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Case Report

Reversible Cortical Blindness with Bilateral Ptosis: A Rare Feature of Hepatic Encephalopathy. A Diagnosis Rarely Thought!

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ABSTRACT

Hepatic encephalopathy (HE) reflects a spectrum of neuropsychiatric abnormalities seen in patients with liver dysfunction and is diagnosed after the exclusion of other known causes of cerebral dysfunction. Rarely, visual cortex may be affected in patients with liver disease and cortical blindness (CB) may occur. It may be the presenting feature of HE and may be mistaken for cerebral stroke, if it other features of HE are not present. It is usually transient and resolves with efficient treatment of encephalopathy. We report a case of transient CB with bilateral ptosis in a young patient of hepatic encephalopathy due to hepatitis B virus related chronic liver disease in view of its rarity and unusual presentation.

Key words: Visual loss, ptosis, liver disease, coma.

INTRODUCTION

Hepatic encephalopathy (HE) frequently occurs in patients with chronic liver disease. [1] It is a constellation of potentially reversible neuropsychiatric abnormalities seen in patients with liver dysfunction and/or portosystemic shunting. Signs and symptoms vary, but usually include disturbed consciousness, personality changes, intellectual deterioration, speech disturbance, asterixis and sleep disturbance (Sherlock and Dooley 1993). In rare cases of hepatic encephalopathy, visual cortex may be affected and cortical blindness (CB) may occur, which resolves following treatment of hepatic encephalopathy. There has been only few case reports of transient CB associated with hepatic encephalopathy. We report a previously unreported complication of transient CB with bilateral ptosis as a presenting feature of hepatic encephalopathy in a young man.

CASE REPORT

36 year old gentleman presented to our hospital with history of painless visual impairment which progressed to complete visual loss in both eyes over 2 days. It was associated with forgetfulness and confusion. There was no history of fever, double vision, motor and sensory symptoms in upper and lower limbs, urinary and fecal incontinence, convulsions and head injury. Patient had consumed herbal medicine for common cold, 5 days prior to the development of above symptoms. There was no history of alcohol and tobacco abuse and no exposure to blood and blood products in the past.

In May 2014 patient was diagnosed to have chronic Hepatitis B infection (e antigen positive) with decompensated cirrhosis of liver (COL) and portal hypertension.

However, he was not on any specific treatment for the same. Clinical examination

revealed confused, restless and drowsy patient with pallor, jaundice, few small ecchymotic patches over forearm and bilateral pitting pedal edema. His body temperature was 98.6^0F, pulse rate 78/minute, BP- 140/70 mm Hg and respiratory rate was 28/minute with accessories working. Abdomen was soft and non tender. There was mild splenomegaly with ascites. Liver was not palpable. His Glasgow Coma Scale was 12/15. Pupils were normal sized. Direct and consensual light reflexes were normal in both eyes. Examination of the anterior segment of the eyes was normal. Fundi examination revealed healthy disc, macula and peripheral retina in both eyes. There was partial ptosis in both eyes. Extra ocular movements were normal and painless in both eyes. Motor system examination revealed normal tone and power. The deep tendon reflexes were normally elicitable and plantars were flexors. He demonstrated hepatic flap. There was no sensory deficit. Neck was soft. His cardiovascular and respiratory system examination was normal. His hemoglobin was 10gm/dl, total WBC count- 6,500 cu mm with normal differential count, macrocytosis on peripheral smear, MCV-104.6fl and platelet count- 1.02 lakhs/cu mm. His liver function test revealed total bilirubin-5.5 mg/dl, direct- 2.6 mg/dl, indirect 2.9 mg/dl, ALT-43U/L, AST-75U/L, ALP-150U/L, total serum proteins 6.9 mg/dl, serum albumin- 2.3 mg/dl and globulin-4.6 mg/dl PT-19 sec, control-11sec, PT (INR)-1.7. Cerebrospinal fluid examination was unremarkable. Electroencephalogram (EEG) done on 3rd day of admission showed diffuse delta-theta activity of the cerebral cortex. Ultrasound of abdomen revealed hyperechoic, nodular, shrunken liver with splenomegaly (16cms) and moderate ascites. Ascitic fluid analysis showed transudative picture and its culture was sterile. Doppler study of hepatic and portal veins did not reveal any thrombus. Upper gastrointestinal endoscopy showed portal hypertensive gastropathy without esophageal varices. Computerized tomography (CT) scan of brain was normal. Magnetic resonance imaging (MRI) brain (Fig.1) with gadolinium enhancement showed bilateral hyperdensities in the basal ganglion in T1 weighted image. Rest of the brain parenchyma was normal. Flash visual evoked potentials (VEPs) were absent in both eyes. Normal papillary reaction and retina with absent flash VEP was consistent with cortical blindness.

