Proptosis in a Child—A Rare Manifestation of Ventriculoperitoneal Shunt Obstruction

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Abstract

Keywords

- proptosis
- ventriculoperitoneal shunt
- obstruction

Ventriculoperitoneal shunt (VPS) obstruction may have a myriad of presentations. We reported a case of an 11-year-old girl presenting with acute, bilateral proptosis secondary to VPS obstruction. While neuroimaging was interpreted as unremarkable, fundoscopy revealed bilateral papilledema and lumbar puncture showed elevated intracranial pressure. Neurosurgical exploration demonstrated VPS valve obstruction and a new VPS was inserted. Postoperatively, she developed a recurrent extradural hematoma, which was initially evacuated and later managed conservatively. To our knowledge, this is the first report of bilateral proptosis secondary to VPS obstruction. This case highlights the value of key clinical findings and limitations of neuroimaging.

Introduction

Ventriculoperitoneal shunt (VPS) placement is a commonly performed neurosurgical procedure among children¹ with shunt obstruction being the most common associated complication. In a cohort of 1,719 patients, 56% developed at least one episode of shunt obstruction in 12 years following insertion.²

VPS obstruction tends to cause late shunt failure,³ with peak incidence within the first year after insertion in children,⁴ classically presenting with symptoms of headache, lethargy, nausea, and vomiting. However, atypical presentations may include visual impairment, cranial nerve palsies, seizures, hemiparesis, rigidity, and developmental arrest.⁵ Conversely, shunt infections, which are the second most common cause of shunt malfunction (8–15%), tend to cause early shunt failure.¹ Other shunt complications include overdrainage that can lead to subdural hematoma (SDH) formation (5%), abdominal pseudocyst formation (1–4.5%), and bowel perforation (0.1–0.7%).¹

Risk factors contributing to VPS obstruction remain elusive. Several theories regarding causative factors have been proposed, including clogging of the proximal catheter with

brain parenchyma during VPS placement, proximal catheter obstruction by pieces of choroid plexus when placed near the foramen of Monroe and occlusion due to the gradual accumulation of debris, such as blood and proteinaceous fluid.¹

In this article, we presented an unusual case of an 11-year-old girl with a VPS presenting with acute onset of bilateral proptosis, headache, and neck pain in the setting of falsely reassuring shunt reservoir evaluation and neuroimaging. Comprehensive neurological investigations were undertaken, with lumbar puncture (LP) revealing significantly elevated intracranial pressure (ICP). Neurosurgical exploration demonstrated an obstructed VPS valve, and subsequently a new VPS was inserted. Following placement of the new VPS she developed an extradural hematoma (EDH), which is a rare but known complication of VPS insertion, and the EDH was managed accordingly.

We performed a literature review of atypical presentations of VPS malfunction, rare causes of proptosis, and discuss the value of key clinical findings along with limitations of neuroimaging in the management of this case. We also briefly discussed the pathophysiology and preventive measures for EDH following VPS insertion.

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Case Description

An 11-year-old girl with past medical history significant for congenital communicating hydrocephalus had a right parieto-occipital VPS insertion at 6 months of age, and later underwent revision to a left parieto-occipital VPS at 20 months of age for concerns of shunt obstruction. She remained in a state of good health until she presented to our institution with acute onset headache, blur vision for 5 days, and bilateral proptosis for 2 days. The patient's headache was associated with neck pain, but no vomiting, photophobia, fever, or neurological deficits were reported. In the emergency department, she had a seizure characterized by blank staring and stiffening of all four limbs for a duration of 1 minute. Urgent computed tomography (CT) of the brain showed no focal abnormality or ventricular dilatation with a left parieto-occipital VPS in place, unchanged in position compared with previous imaging obtained 10 years prior.

