Case Report

Hepatic Encephalopathy Induced Transient Cortical Blindness

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ABSTRACT

Hepatic encephalopathy describes a spectrum of potentially reversible neuropsychiatric abnormalities seen in a patient with severe liver dysfunction with porto-systemic shunting. Cortical blindness can be a rare presentation of hepatic encephalopathy and can even precede the onset of altered sensorium. We report a case of 57 years female with chronic liver disease who presented with bilateral loss of vision, with no focal neurological deficits. From clinical and laboratory examination, a diagnosis of hepatic encephalopathy with cortical blindness was proposed. Her visual disturbances gradually improved with the treatment of hepatic encephalopathy.

Keywords: Cortical blindness; end stage liver disease; hepatic encephalopathy; papilledema.

INTRODUCTION

Hepatic encephalopathy (HE) affects several neurological domains including consciousness, personality, emotional status, motor function, memory, and cognition.[1] It's pathogenesis is believed to be multifactorial and includes impaired blood-brain barrier (BBB) function, neurotoxic substance accumulation, alterations in the synthesis and catabolism of neurotransmitters, and a lack of nutrients such as glucose.[2] Cortical blindness (CB) is a rare presentation of HE and only a few such cases have been reported. Here, we describe a sudden acute bilateral loss of vision in a patient suffering from HE who also had a finding of mild papilledema.

CASE REPORT

A 57 years female, who was a known case of alcohol induced chronic liver disease with a history of upper gastrointestinal (UGI) endoscopy with variceal ligation, presented with painless bilateral visual impairment for two days. She also complained of a sensation of weakness of her lower limbs and headache over bilateral parietal regions. Her history revealed that she had not passed stool for the last two days.

At the presentation to our emergency unit, her oxygen saturation was 95% in ambient air and blood pressure of 150/90 mmHg. Heart rate was 100 beats per minute with a respiratory rate of 18 breaths per minute, a temperature of 97.4 degrees F, and random blood sugar of 222mg/dl. Her consciousness level on the Glasgow Coma Scale was 14 (E4V4M6). Neurological examination showed no signs of a focal neurological deficit with bilateral downgoing plantar reflexes. On ophthalmic examination, pupils were round and isocoric with normal light reflexes. Visual examination showed that she had no perception of light. On fundoscopic examination, there were signs of early papilloedema with no diabetic or hypertensive retinopathic changes. Her initial lab investigations are as shown in (Table 1).

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Table 1. Patient's lab reports on the day of admission.

Hematology		LFT		RFT	
TC	3,400/cumm	T. Bil	6.46 mg/dl	Urea	11.9 mg/dl
Neutrophil	81 %	D. Bil	4.7 mg/dl	Creatinine	0.4 mg/dl
Lymphocyte	13 %	SGOT	68 U/L	Na	143 mEq/l
Eosinophil	0 %	SGPT	26 U/L	K	3.3 mEq/l
Monocytes	6 %	ALP	103 U/L	Chloride	109 mEq/l
Basophil	0 %	Protein	7.6 gm/dl		
Hemoglobin	12.2 gm%	Albumin	2.8 gm/dl		
PCV	36.4 %				
MCV	79.1 fl	**Coagulation**		**TFT**	
MCH	26.6 pg	PT	18.7 sec	T3	6.24 p mol/L
MCHC	33.6 %	INR	1.5	T4	20.7 p mol/L
Platelets	76,300/cumm			TSH	0.877 µIU/ml

Plain computerized tomography scan (CT scan) head was done which was essentially normal. Fluid-attenuated inversion recovery (FLAIR) magnetic resonance imaging (MRI) showed high signal intensity changes in bilateral globes pallidus, cerebral peduncles, posterior aspect of pons, and midbrain with involvement of periaqueductal grey matter, suggestive of hepatocerebral degeneration secondary to hepatic encephalopathy (Figure 1).