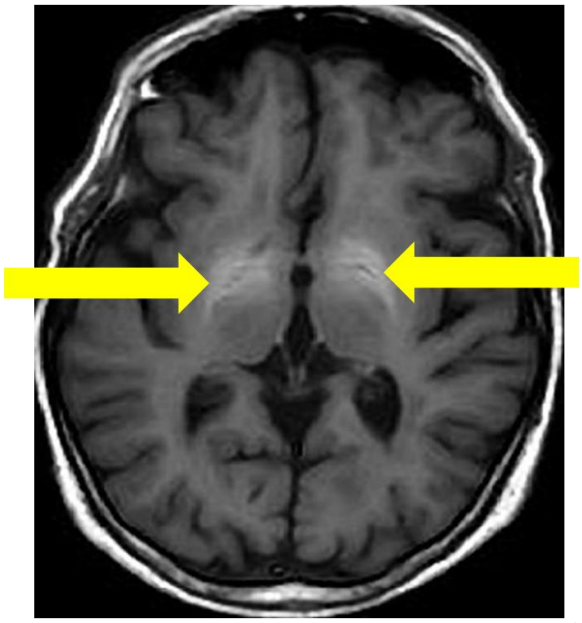

Figure 1: MRI brain - T1 weighted image showing areas of hyperdensities in the region of basal ganglion

In view of further deterioration in his mental status, he was managed in intensive care with endotracheal intubation, supplemental O_2 @ 3L/mt, syrup lactulose 30 ml qid, cap rifamixin 555 mg bd, ceftriaxone 1 gm bd intravenously (IV), 20% Human Albumin 100 ml for 3 days IV, L-ornithine L-aspartate 15g tid IV, tab adefovir dipivoxil 5 mg od, vitamin K 10 mg od IV, and IV fluids according to the central venous pressure. Patient improved completely with above treatment. His EEG normalized, his ptosis disappeared and he was discharged on 21st day of admission with diagnosis of HBV related decompensated COL-Child- Pugh's class C, transient cortical blindness (CB) with bilateral ptosis due to grade IV hepatic encephalopathy.

DISCUSSION

Cortical blindness (CB) refers to visual loss (complete/incomplete) in the presence of normal papillary reflexes, extra ocular movements and ocular fundi. It is caused by bilateral damage to the geniculocalcarine fibres or striate cortex of the occipital lobes (V1). [1] This may mimic functional loss, as fundi and pupillary reflexes are normal. The common cause of CB are thromboembolic disease involving the vertebrobasilar system, cerebral and coronary angiography (Aldrich et al 1987), sudden cardiac arrest with hypoxic encephalopathy, hypertensive encephalopathy, drugs and preeclampsia. [2,9] Metabolic causes reported in literature are hepatic encephalopathy, uremia, hypoglycemia, diabetes mellitus, CO_2 narcosis and acute intermittent porphyria. Patient with significant neuroradiologic lesion has poor chance of complete recovery of vision.

In rare cases of hepatic encephalopathy (HE), visual cortex may be affected and CB may occur before the alteration in consciousness. The reason for the preferential injury to the visual cortex and its exact mechanism remains obscure. The highly permeable blood brain barrier (BBB) in HE allows selective entry of toxic neurotransmitters to occipital cortex. [3] With treatment when the toxic level of these neurotransmitters diminishes and the blindness improves. Vascular insufficiency is also a proposed mechanism. CB due to hepatic encephalopathy has good prognosis (Dalman et al 1997). [4] It usually recovers following treatment.