She was admitted to evaluate for a broad differential diagnosis including VPS malfunction, meningoencephalitis, and cerebral venous sinus thrombosis. Neurosurgery, neurology, and ophthalmology services were urgently consulted. Physical examination was significant for intermittent drowsiness, bilateral proptosis, reactive but sluggish pupils, bilateral abducens nerve palsy, bilateral chronic papilledema with neck stiffness, and positive Kernig's sign. Hence, the clinical findings were consistent with elevated ICP. Of note, the VPS was palpable with no suggestion of shunt fracture. Aspiration of cerebrospinal fluid (CSF) from the VPS reservoir yielded 3 mL of clear fluid without resistance and did not suggest high

pressure within the system. The shunt reservoir also refilled easily after being emptied by compression. Although CSF analysis was not suggestive of meningoencephalitis, empirical intravenous antibiotics were administered. Magnetic resonance imaging (MRI) and magnetic resonance venography of the brain and orbits were both reported as normal (►Fig. 1). There was also no evidence of cavernous sinus lesions or cerebral venous sinus thrombosis.

On the second day of admission, she developed areflexia, with absent biceps, knee and ankle reflexes bilaterally, but bladder and bowel functions were preserved. At this point, other differentials considered were Miller Fisher syndrome or autoimmune encephalomyelitis. However, MRI of the spine and nerve conduction studies were unremarkable. She then developed acute hypoxic respiratory failure due to hypoventilation while asleep despite initiation of noninvasive ventilation, and so the patient was intubated and placed on mechanical ventilation. In view of concerns for progressive autoimmune encephalomyelitis, intravenous immunoglobulin was administered, and LP was performed for further autoimmune, metabolic, and microbiological evaluation. Opening pressure during the LP measured 50 cmH₂O that decreased to 35 cmH₂O after draining 10 mL of CSF. Emergent repeat head CT revealed stable findings, with no evidence of ventriculomegaly or elevated ICP. Shunt series radiography also did not reveal any fractures or discontinuity in the VPS.

Serial ophthalmological exams demonstrated evidence of bilateral optic atrophy with new onset flame shaped hemorrhages, but normal intraocular pressure. Despite counselling by the medical team in recommendation of shunt exploration

Fig. 1 Axial T2-weighted magnetic resonance imaging scans of the brain and orbits obtained on admission demonstrate (A) a left parieto-occipital ventriculoperitoneal shunt in situ, with no evidence of hydrocephalus, space-occupying lesions, or mass effect. (B) Of note, there is increased perioptic nerve cerebrospinal fluid space and posterior globe indentation from the optic nerve sheaths, suggestive of intracranial hypertension.

or invasive ICP monitoring, the child's parents were reluctant to proceed. The patient subsequently failed a spontaneous breathing trial and repeat LP yielded markedly elevated ICP as high as 48 cmH₂O. Given the persistently elevated ICP, hypoventilation, and progression of ocular findings, the patient's parents consented to VPS exploration. Intraoperatively, the VPS valve was obstructed with no proximal flow of CSF. A new left frontal VPS was inserted together with a Codman Certas programmable valve (initially set at level 3, corresponding to 8 cmH₂O),⁶ which was used in anticipation of potential issues of over- or under-drainage. Postoperatively, she tolerated extubation, began neurorehabilitation, and had resolution of headache and hypoventilation. She also regained normal reflexes by the 5th postoperative day. However, her vision had deteriorated significantly, with poorly reactive pupils and inconsistent perception of light.

Interval MRI of the brain performed 1 week postoperatively revealed a 2.8 cm right EDH with 1 cm leftward midline shift (►Fig. 2). This prompted an emergency craniotomy and EDH evacuation. The patient responded well, cerebral compression was decreased, and the VPS valve setting was reprogrammed from 3 to 5, corresponding to 15 cmH₂O. The next day, postoperative head CT showed a small, residual, extradural collection measuring 1.7 cm with residual midline shift of 0.3 cm and hence the valve setting was increased from 5 to 6, corresponding to 18 cmH₂O. The next interval MRI brain performed a week after craniotomy showed right EDH reaccumulation measuring 2.4 cm with unchanged leftward midline shift of 0.3 cm. No hydrocephalus was appreciated and the VPS position was unchanged. After

discussion with her parents, a decision was made to manage the EDH conservatively, but the valve setting was increased further from 6 to 7, corresponding to 20 cmH₂O. Thereafter, two more interval head CT scans revealed stable EDH with minimal midline shift.