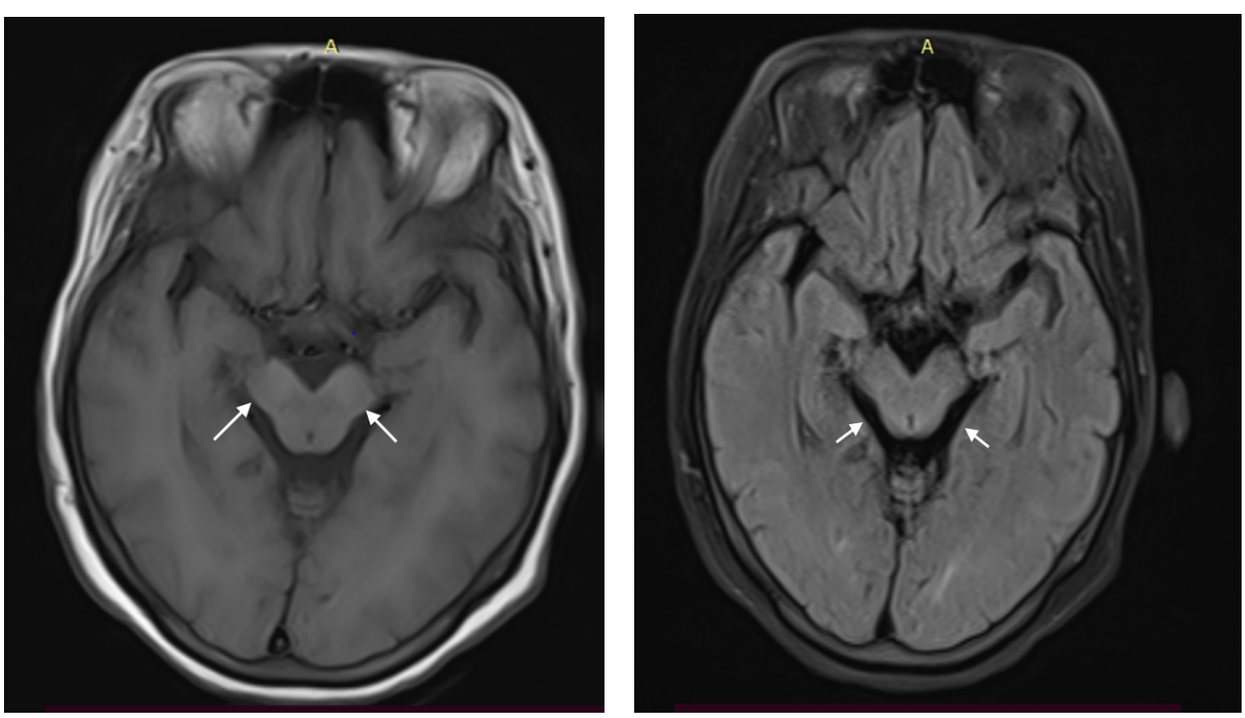

Figure 1. MRI brain: 1a. T1 axial image showing signal change in bilateral cerebral peduncle, 1b. FLAIR axial showing signal change in mid brain.

The patient was admitted to the intensive care unit with the diagnosis of HE grade 1, HE induced CB with papilledema and CLD (Child-Pugh C Classification, MELD-Na score 18). Point of care ultrasound showed a raised optic nerve sheath diameters (ONSD) of 6.2mm and 6.3mm in right and left respectively (Figure 2).

