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Chronic Alcoholic Liver Disease with Degenerative Cervical Myelopathy - Quadriparesis

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Abstract: ***Hepatic myelopathy (HM) is a rare neurological complication of chronic liver disease[1]. We report a rare case of 70- year old male patient with chronic alcoholic liver disease presenting as degenerative cervical myelopathy with normal sensory examination, impaired cerebellar system, progressing as spastic quadriparesis without bowel and bladder complaints.***

Keywords: Hepatic myelopathy, quadriparesis, alcoholic liver disease

1. Introduction

Degenerative cervical myelopathy (DCM), earlier referred to as cervical spondylotic myelopathy, involves spinal cord dysfunction from compression in the neck. Here we report a case of 70- year old male presenting with pure motor quadriparesis without bowel and bladder complaints and normal sensory examination with impairment of cerebellar system. Myelopathy is a rare complication of chronic alcoholic liver disease. It's main clinical features is spastic paraparesis without sensory or sphincteric impairment. A progressive spastic paraparesis in patients with hepatic failure was first described by Leigh and Card[1-3]. Patient with chronic liver disease frequently experience neurological problems, with hepatic encephalopathy being the most common. Comparatively rare is the involvement of the spinal cord; the so-called hepatic myelopathy. The unusual thing about our patient is that cervical spinal cord was affected, which is rare compared with thoracic cord involvement in hepatic myelopathy. Spinal cord signal intensity abnormality on MR imaging should raise suspicions of HM.

2. Case Report

A-70- year old male patient, farmer by occupation, from Karaikal, Puducherry, presented in general medicine OPD with a history of difficulty in walking because weakness and stiffness of both the lower limbs associated with bilateral leg swelling for 2 years. The weakness gradually progressed over 8 months to involve bilateral upper limbs with difficulty in buttoning and unbuttoning his shirt. Patient was walking with support and had tremors during household work. There was no history suggestive of sensory impairment, symptoms of bowel and bladder involvement, ocular or vision abnormalities, seizures, diabetes mellitus, hypertension, tuberculosis, trauma, exposure to toxins or radiation, blood or blood component therapy, bleeding disorders, promiscuity, or similar complaints in the family. There was no history of fasciculations or Wasting. He had significant alcohol intake of about 70-90 grams of alcohol at least 3 times a week since the past 30 years. He is occasional smoker and betel chewer. General examination revealed normal vitals, average nutrition, pallor, pedal edema in both the lower limb and mild hepatomegaly along with splenomegaly. Other physical markers of liver disease such as icterus, spider angioma, and palmar erythema, were not present. On central nervous system examination patient was conscious, alert with mild impairment of attention. Cranial nerve examination and sensory system was normal. Motor power of all limbs muscles was grade 3/5 (MRC grading). Muscle tone was increased in all limbs (Grade 1 spasticity according to modified Ashworth Scale) with brisk deep tendon reflexes and plantar response was bilaterally extensor. He had ankle and patellar clonus. All modalities of sensation in upper and lower extremities were normal. There was evidence of cerebellar signs, butno sign of meningeal irritation.

3. Investigations

Investigation revealed mild anemia 10.5 g/dl (N-14-16 g/dl), normal total white blood cell count and platelet count. Liver function test showed mild hyperbilirubinemia and raised liver enzymes. Total bilirubin/direct bilirubin-1.8/0.5 (mg/dl), AST-120 U/L (N-12-38 U/L), ALT-95 U/L (N-7-41 U/L), ALP-220 (N-30-180 U/L), total protein (4.8 gm/dl) and albumin (2.5 gm/dl). Renal function tests and serum electrolytes were normal. Serum ammonia level was high68 mg/dl (N 15–60 mg/dl). Serum vitamin B12 (N 279–996 pg/ml) and folic acid levels (N 5.4–18.0 ng/ml) were normal. Ultrasound of abdomen showed coarse echo texture of liver with mild splenomegaly, a dilated portal vein and mild as cites. Upper gastrointestinal endoscopy and Electroen cephalography was normal. Patient serum was non-reactive to viral markers of hepatitis A, B, C, HIV 1 &HIV 2.HbA1c level was normal 5.1.

