CASE REPORT

Reversible Vision Loss Due to Transependymal Oedema of the Optic Apparatus Secondary to Ventriculoperitoneal Shunt Malfunction

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ABSTRACT

A 29-year-old male with a history of pilocytic astrocytoma status post-multiple ventriculoperitoneal shunt (VPS) revisions presented with vision loss, ocular motor deficits, and headaches. His shunt was revised but he experienced persistent vision loss despite resolution of his headaches and diplopia. Magnetic resonance imaging (MRI) of the head revealed hydrocephalus and T2 hyperintensity consistent with transependymal oedema surrounding the midbrain, pons, and optic apparatus. His shunt was again revised, but was complicated by an infection that required removal. He underwent an endoscopic third ventriculostomy and an external ventricular device was temporarily placed. Repeat MRI revealed marked improvement of the transependymal oedema and hydrocephalus. His vision returned to baseline and remained stable at 6-months follow-up.

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Introduction

Ventriculoperitoneal shunt (VPS) malfunction may occur secondary to infectious or mechanical etiologies. Shunt infections occur in approximately 11–19.7% of patients¹ whereas mechanical complications, including shunt obstruction, fracture, or displacement, occur in up to 45% of patients who require revision.² The most common symptoms are related to increased intracranial pressure (e.g., headache, nausea, and vomiting or blurred vision from papilloedema). Less commonly, involvement of the optic apparatus, including the optic nerves, optic tracts, optic chiasm, or optic radiations can result in visual field defects and decreased visual acuity (VA).¹ We report an unusual case of acute onset vision loss due to transependymal oedema secondary to VPS malfunction that resolved clinically and radiographically after revision of the shunt and remained without recurrence at 6-month follow-up. To the best of our knowledge, this is the first such case in the transependymal oedema involving the visual pathway secondary to

VP shunt malfunction in the English language ophthalmic literature.

Case presentation

A 29-year-old male presented to an ophthalmologist at an outside hospital with a 3-day history of vision loss, ocular motility deficits, headaches, nausea, and vomiting. His past medical history included cerebellar pilocytic astrocytoma status-post resection at age 3, with multiple VPS placements and revisions, most recently at age 13. Circumstances surrounding previous VPS revisions are unknown. Serial imaging studies showed stable post-treatment ventriculomegaly without transependymal oedema and no recurrent or residual cerebellar astrocytoma. His past ocular history included long standing primary position and gaze evoked nystagmus with mild amblyopia OD. He has a persistent bilateral cranial nerve VI palsy resulting in a mild esotropia that worsened secondary to VPS malfunction. His baseline VA is 20/40 in the right eye (OD) and 20/25 in the left eye (OS). The remainder of his history was unremarkable.

Ophthalmology examination at presentation revealed VA of count fingers (CF) in both eyes (OU) and a slow amplitude uniplanar horizontal nystagmus, mildly dampened by the chin up position. 24–2 Humphrey visual field (HVF) testing revealed a mean deviation of -26.26 dB and an inferior altitudinal and superior paracentral defect OD. The reliability indices revealed fixation losses of 4/15 and a false positive response error of 0% OD. Testing in the left eye revealed a mean deviation of -16.27 dB and a superior altitudinal defect. The reliability indices revealed fixation losses of 0/0, a false positive response error of 3%, and a false negative rate of 53% OS. He was subsequently admitted to the hospital for further workup. Computed tomography (CT) of the head revealed moderate dilation of the lateral and third ventricles and a VPS catheter. VPS malfunction was suspected and the valve was replaced with a programmable Strata II valve set at 1.5, and the distal catheter was replaced. His headaches and diplopia improved; however, vision loss persisted. He was discharged with recommended neuro-ophthalmology follow-up. On neuro-ophthalmologic exam one week later, VA remained 20/CF OU. He was noted to have vertical gaze palsy, convergence-retraction nystagmus, “sun setting” eyes with both eyes in the downgaze position, and bilateral esotropia of 30 prism diopters (PD) that was worse than his baseline chronic esotropia of 20 PD. No optic nerve oedema was found. Cup-to-disc ratio was 0.3 OU. There was a mild optic atrophy inferiorly and temporally, average retinal nerve fibre layer thickness on OCT was $87\text{ }\mu\text{m}$ OD and $84\text{ }\mu\text{m}$ OS. The patient was admitted to neurosurgery. VPS series suggested shunt disconnection/malfunction. Magnetic resonance imaging (MRI) revealed ventriculomegaly and atypical hyperintense T2 signal in the white matter surrounding the midbrain, pons, and optic apparatus. Transependymal oedema was present in both hemispheres, extending into the visual pathways (Figure 1). Due to concern for demyelinating disease, testing for serum anti-myelin oligodendrocyte glycoprotein (MOG) antibodies was requested and returned positive with a titre of 1:20 while aquaporin-4 antibodies were negative. The VPS was replaced. Two days post-operatively, VA measured at near