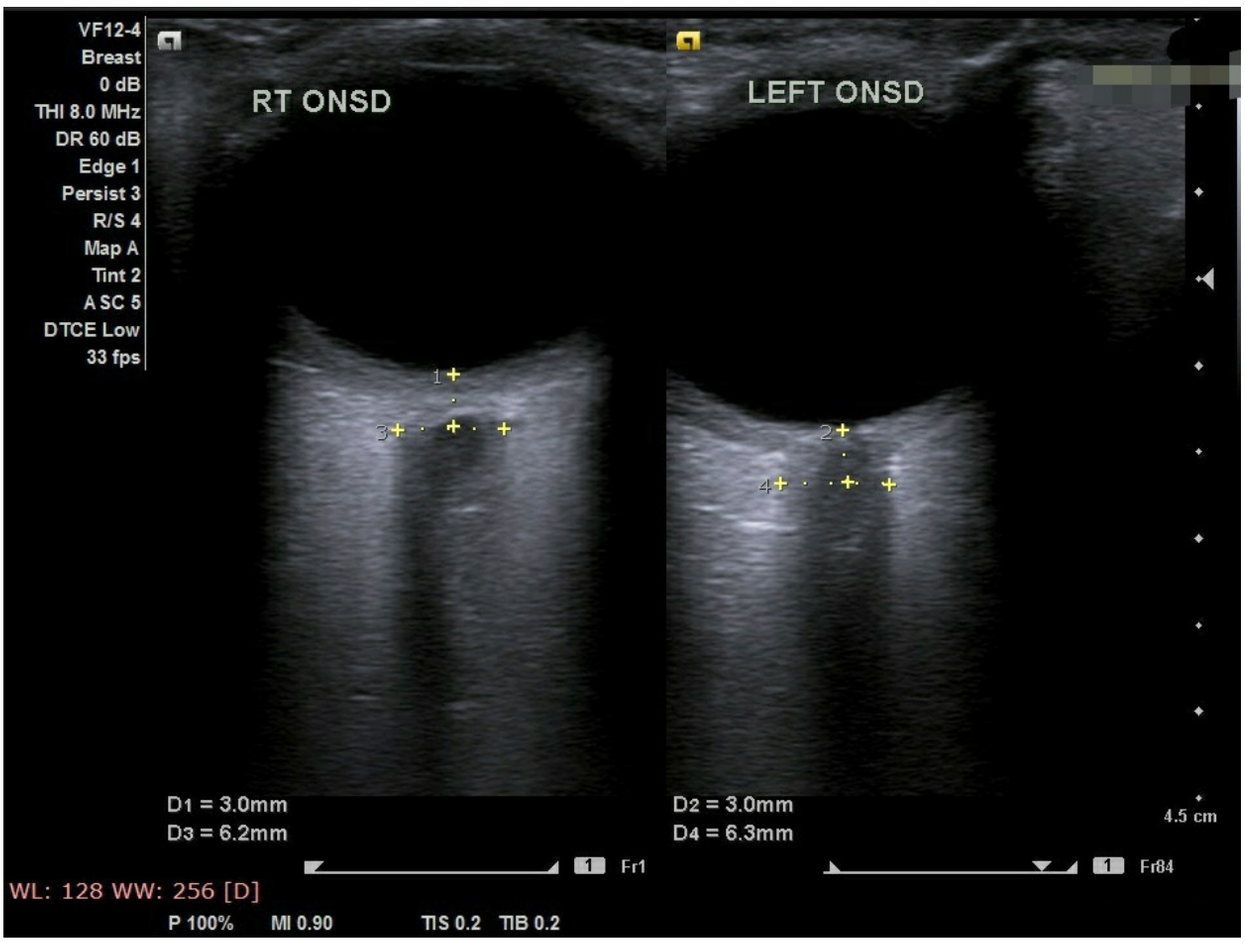

Figure 2. Bilateral optic nerve sheath diameter (ONSD).

Hyperosmolar therapy with 3% sodium chloride was initiated along with supplements of branched-chain amino acids and lactulose. Daily fundoscopy and measurement of ONSD were done in addition to neurological examination. On the second day of ICU admission which corresponds to the fourth day of her visual disturbances, her GCS slowly began to fall to 10/15 (E3V2M5) in the morning and further reduced to 8/14 (E2V2M4) by the night. The stool for occult blood was negative which effectively ruled out UGI bleed. An electroencephalogram (EEG) was done which ruled out non-convulsive seizure activity and any other lateralizing sign of seizure. Visually evoked potential could not be conducted. A lumbar puncture was done which showed raised opening pressure of 23 cm of cerebrospinal fluid (CSF) with no cells with sugar and protein within normal ranges. CSF for adenosine deaminase was found to be negative along with negative CSF culture.

Same treatment was continued. On third day of ICU admission, her GCS started to improve. Fundoscopy showed decreasing papilledema. ONSD also started to decrease to bilateral 5.7mm. In the morning, her GCS was 14/15 (E4V4M6), and then 15/15 (E4V5M6) by evening. On the fourth day of ICU admission, her visual acuity started to improve to a perception of light and gradually to hand movement. She was shifted to ward on day six with full recovery of her vision. She was discharged home after three days.

DISCUSSION

Cortical blindness refers to visual loss due to bilateral lesions of the visual pathways in the temporo-occiptal lobes, with normal pupillary light reflexes and normal fundi.[2] In HE, metabolic disturbance can lead to various symptoms, such as personality changes, disturbances in sleep rhythms, stroke simulation, periodic alternating gaze deviation, and CB.[3] There are only a few case reports of cortical blindness in hepatic encephalopathy. These symptoms can even precede the onset of altered sensorium.

In HE induced CB, the blindness is severe, but transient and reverts to normal as patients recover from HE. However, in one case study of acute liver failure following paracetamol overdose, CB following HE was permanent.[2] Cerebral edema is thought to contribute to

neurologic dysfunction seen in HE. Glutamine, produced by the metabolism of ammonia via glutamine synthetase, attracts water into the astrocytes, leading to swelling and induces oxidative dysfunction of the mitochondria.[4] One study has suggested that further insults like hypotension following gastrointestinal bleeding, in addition to the encephalopathy, may also contribute to vision loss.[2] Contrary to this study, our patient was normotensive with no active gastrointestinal bleeding at the time of developing her visual disturbances.

In all of the reported cases on HE induced CB, none of the cases had associated papilledema in their finding. Although increased intracranial pressure is present in more than 80% of patients with fulminant hepatic failure, patients with encephalopathy secondary to chronic liver disease have shown not to develop elevated intracranial pressure.[5] However, one study has reported two cases of chronic liver disease in a hepatic coma with raised intracranial pressure (ICP) documented, among which one of them had papilledema associated with generalized tonic-clonic seizures and decerebrate posturing.[5] Our case had raised ICP as shown by the increased ONSD and raised CSF opening pressure with papilledema. Hyperosmolar therapy was helpful to reduce the raised ICP as evident with the decreasing ONSD and resolution of papilledema. Seizure activity was ruled out with EEG.

HE presenting with atypical symptoms is not uncommon. These atypical symptoms related to HE can mimic the features of cerebral vascular disease (e.g., the onset of confusion and disorientation, and sometimes neurological deficits).[3] Wishart et al reported an unusual case of HE who presented with global aphasia.[6] Similarly, another case study reported a case of HE with reversible focal neurologic signs resembling an acute stroke.[7] Therefore careful investigation of physical findings, radiological imaging must be done to rule out cerebrovascular events to make an early diagnosis.

CONCLUSIONS

Cortical blindness is a rare presentation of hepatic encephalopathy which can be associated with papilledema and raised ICP. However other causes must be ruled out before associating these two entities. As seen in our case, resolution of HE and ICP usually leads to improvement in the vision.

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CONFLICT OF INTEREST

None.

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