Visual Disturbance Following Shunt Malfunction in a Patient With Congenital Hydrocephalus

—Case Report—

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Abstract

A 25-year-old woman presented with complaints of nausea and headache. She had been treated with a ventriculoperitoneal shunt for hydrocephalus when she was 7 months old. Her bilateral optic discs showed moderate atrophy. Right visual acuity allowed only perception of hand movement and left visual acuity was 0.02 (1.2). Computed tomography (CT) showed mild ventricular dilation but no periventricular lucency. Intracranial pressure (ICP) was not high when the shunt valve was punctured. Her visual acuity deteriorated 5 days after the consultation. She was referred again 8 days after the first consultation. The bilateral optic discs were completely pale. Both pupils were dilated on admission, and the bilateral direct light reflexes were absent. The patient could slightly detect only green light stimulus. CT showed moderate enlargement of the ventricle. ICP was 47 cmH₂O when the shunt valve was punctured. Shuntgraphy showed obstruction of the shunt at the distal end of peritoneal catheter. Emergent total shunt revision was performed. She could detect dark stimulus and the still-dilated left pupil had recovered direct light reflex on the next day. The visual acuity was 0.01 (0.7) on the left 6 months after the operation, although she was blind in the right eye and the bilateral optic discs were completely pale. Visual loss associated with shunt failure remains a major morbidity in shunted congenital hydrocephalus. Early diagnosis and shunt revision may allow visual recovery.

Key words: congenital hydrocephalus, visual disturbance, ventriculoperitoneal shunt, shunt malfunction, intracranial pressure

Introduction

The neonate skull can enlarge and act as a “release valve” for elevated intracranial pressure (ICP), so elevations in ICP do not cause papilledema in the majority of infants with congenital hydrocephalus.⁴⁾ However, once a patient with hydrocephalus undergoes a shunt procedure, the intracranial sutures can fuse, and subependymal gliosis may develop that can greatly reduce ventricular compliance.⁴⁾ Subsequent shunt failure can produce marked papilledema without ventricular dilation. A 10-year study of a Visually Impaired Program revealed that 1.8% of the children became permanently blind during an episode of increased ICP secondary to shunt malfunction.^{2,3,5,7,10,11)} We treated a patient with congenital hydrocephalus who experienced rapid deterioration of her vision after shunt malfunction.

Case Report

A 25-year-old female was treated with a ventriculoperitoneal shunt for hydrocephalus when she was 7

months old. She also developed glaucoma. ICP was 24 mmHg on the right and 21 mmHg on the left. One of her parents and her brother also had glaucoma. Her visual acuity was 0.01 (0.9) on the right and 0.03 (0.9) on the left when aged 14 years, and the bilateral optic discs showed moderate atrophy. Her visual acuity was 0.05 (0.8) on the right and 0.05 (1.5) on the left when aged 23 years, and the bilateral optic discs were pale. Intraocular pressure was 18 mmHg on the right and 19 mmHg on the left at that time.

She had complained of nausea for 2 weeks. She also had headache and painful sensation in the neck and shoulder for 4 days, so she sought medical treatment. The right eyeball was in the abducted position. Right visual acuity allowed only perception of hand movement and left visual acuity was 0.02 (1.2). Intraocular pressure was 17 mmHg on the right and 25 mmHg on the left. The ocular fundus showed glaucoma-like changes in both optic disks. Computed tomography (CT) showed mild ventricular dilation but no periventricular lucency (Fig. 1). The shunt device felt slightly resistant when compressed through the scalp. However, the ICP was not high when the shunt valve was punctured. Therefore, the patient returned home after removal of 25 cm³ of cerebrospinal fluid (CSF).

Fig. 1 Computed tomography scans at the first consultation showing mild ventricular dilation but no periventricular lucency.

Fig. 3 Computed tomography scans on the day after the operation showing the ventricular size had returned to the same size as seen 9 days earlier.

Fig. 2 A, B: Computed tomography scans at the second consultation showing enlargement of the ventricle in comparison with the finding 8 days earlier. C: Shuntgraphy image showing obstruction of the shunt at the distal end of the peritoneal catheter.

Her visual acuity deteriorated 5 days after the consultation. However, her consciousness was clear and she did not complain of appetite loss. The ophthalmologist again referred her to this department 8 days after the first consultation, because she experienced visual deterioration

that was not caused by the glaucoma. Intraocular pressure was 14 mmHg on the right and 24 mmHg on the left at that time. The bilateral optic discs were completely pale.

Both pupils were dilated on admission, and the direct light reflex was absent bilaterally. The patient could slightly detect only green light stimulus. Her consciousness was clear. The patient reported that the headache and painful sensation in her neck and shoulder had not improved, even after removal of CSF through the shunt device. CT showed enlargement of the ventricle in comparison with the CT scan taken 8 days earlier (Fig. 2A, B). The shunt device felt hard and the ICP was 47 cmH₂O when the shunt valve was punctured. She could detect slight light sensation after removal of 30 ml of CSF. Shuntgraphy showed obstruction of the shunt at the distal end of the peritoneal catheter (Fig. 2C).