was 20/25 OU. 24–2 HVF testing revealed a mean deviation of -8.16 dB OD and -18.9 dB OS. Unfortunately, he was readmitted 6 days after with an infected shunt. CSF cultures grew methicillin-resistant staphylococcus aureus and antibiotics were administered. The shunt was removed. He underwent an endoscopic third ventriculostomy (ETV) and an external ventricular device was temporarily placed, then removed. A repeated MRI revealed marked resolution of the transependymal oedema and hydrocephalus (Figure 1). Repeat testing for MOG antibodies was negative. The patient’s vision recovered to baseline by 6-month follow-up after shunt removal. At 12-month follow-up, the patient’s vision remained stable and the MRI demonstrated complete resolution of the transependymal oedema. 24–2 VF was normalised on repeat testing and revealed a mean deviation of -6.74 dB OD and -3.79 dB OS.

Discussion

VPS failure occurs in up to 40% of patients within the first post-operative year.² Common symptoms include headache, nausea, and vomiting and rarely, visual field defects, and blindness.³ Blindness may be transient or permanent and usually occurs secondary to papilloedema, leading to ischaemia and atrophy of the optic nerve. Posterior visual pathway (e.g., homonymous hemianopsia, cortical visual loss) involvement is less common.¹ Our patient experienced the common symptoms of shunt failure in addition to decreased VA, diplopia, vertical gaze palsy, and nystagmus.

The diagnosis of VPS malfunction may be difficult due to insignificant findings on imaging or a lack of increased intracranial pressure symptoms.³ Shunt malfunction may be diagnosed after complications arise, such as the development of interstitial oedema or hydrocephalus. Fluid accumulates due to the drainage failure from the parenchyma into subependymal veins, resulting in oedema. Interstitial oedema may be associated with demyelination; however, the mechanism has not been described. Additionally, hydrocephalus can disturb nearby structures, such as the optic radiations, leading to axonal degeneration.⁴

Figure 1. Magnetic resonance imaging of the brain before (A–D) and after (E–H) shunt removal. (A) Axial T2 FLAIR image reveals prominent dilated ventricles with transependymal hydrocephalus (white arrows). (B) Coronal fat-saturated T2-weighted image shows evidence of a VP shunt in the right hemisphere and marked lateral ventriculomegaly and ballooning of the third ventricle. There is evidence of oedema involving the optic tracts bilaterally (black arrows). (C–D) Axial T2 FLAIR demonstrates markedly dilated ventricles with hyperintensities (white arrows) evident at the level of the midbrain (unfilled arrowhead), periaqueductal grey (filled arrowhead), optic chiasm and optic nerves. The brainstem is inferiorly displaced. (E) Axial T2 FLAIR demonstrates significant improvement with mild residual oedema (white arrow) in the right posterior parietal region. (F) Coronal T2-weighted image reveals improvement of hydrocephalus. (G–H) Axial T2 FLAIR images demonstrate resolution of transependymal oedema and ventriculomegaly at the level of the optic nerves (G) and optic tracts (H). The transependymal oedema involving the midbrain (unfilled arrowhead) and periaqueductal grey (filled arrowhead) has resolved.

