Portosystemic Shunt Presenting as Rapidly Progressive Dementia

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Abstract
Rapidly progressive dementia (RPD) is a challenging clinical problem in the elderly. It encompasses a wide range of diseases. Thorough clinical examination and a systematic approach is essential to find the cause of RPD. Early recognition of causes of RPD is important for early treatment and reversal of the pathology. We report a case of RPD due to portosystemic encephalopathy secondary to a large portosystemic shunt with preserved liver functions, in a previously healthy elderly male. He had pallidal hyperintensities on MRI brain that can be the clue to the diagnosis of portosystemic shunt and chronic liver disease in patients with RPD.

Introduction
Portosystemic shunts (PSS) remain an unrecognized cause of neurological or psychiatric disorders. This condition has been essentially reported in patients with portal hypertension secondary to chronic liver disease such as hepatic cirrhosis, often linked to liver damage due to alcohol or viral hepatitis. Very few cases have been reported so far, in which a portosystemic shunt presented as psychiatric and neurological disorders without prior history of liver disease. We are presenting one such case that remained diagnostic dilemma for a long time.

Case History
An 80 years old male retired school teacher, premorbidly a well-adjusted cognitively intact individual, functioning at an age appropriate level, presented to the outpatient psychiatric service with a history of abnormal behaviour of 20 days duration. He had outbursts of anger and had assaulted his wife accusing her of infidelity. Subsequently he had recurrent episodes of aggression towards his wife. He admitted visual hallucinations but denied auditory hallucinations. He was perplexed and had crying spells. There was no past history of alcohol intake, head injury, cognitive impairment, seizures, confusion or similar change in behaviour. He admitted visual hallucinations but denied auditory hallucinations. He was disoriented to time and person and had recent memory impairment. His mini mental state examination score was 13/30. There was no focal neurological deficit. There were no signs of chronic liver disease (CLD) or liver cell failure like asterixis, constructional apraxia, spider angioma, loss of body hair, ascites, splenomegaly or shrunken liver. Systemic examination was otherwise unremarkable. He was admitted under Geriatrics for evaluation of cognitive impairment and to rule out organic causes of psychosis.

Considering cognitive decline and psychotic symptoms of short duration, he was investigated for causes of subacute confusional state and treatable causes of dementia. A possibility of Lewy body dementia was considered as the patient had prominent visual hallucinations. However, acute onset, short duration of symptoms and absence of features of parkinsonism were against this diagnosis. Biochemical investigations revealed normal liver functions with an albumin of 3.5 g/dl. Serum electrolytes, renal functions, calcium and vitamin B12 levels were normal. VDRL was non-reactive. His EEG was normal. Magnetic resonance imaging (MRI) of the brain revealed hyperintensities in the globus pallidus bilaterally suggestive of manganese deposition raising the possibility of CLD. He was started on Quetiapine 25 mgs twice daily as a symptomatic measure.

Ultrasound examination of the abdomen revealed liver with mild coarse echotexture, mild splenomegaly and normal portal vein. There was no ascites. Serum ammonia levels were normal. HBsAg by ELISA was weak positive but HBeAg was negative and HBV DNA levels were negligible. Prothrombin time was normal.
In the ward, the patient had persistent visual hallucinations and disorientation. The Quetiapine dose was gradually increased to 400 mg daily. Gastroscopy was planned but could not be done as patient was not cooperative. Suspecting CLD, oral amoxicillin and lactulose were initiated.

As the picture was not typical of hepatic encephalopathy, further evaluation for RPD was done in consultation with hepatologist. CSF analysis and blood and CSF manganese levels were normal. Thyroid antibodies and anti-TTG antibody were negative. Whole body Positron Emission Tomography CT done to rule out paraneoplastic limbic encephalitis and occult malignancy was negative. The images however showed tortuous collaterals at gastoesophageal junction suggestive of portosystemic shunt.

Soon afterward on the 28th day of admission the patient developed hematemesis. Emergency upper GI endoscopy revealed grade 4 varices with stigmata of recent hemorrhage, and sclerotherapy was done. Serum ammonia repeated at this time was high (110 mg/dl). At this juncture the patient had asterixis and constructional apraxia. He was managed with octreotide and pantoprazole infusion. Suspecting a large PSS, the patient was started on rifaximin, propranolol, low protein diet, oral lactulose and twice daily lactulose enema. He made a good recovery over the next week with improvement of MMSE to 23/30, disappearance of visual hallucinations, asterixis and constructional apraxia. His episodes of aggression also gradually declined.

This patient, who presented as RPD was diagnosed to have PSE on the basis of a large PSS with early CLD and preserved synthetic liver function. Patient was under regular follow up with geriatric medicine showing steady improvement in his mental status. Nine months after discharge patient died as a consequence of head injury following an accidental fall.

**Discussion**

Very few cases of PSE due to PSS without a prior history of liver disease have been reported in the literature since the first description in 1964.1 Our patient presented with RPD and psychosis and MRI revealed hyperintensities in the globus pallidus bilaterally suggestive of manganese deposition which was suggestive of chronic liver disease. As per the literature upto 90% of MRI brain studies in patients with CLD are abnormal with hyperintensities in the globus pallidus bilaterally on T1W images and have abnormal MR spectroscopy. These hyperintensities are thought to be due to manganese deposition but do not correlate well with the presence of hepatic encephalopathy. They increase after doing shunt procedures and disappear after liver transplantation or closure of the shunt.2-4 Patients can present with predominant neurological and psychiatric symptoms without showing any features of CLD in cases of large PSS with relatively preserved synthetic function of the liver. This was seen in our patient with normal albumin, transaminases, serum ammonia and manganese levels along with absence of asterixis and constructional apraxia at presentation.

In a recent review article, only 21 cases of PSS revealed by neurological or psychiatric symptoms with preserved synthetic liver function were found in the English literature between 1964 and 2011. Inclusion criteria were: (a) neurological or psychiatric disorders unexplained by another cause, (b) no known history of liver cirrhosis or liver disease prior to neuropsychiatric symptoms, (c) normal or slightly disturbed liver function tests including prothrombin rate above 70%, albumin above 30 g/L, normal total and conjugated bilirubinemia, (d) liver enzymes below 2.5-fold the normal values and (e) evidence for significant PSS on abdominal imaging by ultrasound or CT.5 Our patient fulfilled all the above criteria.

Serum ammonia levels are highly labile and should be repeated if there is a high index of suspicion. Though hyperammonemia has a poor sensitivity for hepatic encephalopathy, in the recent review hyperammonemia was reported as a good screening test for patients presenting with PSE due to a PSS.6 In our patient, initial ammonia was normal but repeat ammonia levels were elevated. Demonstration of the shunt either by gastroscopy, ultrasonography or by CT abdomen is essential in order to diagnose PSS. The majority of similar patients reported in literature had a congenital shunt or post-surgical shunt and a high index of suspicion is required for the early diagnosis.6 Almost all the patients responded to low protein diet and lactulose. To conclude, pallidal hyperintensities on MRI can be the clue to the diagnosis of underlying chronic liver disease or encephalopathy in patients with RPD.

**References**


6. Watanabe A. Portosystemic encephalopathy in non cirrhotic patients.