Emergent total shunt revision was performed using a Codman® Hakim® Programmable Shunt (Codman & Shurtleff, Inc., Raynham, Massachusetts, USA; 100 mmH₂O resistant pressure). She could detect dark stimulus and the still-dilated left pupil had recovered direct light reflex on the next day. The ventricular size decreased after the operation and returned to the same size as seen on the CT scan taken 9 days earlier (Fig. 3). She was able to detect light stimulus in both eyes by 2 days after the operation. She felt no headache and nausea 3 days after the operation. She could feel the texture of food and dishes, and recognize the presence of a person in front of her. Her visual acuity continued to improve after the operation. Visual acuity was 0.01 (0.7) on the left 6 months after the operation, although she was blind in the right eye and the bilateral optic discs were completely pale. Visual field in the left eye had a range of 95 degrees in the horizontal direction and 90 degrees in the vertical direction. ICP was 17 mmHg on the right and 18 mmHg on the left at that time. The patient could manipulate a cell phone and compose an e-mail using glasses.

Discussion

CT is not necessarily effective for judging the degree of increased ICP. In our case, CT showed only mild enlarge-

ment of ventricular size at the first visit. The mild or intermittent increase in the ICP due to shunt malfunction could not effectively enlarge the entire ventricular system, but was sufficient to cause visual disturbance, headache, and nausea, which indicated increased ICP.^{6,12} The current case showed that elevated ICP due to shunt malfunction cannot be excluded, even if CT shows only slight enlargement in ventricular size.^{6,11} Although the second preoperative CT showed deterioration of the hydrocephalus, the ventricular dilation was moderate.

Fundoscopy examination at the onset showed only glaucoma-like change in our patient. Furthermore, the ICP was not high when the shunt valve was punctured in the outpatient department. However, the intermittent increases in ICP may have had a serious effect on the retinal circulation, during intermittent obstruction of the peritoneal catheter. Fluorescein angiography could have shown a profound delay in retinal and choroidal perfusion at this stage.¹²

Hydrocephalus is associated with a pre/perinatal developmental retinal disorder.¹ One study showed that 14% of patients with congenital hydrocephalus suffer from optic atrophy.¹ Children with hydrocephalus have smaller disc areas, which might reflect reduced number and diameters of the optic nerve axons.¹ Children with hydrocephalus also have abnormal retinal vascular pattern, with straighter arteries and fewer branching points which are thought to reflect the reduced nutritional demands of fewer neural elements.¹ The fragility of the visual system associated with congenital hydrocephalus may result in rapid deterioration of visual ability. When the present patient was 14 years old, the bilateral optic discs had already shown moderate atrophy. The patient became almost blind over only 8 days, during which the shunt failure progressed from intermittent obstruction to permanent obstruction and ICP increased markedly.

Various other mechanisms can cause loss of vision under increased ICP following shunt failure. The lesions are usually classified as pregeniculate (anterior visual pathway) if optic atrophy or decreased light reflex are present, or postgeniculate if these findings are absent.⁷ In the present patient, the bilateral optic discs were completely pale and the pupils were dilated without direct light reflex on admission. Therefore, the lesion was considered to be in the anterior visual pathway. The anterior visual pathway can be disturbed in various portions. So-called "papilledema" is an intraocular lesion caused by increased ICP. The network of small arteries passing over bone, which supplies the intracranial optic nerve, is compressed by generalized high pressure and can cause circulatory stasis and ischemia.^{3,6,7} Enlarged third ventricle can bulge the infundibulum downwards, pressing the posterior angle of the chiasm.^{3,6,7} In the present patient, CT on admission showed enlargement of the ventricle in comparison with the CT scan taken 8 days earlier, but the ventricular dilation was not prominent enough to compress the intracranial small arteries or optic nerves. Therefore, we considered that the lesion was in the intraocular portion rather than in the intracranial portion. The edema of the optic discs possibly did not develop because of the

raised ICP, as the bilateral optic discs had already undergone moderate atrophy.

In some patients with open-angle glaucoma, eyes with normal discs and fields could tolerate a tension of 30 mmHg for many years.⁸ However, if abnormalities ranging from early glaucomatous cupping to advanced visual field defects were present on initial evaluation, progressive loss of field tended to occur at lower tensions.⁸ Similarly, many eyes in patients with acute closed-angle glaucoma suffered visual loss from optic nerve damage despite good control of intraocular pressure.⁹ The intraocular pressure in the present patient was moderately high at the onset, so the glaucoma might have been an aggravating factor of visual impairment. However, the patient did not show any symptoms of acute glaucoma attack such as glaucomatous halo, corneal edema, conjunctival hyperemia, or palpebral edema, except for the nausea and headache. Furthermore, the visual symptom recovered markedly after reconstruction of the shunt system. Therefore, the deteriorated visual symptom was suspected to be caused by the acute hydrocephalus following the shunt failure rather than the glaucoma.

Visual loss associated with shunt failure remains a major morbidity in patients with shunts for congenital hydrocephalus.⁴ Rapid deterioration of vision can occur after shunt malfunction in infants, and serious permanent visual dysfunction follows, if the cause of the visual deterioration is difficult to detect.⁴ Therefore, shunt malfunction should be suspected if patients treated for hydrocephalus present with visual deterioration, headache, and nausea, even if the ventricular dilatation is unremarkable.¹¹ Prompt treatment of increased ICP is important.³ Early diagnosis and shunt revision may allow visual recovery.^{6,7,12}

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