Case Report

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Transient cortical blindness secondary to hepatic encephalopathy in a pediatric patient: A case report and literature review

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Abstract:

Cortical blindness, also known as cerebral visual impairment, may occur in pediatric patients. Hepatic encephalopathy is a rare cause of cortical blindness in children. This report describes a girl with underlying type 1 autoimmune hepatitis, who complained of sudden-onset, painless visual loss in both eyes, which was associated with generalized headache and altered mental status. She was treated with intravenous antibiotics and syrup lactulose. The patient regained full visual recovery after 1 week. Prompt diagnosis and treatment are mandatory in such uncommon instances.

Keywords:

Cortical blindness, hepatic encephalopathy, pediatric patient, visual recovery

Introduction

Cortical blindness or, as it recently called, cerebral visual impairment is a common cause of permanent visual impairment/loss in children. Common causes that have been reported in pediatric patients include hypoxic–ischemic encephalopathy, periventricular leukomalacia, traumatic brain injury, meningitis, cardiac arrest, hypoxia, status epilepticus, intracranial hemorrhage, and cerebral thrombosis.[1]

Hepatic encephalopathy is a rare cause of cortical blindness in the pediatric age group. This report describes the case of a young girl who developed hepatic encephalopathy and presented with sudden-onset, painless visual loss, headache, and altered mental state. Prompt diagnosis and management are very crucial for this uncommon clinical condition.

Case Report

A 13-year-old girl complained of sudden-onset, painless visual loss in both eyes, with generalized headache. Two hours later, she spoke incoherently, was delirious, and was not obeying commands. There was no history of recent trauma or fever.

The patient was diagnosed with type 1 autoimmune hepatitis 4 years prior. She first presented with generalized yellowish discoloration of the eyes and skin, abdominal pain, and lethargy. She was initially treated for leptospirosis and recovered. However, she had persistently elevated liver enzymes. A liver biopsy revealed autoimmune changes, which confirmed her diagnosis.

Two years later, she developed liver failure and portal hypertension. She was placed on the list for a liver transplant procedure. She requires oral azathioprine 50 mg once daily, oral prednisolone 20 mg once daily, syrup lactulose 15 ml once at night, and

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oral propranolol 10 mg twice daily. She had ocular assessments every 6 months, which have revealed visual acuity of 6/6 in both eyes (20/20 OU).

Five days before her current presentation, she visited the emergency department with vomiting and hematemesis. She had reduced oral intake and appeared lethargic. Her temperature was 37.8°C. Her blood pressure was 85/54 mmHg, which subsequently improved with an intravenous infusion of dopamine 280 mg in 50 ml of dextrose 5%. She was admitted for stabilization and further treatment.

She had persistent coffee ground vomitus in the ward, and a diagnosis of hypovolemic shock secondary to bleeding esophageal varices was considered. She was started on intravenous octreotide infusion at 40 mcg/h and was transfused with 1000 ml of fresh frozen plasma.

The patient was drowsy, lethargic, and appeared pallor. Her Glasgow Coma Scale measurement was 13/15 (4 for eye response, 4 for verbal response, and 5 for motor response). Her visual acuity was hand motion in both eyes, and her primary gaze was orthophoric. The pupillary reflexes were normal in both eyes, with no evidence of relative afferent pupillary defect. The optic nerve functions could not be assessed because of profound visual loss. The anterior segment examination of both eyes was normal, and the funduscopy examination showed healthy disc, macula, and retina in both eyes [Figure 1].

Laboratory investigations revealed an elevated serum ammonia level (190 mmol/l) and altered liver enzymes. These included aspartate transaminase (111 IU/l), alkaline phosphatase (292 IU/l), and alanine aminotransferase (85 IU/l). The total serum bilirubin was 560 μmol/l. The total white cell count was 5.48 × 10^9/l, hemoglobin level was 10.8 g/dl, and the platelet count was 30.0 × 10^9/l. Magnetic resonance imaging of the brain and orbit showed normal findings [Figure 2]. The visual evoked potential showed delayed P100 latencies in both eyes [Figure 3]. The clinical presentation was, therefore, consistent with acute cortical blindness.

The systemic manifestations suggested a diagnosis of grade II hepatic encephalopathy. The patient began receiving intravenous cefotaxime 1 g three times daily for 10 days and intravenous metronidazole 500 mg three times daily for 7 days. Syrup lactulose 15 ml twice daily was also given. An emergent esophagogastroduodenoscopy was subsequently deferred because she had stopped vomiting.

