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CASE REPORT

Failure of the Clinical Examination to Predict the Presence of Functional Vision

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ABSTRACT

The absence of a pupillary light reflex and the lack of any clinical response to visual stimulation are generally considered reliable signs of poor visual function in patients with abnormal mental status. Two case reports are presented, one with no pupillary light reflex and no response to visual stimuli; and another with no clinical response to visual stimulation. After several months with no signs of vision, both patients regained significant visual function. It is important to recognize that all signs of visual function can be absent despite the potential for good vision. This should be considered before concluding that visual loss will be permanent.

KEYWORDS optic nerve, pupillary light reflex, trauma, vision

INTRODUCTION

Optic nerve injury is a common accompaniment to head trauma and is often missed early on, as the patient is unable to communicate a subjective change in visual function. A careful clinical examination, however, has generally been considered a reliable way to detect total unilateral or bilateral loss of vision even in the absence of an ability to communicate. We report below two patients who presented with no clinical evidence of visual function of one or both eyes. Later evaluation found that both had good residual vision.

CASE REPORTS

Case 1

A 26-year-old previously healthy woman was involved in a skiing accident, sustaining multiple skull fractures, subdural and subarachnoid haemorrhages, and bifrontal intraparenchymal haemorrhagic contusions. She recovered from her acute injuries and after 1 month in an inpatient rehabilitation unit, a neuro-ophthalmology consultation was requested concerning poor vision in one eye. She was awake but neither spoke nor followed any commands. She would blink in response to sudden noises. A neuro-ophthalmological examination at that time found the patient to have a normal blink to bright light in the left eye but a response neither to light nor to threat on the right. Both pupils were round and regular. In the dark the right pupil measured 7 mm in diameter whereas the left was 4.5 mm. In the light the right measured 6.0 mm and the left was 3.0 mm. There was a large relative afferent pupillary defect (APD) in the right eye. Funduscopic examination revealed a diffuse loss of nerve fibres in the right eye with markedly narrowed vasculature. The left fundus was normal. The right eye did not have any vertical movements. Adduction was limited to 50–60% but there was full abduction. Extra-ocular movements of the left eye were normal. Over the next several weeks she was evaluated frequently for any evidence of vision in the right eye. No response of either pupil was ever noted to an indirect ophthalmoscope light directed at the right eye while the pupil was observed under a magnifying lens. No eye movements were evoked in either eye using an optokinetic drum, a large moving mirror, or any other attempt at visual stimulation in front of the left eye. Visually evoked potential (VEP) testing was initially done using flash stimuli, as the patient was not able to fixate consistently on a target. There was a normal response of the left but none was seen on the right.

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She was evaluated at least every other week over the next 3 months. There was a continuing improvement in mental status and in III nerve function. She began to follow some commands and to fix and follow with the left eye but not the right. Visually evoked responses were repeated using both flash and pattern stimuli. Again, responses were normal on the left but no repeatable response of the right eye was present to either stimulus. She began identifying letters and colours with the left eye but reported no vision with the right. The third nerve paresis improved and the consensual pupillary responses of the right eye to a left sided stimulus became more brisk. Nonetheless there continued to be a total lack of a direct or consensual response after stimulation of the right eye using the light of an indirect ophthalmoscope.

After 3 months a therapist reported that the patient seemed to be able to identify colours with the right eye. The initial clinical impression was that she must have been using the left eye. Total careful patching of the left eye, however, proved that she was clearly seeing from the right. In fact her visual acuity was 20/200. By confrontation, finger counting in the right eye was present in the inferior visual field, but absent in the superior visual field. On funduscopic examination, the right optic disc was markedly atrophic with perivascular sheathing. The left disc was within normal limits. The pupils were 6 mm in diameter. There remained an absolute right APD on repeated examinations with no detectable direct or consensual pupillary light reflex to stimulation of the right eye. Both pupils reacted briskly with stimulation of the left eye. At that time her external ocular movements were full in both eyes.

When re-examined a year later, the pupillary findings were unchanged but her visual acuity had improved to 20/70. Although none was seen grossly, slit lamp examination revealed a minimal reaction of the right pupil to direct stimulation. Visual fields by confrontation were normal on the left, but showed a dense loss to hand movement superiorly on the right.