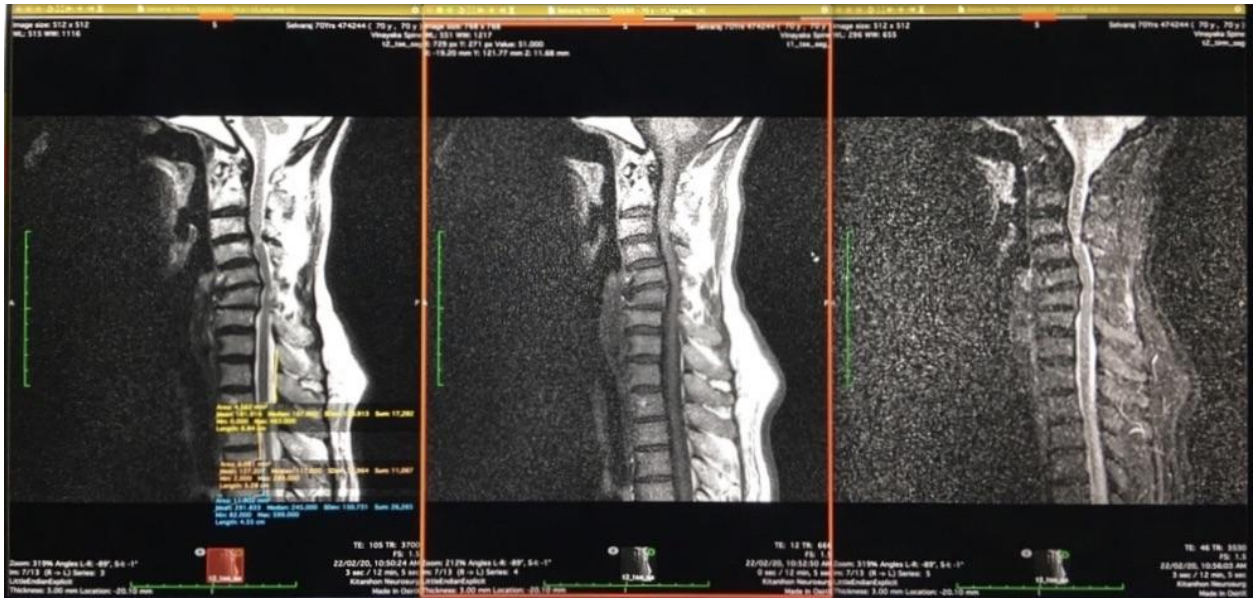
4. MRI Findings

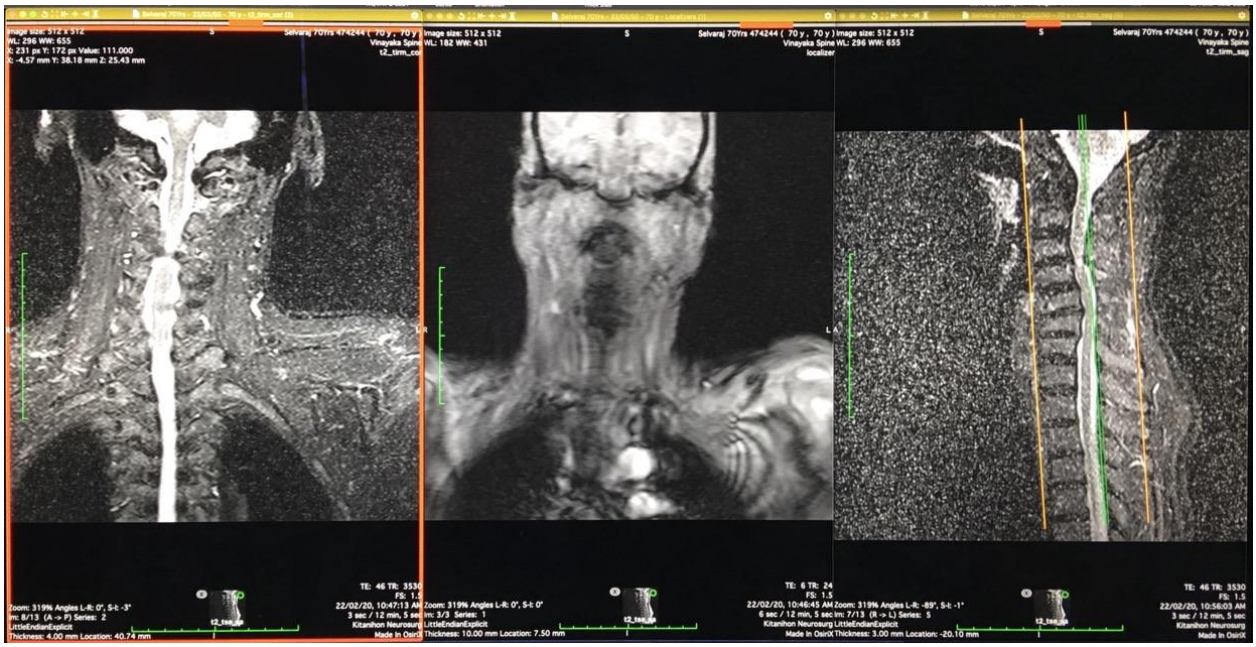
MRI brain (plain)-showed a marked cortical and subcortical cerebral atrophy. No evidence of infract or intracranial