Her visual acuity gradually improved, alongside the improvement of her general condition over the next 4 days. She regained full consciousness and orientation, with a full Glasgow Coma Scale (4 for eye, 5 for verbal, and 6 for motor responses). The best-corrected visual acuity was 6/6 in both eyes (20/20 OU) after 1 week of treatment. Ocular examination was essentially unremarkable. The serum ammonia level reduced to 154 mmol/l. She was discharged home well.

Discussion

Hepatic encephalopathy is an uncommon cause of cortical blindness. Most published case reports were

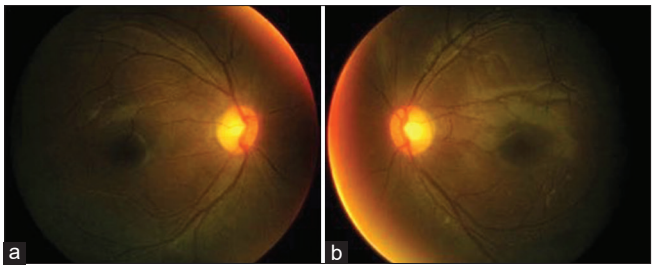

Figure 1: Color fundus photographs (a and b) with normal findings in both eyes

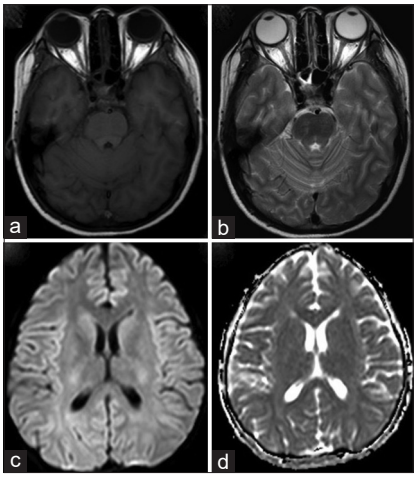

Figure 2: Axial images of magnetic resonance (a, T1; b, T2; c, diffusion-weighted imaging; d, apparent diffusion coefficient). All show normal findings

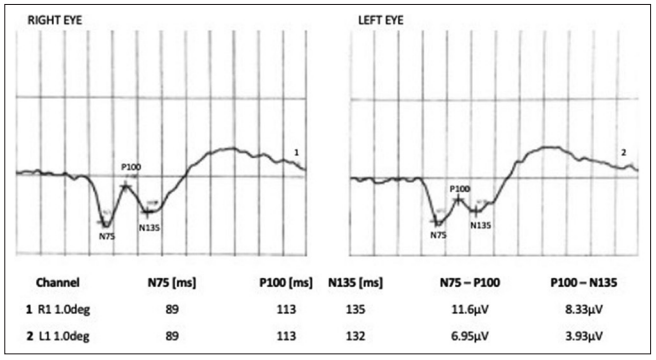

Figure 3: Recorded visual evoked potential, showing delayed P100 latencies

on hepatic encephalopathy causing cortical blindness in adult patients.[2-9] Table 1 summarizes the published reports on cortical blindness resulting from hepatic encephalopathy in patients aged <20 and including the present patient.

Acute liver failure or chronic liver disease can cause hepatic encephalopathy, which results from hepatocyte failure affecting brain function. The present patient developed hepatic encephalopathy secondary to liver failure that has resulted from chronic autoimmune hepatitis type I. Arikan *et al*. reported that an acute fulminant hepatitis of unknown origin as the cause of hepatic encephalopathy in their patient.[10] Ammar *et al*. described a 19-year-old male who developed hepatic encephalopathy secondary to acute paracetamol/ecstasy overdosage.[3] Chronic infective hepatitis B and C[2,6-9] and unknown etiologies[4,5] have been reported in adult patients who have presented with cortical blindness resulting from hepatic encephalopathy.

Visual loss in hepatic encephalopathy is believed to be caused by an impaired blood–brain barrier and accumulation of toxic neurotransmitters in the visual cortex. Second, visual loss can also be due to infarction of parietal and occipital lobes resulted from profound hypotension and anemia.[3] The present patient had a good visual recovery after 1 week. The patient described by Arikan *et al*. improved after 3 weeks.[10] The duration of visual loss reported in adult patients has ranged from 12 h to 3 weeks.[2,4-9] Good visual recovery has also been reported by Cheng-Tagome *et al*., Miyata *et al*., Naparstek *et al*., Eguchi *et al*., and Chen and Chen.[2,4-6,9] This suggests that the condition is reversible with prompt treatment. In contrast, Ammar *et al*. reported that their patient did not regain visual improvement, even after a year.[3] This could be due to irreversible, neurotoxic effect of paracetamol and ecstasy, which resulted in permanent damage to the visual cortex.[3]

