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Case report

Accelerated visual recovery from protracted hypoxic cortical blindness in a child

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ABSTRACT

Purpose: This report describes accelerated visual recovery in a child following protracted hypoxic cortical visual loss and reviews mechanisms responsible for visual recovery.

Observations: A 12-year-old boy developed cortical blindness after a severe snowboarding crash. Magnetic resonance imaging showed severe multifocal hypoxic brain injury, with multifocal restricted diffusion and extensive T2/FLAIR hyperintensities throughout the visual cortex, basal ganglia and midbrain. The mismatch of affected areas on FLAIR and DWI sequences indicated a combination of cytotoxic and vasogenic edema, which suggested partial reversibility with potential for recovery. Two weeks after his injury, he began to experience an accelerated improvement in vision with recovery of 20/20 visual acuity and 40 sec/arc stereoacuity over the following week. Three months later, visual field examination showed a steep-margined horizontal band of spared visual field, which showed further expansion on repeat testing 1 year later.

Conclusions and importance: Protracted hypoxic cortical visual loss can be followed by dramatic visual recovery in children. Magnetic resonance imaging can provide useful prognostic information.

1. Introduction

Cortical blindness can develop following hypoxic injury to the brain.[1–6] Visual recovery to varying degrees has been reported previously, although the mechanisms responsible are unclear.[1–6] We document the unusual clinical course and neuroimaging findings in a child who displayed accelerated visual recovery following hypoxic brain injury resulting from a snowboarding crash.

2. Case report

A 12-year-old boy sustained a severe concussion to the head and injuries to the face and chest from a snowboarding crash on a school trip. He required cardio-pulmonary resuscitation and had needle thoracotomy for pneumothorax on the scene. He was airlifted to the hospital, where he was found to be comatose with bilateral pneumothoraces and pulmonary contusions. A computed tomography (CT) scan of the head showed no intracranial hemorrhage or fractures. Over the course of the first week, he received intravenous midazolam and fentanyl, piperacillin and tazobactam for suspected aspiration pneumonia with acute respiratory distress syndrome, and norepinephrine as vasopressor therapy. On the 7th day after the injury, he began recovering consciousness and was extubated. He showed no clinical signs of seizure activity before or after regaining consciousness. As his cognition recovered, it became evident that he could not see people and objects around him.

On day 10 after the injury, his vision acuity was light perception without projection with absent optokinetic nystagmus, although he seemed unaware of his blindness. The pupils were 7 mm in diameter with brisk reactions to light and no relative afferent pupillary defect. The retinas appeared normal, and there were no signs of optic disc swelling or atrophy. Magnetic resonance (MR) imaging of the brain demonstrated signs of multifocal hypoxic-ischemic injury. Diffusion weighted imaging (DWI) showed several areas of restricted diffusion throughout the cortex and subcortical white matter, most prominent in external vascular border zones between the anterior/middle and middle/posterior cerebral artery territories. On T2-weighted and FLAIR (fluid attenuated inversion recovery) sequences, more extensive areas of hyperintensity were present with symmetric involvement of the deep gray structures, including basal ganglia and, to a lesser extent, the thalami and midbrain. Diffusely abnormal signal was noted throughout the cerebral cortex, including visual areas such as the occipital cortex

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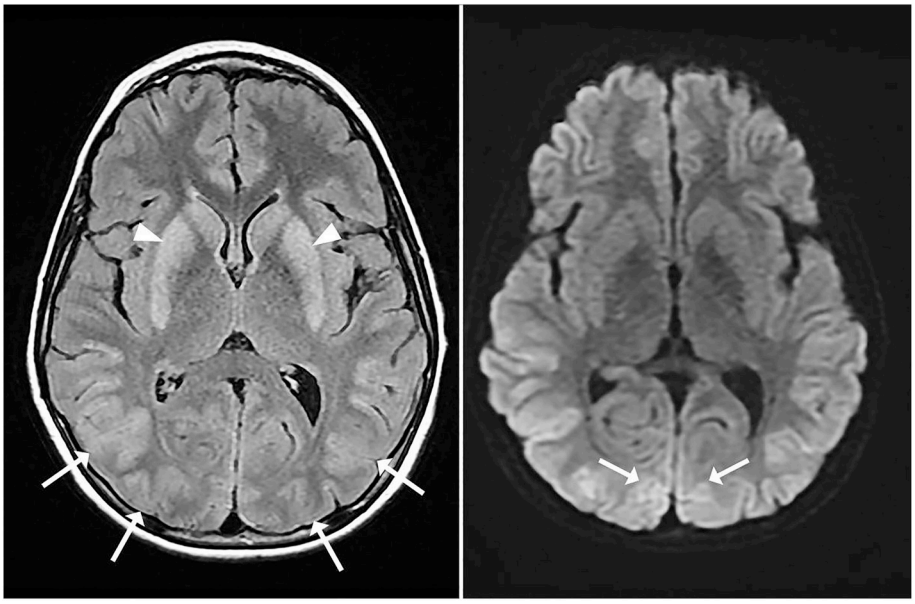