CB is a very rare complication and sometimes may be the initial manifestation of hepatic encephalopathy when other cerebral symptoms are mild. In such cases, differentiating it from posterior circulation stroke becomes difficult. There have been only ten case reports of CB due to hepatic encephalopathy published in literature of which 7 cases had complete recovery. [5-7,9] Van Pesch et al (2006) described a case of recurrent ictal CB associated with occipital lobe MRI abnormalities in a patient suffering from end stage liver disease who had partial improvement of vision. [8] Ammar et al (2003) reported a case of permanent CB following grade II hepatic encephalopathy with normal imaging in 19 years old boy with acute liver failure due to paracetemol and ephedra overdose. [1] Miyata et al (1988) reported CB in a patient with recurrent episodes of hepatic encephalopathy precipitated by upper GI bleed, in whom imaging and EEG failed to reveal any abnormality. [5]

However, none of the cases reported earlier in the literature had associated bilateral ptosis, thus making our case unique and first of its kind to be reported. In our patient, use of herbal medicine probably led to liver injury and precipitated in hepatic encephalopathy, considering the fact that the symptoms started five days after taking it. The exact cause of ptosis could not be explained. MRI brain did not reveal any lesion in the midbrain. The imaging of the occipital cortex was also normal but there was increased intensity in T1 weighted MRI. This is found in patients with chronic liver disease with or without hepatic encephalopathy and is related to manganese deposition. The purpose of this report is to highlight the rare clinical features of hepatic encephalopathy, thereby creating awareness of this entity. This case also emphasis the need for prompt and efficient treatment of hepatic encephalopathy, correction of which leads to reversal of blindness. Taking a lesson from the above case, though it is said that hepatic encephalopathy is diagnosis of exclusion, it is prudent to rule it out before embarking on any other diagnosis.

CONCLUSION

Hepatic encephalopathy can rarely present with cortical blindness which may be mistaken for cerebrovascular disease. Meticulous history, careful physical examination, appropriate investigations and high index of suspicion are required to come to proper diagnosis. Early detection and appropriate therapy for HE may have a good

prognosis and favorable outcome.

REFERENCES

1. Ammar T, Auwzinger G and Michaelides M. Cortical blindness and hepatic encephalopathy. Acta Ophthalmologia Scand 2003:81; 402-4.
2. Aldrich MS, Alessi AG, Beck RW and Gilman S. Cortical blindness: etiology, diagnosis and prognosis. Ann Neurol 1987: 21; 149-58.
3. Biswas AK, Banerjee A and Bala S. Reversible cortical blindness in a case of hepatic encephalopathy. Med J DY Patil Univ 2016; 9:254-6.
4. Dalman JE, Verhagen WIM and Huygen PLM. Cortical blindness. Clin Neurol Neurosurg 1997; 99: 282–6.
5. Miyata Y, Motomura S, Tsuji Y and Koga S. Hepatic encephalopathy and reversible cortical blindness. AM J gastroenterol 1988:83: 780 – 82.
6. Naparstek Y, Shouval D, Auerbach E and Eliakim M. Transient cortical blindness in hepatic encephalopathy. Isr J Medsci 1979:15; 854-6.
7. Chen CM and Chen PC. Transient cortical blindness in liver cirrhosis. Dig Dis Sci 1998:43; 365-7.
8. Pesch VV, Hernalsteen D, Rijckevorsel KV, Duprez T, Boschi A, Ivanoiu A, et al. Clinical, electrophysiological and brain imaging features during recurrent ictal cortical blindness associated with chronic liver failure. Acta Neurol Belg 2006:106, 215-8.
9. Cheng-Tagome S, Yamamoto A, Suzuki K, Katayama N and Imai H. Cortical blindness induced by hepatic encephalopathy: case report and review of published case reports. Acute Medicine & Surgery May 2016. doi: 10.1002/ams2.225

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