Treatment involves shunt revision or removal, typically resulting in rapid improvement.^{1,3} Dadlani *et al.* described a patient with progressive decrease in VA to light perception bilaterally due to shunt malfunction. Their patient did not exhibit signs of increased intracranial pressure despite ventriculomegaly. His VA improved to CF at 3 feet after VPS replacement.³ In contrast, Sunil *et al.* reported a case of transient bilateral vision loss without disc oedema or fundus abnormalities in a patient and a VA of 20/50 OU. He experienced nausea, gait instability, and changes in memory. CT angiography revealed dilated ventricles resulting in mass effect on the internal cerebral vein. Following valve replacement, the patient experienced improvement of his symptoms without recurrence at one-year followup.¹ Although both cases describe symptomatic improvement after shunt revision, neither case reports evidence of transependymal oedema.

Following our patient's initial shunt revision, he experienced persistent vision loss despite improvement of his systemic symptoms of increased intracranial pressure. Therefore, we postulate the resulting transependymal oedema, rather than hydrocephalus alone, accounted for his acute VA loss. Nickels *et al.* proposes several mechanisms of vision loss due to interstitial oedema. Interstitial oedema can cause direct compression of the optic nerves, thereby resulting in decreased VA. Additionally, the oedema can lead to venous infarction or compression of the vessels that supply the optic nerves, resulting in ischaemia.⁵ Although these theories require further investigation, the direct compression of the optic apparatus due to the interstitial oedema may explain the vision loss seen in our patient. Similarly, the patient's ophthalmoparesis and symptoms of dorsal midbrain syndrome are likely due to an analogous mechanism. The ophthalmoparesis can be explained by the

periaqueductal grey transependymal flow while the symptoms of dorsal midbrain syndrome result from the oedema involving the midbrain structures (Figure 1).

Due to the acute vision loss and MRI findings, there was concern for a demyelinating process prompting anti-MOG antibody studies. Despite positive titres, our patient ultimately did not have evidence of demyelination disease. Although anti-MOG antibodies associated with isolated hydrocephalus has not been reported in humans, Del Bigio describes the overexpression of mRNA for MOG and other proteins in hydrocephalus in animal studies. The disruption due to hydrocephalus may result in attempted compensation to maintain integrity of the brain. Further investigation is required to understand the association of hydrocephalus and MOG levels.⁶ We hypothesise our patient had a false positive anti-MOG antibody titre, since it was negative on repeat testing. However, due to inconclusive data we cannot with certainty determine the aetiology of the initial positive result.

We believe that this patient's reversible vision loss was due to transependymal oedema resulting in direct involvement of the optic apparatus. To our knowledge, this is the first case of fully reversible vision loss due to transependymal oedema involving the visual pathway secondary to VP shunt malfunction in the English language ophthalmic literature. Additionally, transependymal oedema involving the optic chiasm and/or visual pathway

has not been previously reported in this setting. This unusual case may be important to determine the medical management of patients with VP shunt malfunction who present with atypical features.

Declaration of interest

The authors report no conflict of interest.

References

1. Sunil M, Payne C, Panda M. Transient binocular visual loss: a rare presentation of ventriculoperitoneal shunt malfunction. *BMJ Case Rep.* 2011;2011:bcr1020114929. doi:10.1136/bcr.10.2011.4929.
2. Erol FS, Ozturk S, Akgun B, Kaplan M. Ventriculoperitoneal shunt malfunction caused by fractures and disconnections over 10 years of follow-up. *Childs Nerv Syst.* 2017;33(3):475–481. doi:10.1007/s00381-017-3342-0. Epub 2017 Jan 17.
3. Dadlani R, Dadlani R, Ghosal N, Hegde A. Occam's razor in the management of ventriculoperitoneal shunt dysfunction: diagnosis and management of an unusual pediatric case. *Asian J Neurosurg.* 2015;10(2):177–180. doi:10.4103/1793-5482.154988.
4. Mallucci C, Sgouros S. *Cerebrospinal Fluid Disorders*. Boca Raton, FL: Informa Healthcare; 2010.
5. Nickels TJ, Manlapaz MR, Farag E. Perioperative visual loss after spine surgery. *World J Orthop.* 2014;5(2):100–106. doi:10.5312/wjo.v5.i2.100. Published April 18, 2014.
6. Del Bigio MR. Future directions for therapy of childhood hydrocephalus: a view from the laboratory. *Pediatr Neurosurg.* 2001;34(4):172–181. doi:10.1159/000056016.