Fig. 1. Magnetic resonance imaging. Fluid attenuated inversion recovery (FLAIR) sequence (left panel) demonstrates hyperintense signal throughout the basal ganglia (arrowheads) and cortex, particularly posteriorly (arrows). Diffusion-weighted imaging (DWI) (right panel) shows restricted diffusion with hyperintense signal in multiple cortical areas, including the temporal-occipital regions and calcarine fissures (arrows).

and calcarine fissures (Fig. 1). T2-weighted FLAIR imaging showed mild signal hyperintensities within the posterior optic radiations; however, the deep white matter tracts appeared relatively preserved on diffusion tensor imaging.

Two weeks after the injury, the patient's vision remained at light perception. He then experienced “an explosion of sight,” and vision improved to 20/20 in each eye over the next week. Three months later, visual acuity was 20/15 with full color vision, normal stereoacuity, and no optic atrophy. Three months after the injury, Humphrey 10–2 visual field testing showed a bilaterally symmetrical superior and inferior altitudinal defect with a 5-degree horizontal band of intact visual field (Fig. 2-top). Repeat testing at one year showed superior and inferior expansion (Fig. 2-middle). Humphrey 30–2 testing showed a steep-margined peripheral visual field defect with persistent sparing of an inferior horizontal band and expansion of the superior visual field (Fig. 2-bottom). His neurologic function is otherwise normal, and he is getting good grades in school and has resumed skiing (but not snowboarding).

3. Discussion

Recovery from cortical blindness can occur in the setting of hypoxia following cardiac arrest.[1–3] Hoyt et al. described a case of partial recovery of vision in a 39-year-old following cardiac arrest, and reported visual field improvement over 2 months.[1] The authors hypothesized that the striate cortex was selectively affected, due to the increased number of granular cells especially in layer 4 of the occipital cortex. Margolin et al. described cortical visual loss with hand motion vision in a 16-year-old, after asystole occurring during a motor vehicular accident. His vision recovered to 20/20 over 12 weeks, although he demonstrated constricted visual fields.[2] In some cases the onset of blindness began several days after the initial injury.[4–6] De Souza et al. reported a case of delayed onset cortical blindness in a 47-year-old one week after cardiac arrest following a myocardial infarction, with recovery to normal visual acuity 1 month later.[6]

The potential for visual recovery in our patient was suggested by the presence of both vasogenic and cytotoxic edema, as evidenced by the mismatch between DWI and T2/FLAIR-weighted images. The pathophysiology of brain injury in cardiac arrest has been described as a “two-hit model”.[7] The initial hypoxia caused by the cardiac arrest results in neuronal death, i.e. the first hit. Primary energy failure results in cell death with cessation of ion exchange and trapping of free water within cells, producing restricted diffusion on DWI sequences (cytotoxic edema). Following cardiopulmonary resuscitation, additional injury-i.e. the second hit—can result from cerebral autoregulatory disturbances and reperfusion insult. As a part of this cascade, there is blood-brain barrier breakdown with a fluid shift from the intravascular compartment to the interstitial space, manifesting as areas of matched hyperintensity on T2/FLAIR and DWI (vasogenic edema). Loss of function in areas of vasogenic edema is potentially reversible if the underlying cause is corrected, as seen with cerebrovascular autoregulatory disorders such as posterior reversible encephalopathy syndrome.[8] The mechanism behind the later visual field recovery could be explained either by retained neuroplasticity because of his young age, by better test-taking ability from repetition of the visual field, or improved cognition.

The congruent, steep-margined visual field defects in our patient are indicative of bilateral occipital injury, but inconsistent with watershed injury isolated to the striate cortex.[9] Rather, the visual field defects and multifocal lesions on MR imaging indicate a watershed event that more broadly involves the striate cortex, extrastriate cortex, and optic radiations to varying degrees. His dilated pupils may have been caused by hypoxic disruption of autonomic efferent pathways, as suggested by the dorsal midbrain signal abnormalities on MR imaging.

4. Conclusions

This case history demonstrates the remarkable potential for recovery of cortical vision in children with hypoxic brain injury. For reasons that are not fully understood, visual recovery can continue long after the injury. MR imaging can provide prognostic information by identifying affected areas and distinguishing potentially reversible vasogenic edema from irreversible cytotoxic edema.

Patient consent

The patient's legal guardian consented to publication of the case. This report does not contain any personal information that could lead to the identification of the patient.

