A rare hydrocephalus complication: cortical blindness

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Received: 29 December 2014, Revised: 27 February 2015, Accepted: 5 March 2015


Cortical blindness related to bilateral occipital lobe infarction is an extremely rare complication of hydrocephalus. Compression of the posterior cerebral artery, secondary to tentorial herniation, is the cause of occipital infarction. Particularly in children and mentally ill patients, cortical blindness may be missed. Therefore, early diagnosis and treatment of hydrocephalus is important.

We present herein a child of ventricular shunt malfunction complicated by cortical blindness.

Key words: hydrocephalus, complication, cortical blindness, ventricular shunt malfunction.

Ventricular shunt malfunction (VSM) is a life-threatening condition and must be treated urgently. Acute hydrocephalus may be complicated by convulsion, loss of consciousness, brain herniation and even death. Cortical blindness secondary to occipital lob infarction is a rare complication of hydrocephalus. Early treatment of hydrocephalus can prevent from serious complication. We present a case of bilateral occipital lob infarction complicated by VSM and hydrocephalus.

Case Report
A 3-year-old, prematurely born boy was referred to our emergency department with a history of blindness noticed a few days ago. He had a history of perinatal hypoxic-ischemic encephalopathy, intraventricular hemorrhage, and ventricular shunt placement. The reason for referring to our hospital was visual loss, which had been assumed to happen soon after the shunt revision surgery. Initially, cranial computed tomography (CT) images for symptoms of lethargy and vomiting which were acquired in referring hospital six days ago, showed acute hydrocephalus, hypodensity and edema (Fig. 1). On the next day after the successful shunt revision surgery, patient’s parent noticed visual loss. Two days after the surgery, diffusion-weighted imaging showed bilateral occipital lobes diffusion restriction due to acute ischemia without ventricular dilatation (Fig. 2 A, B). After the following three days, patient was referred to our hospital.

Although it was challenging to accurately examine poorly cooperative patient, repeated neurological and ophthalmological examinations enabled us to make a precise diagnosis. On examination, the patient was unable to fixate and track a target or light. Eye movements were full with normal fundoscopy findings, and pupil-light reaction was within normal limits. According to prior radiological images and the neuro-ophthalmological examination, this situation was interpreted as cortical blindness. Magnetic resonance (MR) imaging and MR angiography of the brain showed late subacute infarction in the bilateral occipital lobes, no signs of hydrocephalus (Fig. 3 A, B) and patency with normal caliber of bilateral posterior cerebral artery (PCA) (Fig. 4). Despite the patency of PCAs, recovery of visual loss was not established. Parents were informed, and the patient was discharged.

Discussion
Ventricular shunt malfunction is a life-threatening condition and must be treated urgently. Acute hydrocephalus may be complicated by convulsion, loss of consciousness, brain herniation and even death. Non-diagnosed
or non-treated hydrocephalus accompanied by raised intracranial pressure (ICP) is a rare cause of cortical blindness. The pathophysiology of this condition depends on elevated ICP, which results in downward tentorial herniation that eventually leads to entrapment of PCAs by the tentorial incisura. By the reason of cortical blindness is due to compression of PCA, not endoluminal pathologies, early treatment and relief of ICP may provide reperfusion of occipital lobes and total recovery of vision. Cases of tentorial herniation complicated by occipital lobes infarction have been reported in the literature since 1980s. However, in these cases, etiology of raised ICP is generally results from supratentorial, intra/extra-axial hematomas or masses. Also, other cases with tentorial herniation, which are due to colloid cysts of the third ventricle, can be found in the literature. Siu et al. reported a case of third ventricle colloid cyst complicated by spinal cord infarction due to anterior spinal artery occlusion which is a very rare condition. Rich vascular supply lets these areas be less vulnerable than occipital lobe.

Cortical blindness is named if there is a loss of visual information to the occipital cortex due to a lesion of the visual pathways. This condition is usually caused by lesions in the visual cortex, but it can also be caused by lesions in the optic nerve, optic tract, or retrochiasmatic areas. It is characterized by the sudden onset of blindness in one or both eyes, and it is usually permanent. Cortical blindness is a common condition in patients with hydrocephalus, and it can be caused by various factors, including raised ICP, compression of the PCA, and infarction of the occipital lobes. Early diagnosis and treatment are necessary to prevent irreversible damage to the visual cortex.
Fig. 3. Brain MR imaging which was performed on the tenth day after the shunt revision surgery. DWI (A) and ADC map (B) show resolving restriction of diffusion previously seen in the bilateral occipital areas, suggesting subacute infarct. There is no evidence for hydrocephalus.

Fig. 4. Reconstructed 3D TOF MR Angiography shows patency of lumen of bilateral PCAs and other vascular structures with normal diameter.

Cortical blindness due to hydrocephalus

in visual acuity or visual fields in structurally normal eyes and normal appearing anterior visual pathways due to an abnormality affecting the part of the brain responsible for sight. Cortical blindness is an older term for cortical visual impairment (CVI). The term “blindness” can be misleading. Children with CVI usually have some level of vision which can improve over time. CVI is the most common cause of permanent visual impairment in children and can be caused by any process that damages the brain. Examples include: stroke, decreased blood supply, decreased oxygenation, brain malformation or infection, hydrocephalus. Congenital cortical blindness is most often caused by perinatal ischemic stroke, encephalitis, and meningitis.

In the present case, the reason for ischemic cortical blindness was entrapment of PCAs by dilated third ventricle due to VSM. Particularly in children and mentally ill patients, cortical blindness may be missed by clinicians as happened in our case. The cortical blindness was noticed by patient’s mother after the shunt revision operation in our case, so it was questioned whether or not blindness had been a complication of surgery. In fact, it was
presenting symptom which existed on initial CT scan.

REFERENCES
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