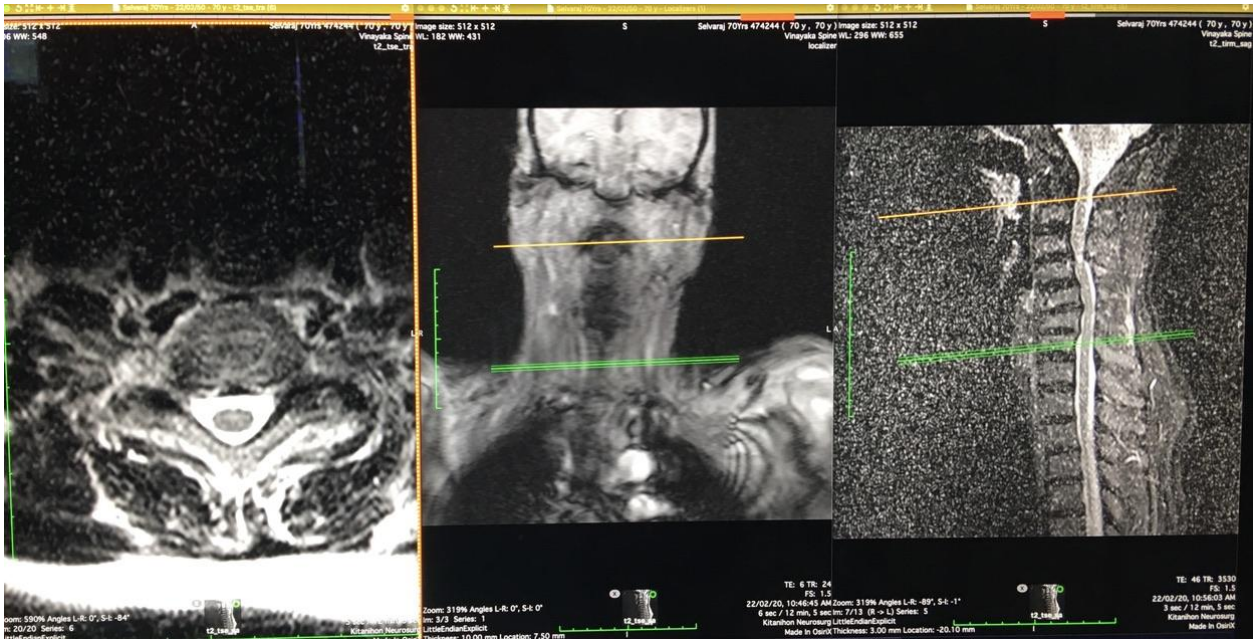
hemorrhage. Cerebellar hemispheres, brain stem and 4th ventricles are normal.

MRI- Cervical Spine

Sagittal and axial STIR and T2-weighted MR imaging of the cervical spinal cord, showed hyperintensities that involve the cervical spinal cord. No spinal cord enhancement on a postcontrast sagittal T1-weighted image. Spinal cord stenosis noted. Spinal cord thinning with atrophy and possible syrinx/edema is also noted at C3-C4 cervical cord.







5. Discussion

The pathogenesis of hepatic myelopathy is poorly understood. The first documentation of hepatic myelopathy from the Indian subcontinent was provided by Pant et al[2, 5].There is a less literature of hepatic myelopathy causing quadriparesis that is mainly affecting cervical cord. Our patient had cervical myelopathy causing quadriparesis wherein the upper limbs as well as lower limb have been equally involved[6]However, owing to variability of clinical presentation, cervical myelopathy due to hepatic cause can be very difficult to diagnose. Our case is interesting for several reasons. Firstly, this patient presented with a long history of progressive quadriparesis with no prior episode of hepatic decompensation. It has been postulated that alteration of the hepatic metabolism leads to deficiencies of essential nutrients for the central nervous system[5]. Nitrogenous products such as ammonia, fatty acids, indoles and mercaptans, bypassing the liver through the portocaval shunt, play a contributory role[8]. These nitrogenous products cause injury tothe axon cylinders, neuronal cell bodies and myelin. Furthermore, a discrepancy exists between the tissue reaction in the corticospinal tract, where axonal degeneration and demyelination, cytoplasmic astrocytosis and round cell infiltration occur and sparing of other systems in the spinal cord. Differential diagnosis of hepatic myelopathy includes amyotrophic lateral sclerosis, multiple sclerosis, paraneoplastic effect, radiation myelopathy, human T-Lymphotropic Virus Type 1 associated myelopathy and vascular spinal cord disease. Our patient had chronic alcoholic liver disease, presented as pure motor quadriparesis, sparing of bowel, bladder and sensory system[3].We believe that the overall clinical picture, results of laboratories and imaging studies described above are sufficient to make a diagnosis of chronic alcoholic liver disease with degenerative cervical myelopathy (Hepatic myelopathy)[7].The treatment of HM is difficult and the progression of spastic weakness is relentless[9]. Conservative treatment strategies for HM include liver protection, neurotropic drugs, and measures to control blood ammonia concentration. However, as previously mentioned, HM responds poorly to conservative medical therapy. In particular, in contrast to hepatic encephalopathy (HE), HM usually does not respond to blood ammonia lowering therapies[10-14].Surgical treatment options in HM currently include liver transplant (LT), surgical ligation, shunt reduction or occlusion by interventional procedures. Surgical ligation has been reported to be effective, but is only used occasionally. Surgery is recommended in patients with moderate or severe degenerative cervical myelopathy.

6. Conclusion

We report an unusual instance of chronic alcoholic liver disease with spastic quadriparesis without any obvious features of hepatic encephalopathy. Hepatic myelopathy is a serious complication associated with portal hypertension and portosystemic shunting. Early identification of this disorder and exclusion of other treatable causes is important. It remains however a diagnosis of exclusion. Early diagnosis and prompt management of liver disease may improve overall prognosis of neurological complications.

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