Ammonia is the by-product of the bacterial metabolism of protein and nitrogenous compounds in the colon and of glutamine metabolism in enterocytes.[11] It is an important factor in producing encephalopathy. Serum ammonia level was elevated (190 mmol/L) in the present patient. Arikan *et al*. report that their patient had a serum ammonia level of 296 mg/dL (normal: <80 mg/dL).[10] Ammar *et al*. do not mention the level of serum ammonia in their patient,[3] but elevated serum ammonia level was also documented in adult patients.[2,4-6,9] This is parallel with Qureshi *et al*., who state that ammonia level correlates with the severity of hepatic encephalopathy.[12] On the other hand, Van Pesch *et al*. note a normal level of serum ammonia in their patient with hepatic encephalopathy and cortical blindness.[8] The current authors are unable to find any published

Table 1: Summary of published case reports of cortical blindness due to hepatic encephalopathy in patients aged <20 years old

Authors/year	Age/gender	Cause	HE grade	Associated symptoms	Serum ammonia (normal value)	Presenting visual acuity	Duration of visual recovery	Final visual acuity	Brain MRI findings	Treatment
Arikan *et al*./2003	5/male	Fulminant hepatitis of unknown cause	III-IV	Not available	296 mg/dl (normal: <80)	Hand motion	3 weeks	6/6 in both eyes	Occipitoparietal lesion on both sides	Syrup lactulose, IV amikacin, liver transplantation
Ammar *et al*./2003	19/male	Fulminant hepatitis due to paracetamol/ecstasy overdosage	Not available	Gastrointestinal bleeding	Not available	No perception of light	No improvement at 8 weeks and 1 year later	No recovery (no perception of light)	Normal	Liver transplantation; no details of medical treatment
Our patient/2020	13/female	Autoimmune hepatitis type 1	II	Gastrointestinal bleeding and hypovolemic shock	190 mmol/l (normal: 18-72)	Hand motion	1 week	6/6 in both eyes	Normal	Syrup lactulose, IV cefotaxime, IV metronidazole

HE=Hepatic encephalopathy, MRI=Magnetic resonance imaging, IV=Intravenous

reports concerning the association/correlation between serum ammonia level and visual recovery in patients with hepatic encephalopathy.

The brain imaging was normal in the present patient. This finding is also similar to the report by Ammar *et al*.[3] Bilateral parietooccipital white matter signal abnormalities with gliosis were documented by Arikan *et al*.[10] Van Pesch *et al*. also report a recurrent occipitoparietal lesion in their patient, a 55-years old male, who also showed evidence of focal occipital status epilepticus on electroencephalogram.[8] Brain imaging is helpful for excluding space-occupying lesions or cerebral vascular accidents, which may be mistaken for cortical blindness.

Treatment strategies consider the triggering event, the presence/absence of associated symptoms, the severity of hepatic encephalopathy, the severity of underlying liver disease, and the age and general condition of the patient.[13-14] Medical treatment includes intravenous antibiotics/protein intake, correction of fluid/electrolyte imbalance, and drugs to reduce the production of ammonia in the colon. Correction of anemia and coagulopathy is also essential. The current patient presented with gastrointestinal bleeding associated with hepatic encephalopathy. A similar presentation is described by Ammar *et al*. and Arikan *et al*.[3,10] The other published cases in adults report the absence of associated symptoms.[2,4-9]

Liver transplantation aims to enhance the survival of patients with severe hepatic encephalopathy. It has been described as the treatment received by two young patients who had hepatic encephalopathy due to fulminant hepatitis.[3,10] However, these patients developed cortical blindness following liver transplantation.[3,10] All the published cases including the present patient survived the encephalopathy attack,[2-10] although Miyata *et al*. report that their patient had six attacks of hepatic encephalopathy in 1 year.[4]

Conclusion

Cortical blindness secondary to hepatic encephalopathy in the pediatric patient is rare. Early diagnosis and prompt medical treatment are essential for a complete visual recovery. The correlation of serum ammonia level with the rate of visual recovery is another issue that should be explored and discussed.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the parents has given the consent for their child's images and other clinical information to be reported in the journal. The parents understand that their child's name and initial will not be published and due efforts will be made to conceal the identity, but anonymity cannot be guaranteed.

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Conflicts of interest

The author declares that there are no conflicts of interests of this paper.

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