Case 2

A 20-year-old previously healthy woman was involved in a horseback riding accident. Computerised tomographic and magnetic resonance imaging studies revealed multiple small intraparenchymal haemorrhages with intraventricular extension into the occipital horns, and the presence of subarachnoid haemorrhage. There was a right frontal white matter hyperdensity at the cortical-subcortical junction consistent with diffuse axonal injury, as well as a right caudate subacute lacunar infarction.

The patient was first seen 3 weeks after trauma for evaluation of cortical blindness. She was confused and unable to communicate verbally but awake. Her pupillary examination was normal, but despite normal pupillary responses to light, there was a blink response neither to visual threat nor to stimulation with the light of an indirect ophthalmoscope. Neither fast- nor slow-phase eye movements were elicited with optokinetic stimulation, and there was no discernible response to movements of a large mirror or any other visual stimulation. She did, however, have normal spontaneous saccadic eye movements in all directions.

Over the next several weeks these manoeuvers were repeated a number of times and no responses were ever noted. She would stare blindly and make grasping movements and saccades as if to imagined objects. At 8 weeks after the trauma her mental status started to improve but repeated evaluations weekly over the next month showed no change. One day after one such examination we were called regarding a marked improvement in her mental status, and she was documented to have 20/20 vision and full fields to confrontation bilaterally. Eye movements were also normal. Formal visual fields were not done.

DISCUSSION

These cases describe two patients who recovered significantly despite no evidence of any visual function. In the first case, there appeared to be a total unilateral visual loss, absent direct and consensual pupillary light reflex, and absent flash and pattern reversal VEPs. This case is striking for three major reasons. Firstly, recovery after having no perception of light (NPL) for 3 months is rare. Recovery after prolonged blindness from compression by a mass lesion has been reported at least twice. In one patient, treatment resulted in recovery after 1 week of amaurosis.[1] In a more striking case, vision recovered 1 year after the patient became NPL from compression by a craniopharyngioma. Although recovery of vision after documented NPL post trauma has been reported, we were unable to find recovery from an optic neuropathy this long after trauma.[2]

Secondly, the clinical examination showing no response to a bright light, no VEP response and no following response to a peripheral full field stimulus led to a false impression of a negligible chance of recovery. This case stands in contrast to the report by Agarwal and Mahapatra who evaluated 100 patients with NPL vision post trauma. Of 15 patients with absent VEPs, none recovered vision.[3]

Thirdly, although a pupillary light reflex can be seen in the absence of clinical evidence of perception of light, we are unaware of a report noting this level of visual function in a patient with no pupillary light reflex. Routine clinical examination carried out at the bedside with a penlight may be unable to detect a pupillary light response when the amplitude is less than 0.3 mm and the maximal constriction velocity is <1 mm/s.[4] This patient, however, was evaluated with a very bright light under a magnifying glass on a number of occasions.

The second case is one with presumed cortical blindness associated with no visible response to threat; to a direct very bright light; or to a large moving full-field visual stimulus. It was striking not only because of the total lack of evidence of visual function by all clinical parameters followed by excellent recovery, but also because of the rapidity with which the patient recovered.

The anatomy underlying the reflex blink to threat is unclear. Liu and Ronthal[5] refer to case reports in earlier studies by Keane[6] and Tavy et al.[7] that suggested this reflex required an intact optic nerve and its connection to the pretectum but not the cortex. However, Liu and Ronthal in their review of five patients with abnormal blink-to-threat reflexes concluded that the blink-to-visual threat reflex requires an intact striate cortex as well as other mechanisms located in the inferior parietal lobules and frontal eye fields. Overall our case would support the hypothesis that cortical regions are involved, as our patient had no evidence of brainstem damage. It is tempting to suggest that the case supports frontal lobe involvement in the blink-to-threat reflex, as she had a clear frontal lesion consistent with diffuse axonal injury and no evidence of occiptal damage. However, the pattern of recovery suggests the possibility of a cortical equivalent of spinal shock involving the occipital lobe.

Overall these cases suggest that even full, careful repetitive examinations, including electrophysiological studies, demonstrating no evidence of response and any visual function cannot rule out the potential for recovery of visual function. Vision is ultimately a subjective process that is difficult if not impossible to adequately evaluate when a patient has no ability to give the examiner adequate feedback. Much care needs to be taken in speaking to the patient and to the family of such patients. Particularly in someone with a decreased mental status, it can be very difficult to be absolutely sure that no visual recovery is possible and therefore we should be extremely cautious about conveying such a bleak prognosis to the patient or the family.

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