Over the next few weeks, she continued to receive neuro-rehabilitation and experienced gradual improvement in vision, from only perceiving light to being able to read. Serial examinations revealed improved color perception, intact ocular movement bilaterally, resolution of proptosis, and resolution of optic disc swelling; however, visual fields were restricted. Repeat MRI of the brain 6 weeks after EDH evacuation demonstrated resolution of the residual EDH and she was discharged after a 2-month hospital stay. She remains well and is coping well in school despite residual visual field restriction.

Discussion

Proptosis in a child is rarely a feature of elevated ICP and is typically secondary to orbital cellulitis or malignancy, with less frequent causes including vascular malformations or inflammatory lesions.⁷ To our knowledge, our case is the first description of a child presenting with proptosis secondary to VPS obstruction. Our case report highlights the value of relying on key clinical signs and the limitations of neuroimaging and shunt examination in the recognition of VPS obstruction. The development of papilledema typically lags behind the onset of elevated ICP and has been reported to be present in 94% of patients with chronically elevated ICP.⁸ This

Fig. 2 Coronal (A) and axial (B) T1-weighted magnetic resonance imaging scans of the brain obtained 1 week after surgical placement of a new left frontal ventriculoperitoneal shunt demonstrate a right extradural hematoma indenting the right cerebral hemisphere with midline shift to the left.

indicates that papilledema should always be interpreted as raised ICP until proven otherwise, even in the presence of atypical manifestations and the absence of common symptoms such as vomiting. In the context of patients with a VPS, shunt obstruction must be excluded while evaluating for other causes of raised ICP and surgical exploration must be strongly considered.

Undoubtedly, neuroimaging is the most valuable investigation in assessing for VPS obstruction, with ventricular enlargement being the most common feature.⁹ Other findings of shunt obstruction on CT imaging include cortical sulci effacement, compressed or obliterated basal cisterns, and periventricular edema due to transependymal CSF absorption.⁹ Of note, while neuroimaging can confirm the diagnosis, a negative scan is insufficient to rule out VPS obstruction. Ventricular dilatation may be absent on neuroimaging in 10 to 30% of cases of VPS malfunction.^{10,11} In addition, a recent, retrospective study involving 95 children with VPS malfunction reported that 45% (27 of 60) of their cohort with VPS obstruction had normal CT imaging.⁹ This is also supported by the observation that up to 15% of shunted pediatric patients have such profound alterations in brain compliance that their ventricles do not enlarge in the event of shunt failure or raised ICP.¹² It has also been hypothesized that VPS insertions at a younger age may be responsible for early sutural closure and subependymal gliosis, which makes the ventricular system resistant to subsequent dilatation when a shunt malfunction occurs, thus being responsible for the normal appearance of ventricles on CT imaging.¹³ In comparison, MRI may be useful to detect more subtle signs.¹⁴ In particular, MRI of the orbits may reveal features of raised ICP such as (1) enlargement of the optic nerve sheath, (2) flattening of the posterior sclera, (3) protrusion of the optic papilla into the globe, and (4) tortuosity of the optic nerve.¹⁵ Although initially reported as normal, a closer inspection of the MRI orbits of our patient revealed increased perioptic nerve CSF space and flattening of the posterior aspect of the globe, which was in keeping with intracranial hypertension. Tapping a VPS reservoir may also provide limited value in assessing shunt function. In a prospective pediatric study of VPS tapping prior to surgical exploration, 93% of VPS taps with poor or no CSF flow were found to have proximal catheter obstruction. Among the patients with good CSF flow and high opening pressure on VPS tapping, 92% and 8% were found to have distal and proximal catheter obstructions, respectively. However, among patients with good CSF flow and normal or low opening pressure on VPS tapping, 64% and 7% were found to have proximal and distal catheter obstructions, respectively.¹⁶