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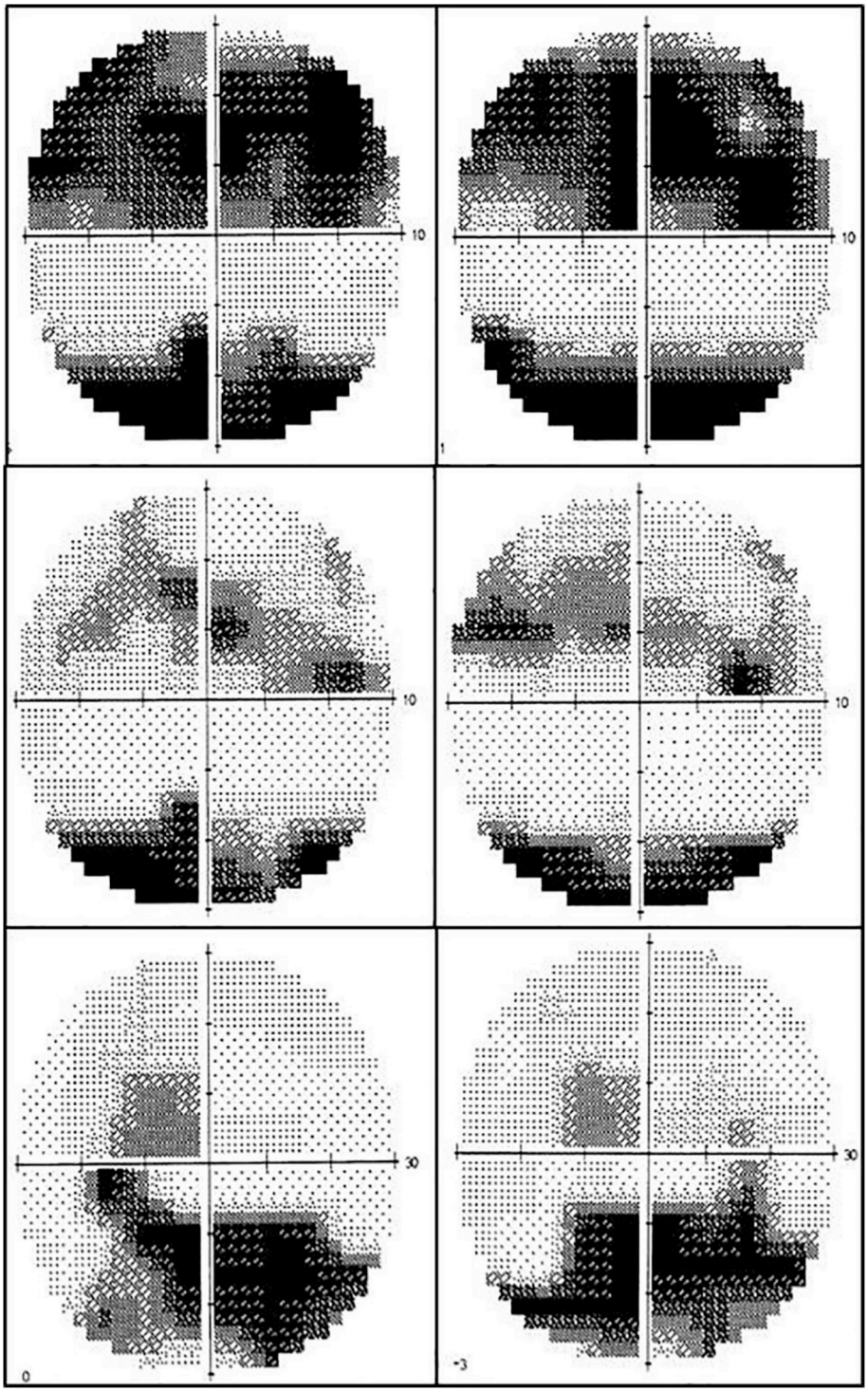

Fig. 2. Humphrey visual fields (HVF). 10-2 HVF right eye (right top panel) and left eye (left top panel) performed at 3 months following injury showing a bilaterally symmetrical superior and inferior altitudinal defect with a 5-degree horizontal band of intact visual field. 10-2 HVF right eye (right middle panel) and left eye (left middle panel) performed at 1 year following injury showing expansion of the field. 30-2 HVF right eye (right bottom panel) and left eye (left panel) performed at 10 months following the injury showed a steep-margined peripheral visual field defect with persistent sparing of an inferior horizontal band and expansion of the superior visual field.

Conflicts of interest

The following authors have no financial disclosures: Sasha Mansukhani, Mai Lan Ho, Elizabeth Bradley, Michael C. Brodsky.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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