Neuro-ophthalmological manifestations have been reported to occur earlier than classic clinical or radiological signs of raised ICP.^{17,18} This may account for our patient presenting with blur vision, proptosis, papilledema, and bilateral abducens nerve palsy in the absence of vomiting or ventriculomegaly. In addition to papilledema and optic atrophy, other neuro-ophthalmological manifestations of VPS malfunction may include impaired visual acuity, transient or permanent blindness, visual field deficits, and

dorsal midbrain syndrome.^{14,19} Rare presentations of VPS malfunction associated with possible hypothalamic, midbrain or lower brainstem compression include manifestations such as seizures, lower cranial nerve palsies, hemiparesis, Parkinsonian-like rigidity, akinetic mutism, reversible opisthotonos, and central diabetes insipidus.¹⁹ In our patient, we postulate that brainstem compression not appreciable on neuroimaging may have resulted in areflexia and sleep-related hypoventilation. To our knowledge, there are no reports of proptosis secondary to VPS obstruction, although proptosis has been described in a few cases of chronic intracranial hypertension or hydrocephalus.^{14,20} In 1978, Osher et al reported a case of a 19-year-old male who presented with right-sided proptosis, papilledema, visual loss, and personality change. He was found to have aqueductal stenosis, resulting in a dilated third ventricle that extended into the parasellar area. The authors postulated that compression of the cavernous sinus obstructed orbital venous return and resulted in proptosis.²¹ A case report from 1975 involved a 32-year-old man with raised ICP secondary to a chronic SDH who underwent LP, which then precipitated acute tentorial coning and aggravated intracranial hypertension. This contributed to intracranial venous congestion, with resultant obstruction of orbital venous return and the subsequent development of bilateral proptosis.²² In 2012, another case report involved a 24-year-old woman with idiopathic intracranial hypertension who presented with unilateral proptosis and dural ectasia of the optic nerve.²³

A large reduction in ICP following over-drainage is known to cause SDH, particularly in the setting of chronic hydrocephalus.²⁴ Excessive CSF drainage results in brain parenchyma sinking away from the skull after ventricular pressure is reduced, stretching and potentially tearing bridging veins. It is likely that the risk of postdecompression SDH is proportional to the duration of exposure to raised ICP and the magnitude of change in ICP pre and postdecompression. However, EDHs rarely occur in such cases owing to the adherence of dura to the skull.²⁵ Collapse of the brain parenchyma may exert traction on the middle meningeal artery and its branches, contributing to vascular tears and EDH formation.²⁶ Seyithanoglu et al suggested that in some patients, skull-dura adhesions may be less significant than dura-arachnoid adhesions. This could contribute to the formation of an EDH rather than SDH if over-drainage occurs.²⁴ Post-VPS EDH tends to occur at the frontoparietal region, which may be due to loose fixation of the dura to this segment of the cranial vault.²⁷ With this in mind, the following few precautions during VPS insertion may help to prevent EDH/SDH: (1) the ventricular catheter should be carefully applied with minimal CSF spillage (2) a programmable pressure valve may be inserted with relatively high pressure settings used initially, followed by a gradual reduction in pressure over time; (3) any change in patient posture, especially the return to an upright position, should be slow and gradual; and (4) routine follow-up neuroimaging should be obtained to guide titration of the programmable valve.^{28,29} Although her postoperative

course was complicated by the development of an EDH with significant midline shift prompting surgical evacuation, the use of a programmable valve in our patient allowed subsequent titration of pressure settings, which facilitated resolution of residual EDH without further surgical intervention, favorable neurological recovery, and improvement in visual function.

Conclusion

VPS obstruction may manifest with a myriad of clinical presentations, including visual abnormalities, seizures, hemiparesis, rigidity, and developmental arrest. We report a case of bilateral proptosis in a patient with VPS obstruction and highlight the value of key clinical findings such as papilledema and subtle findings on MRI to determine the presence of raised ICP. This case illustrated the importance of considering VPS obstruction despite an atypical clinical presentation, falsely reassuring neuroimaging and negative shunt examination. In scenarios of high clinical suspicion for VPS obstruction, surgical exploration must be considered to evaluate shunt function. The extent and duration of raised ICP are also an important consideration in approaching VPS revision, so as to mitigate the risk of postoperative EDH or SDH. Programmable valves may be used effectively to titrate shunt flow according to clinical need.

Authors' Contributions

All authors participated in creating the content of the case report, editing, and providing final approval for submission. No undisclosed authors contributed to the case report.

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Conflict of Interest

None declared.

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