A Rare Complication of Cirrhosis: Hepatic Myelopathy

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ABSTRACT

Introduction: Hepatic myelopathy is characterized by spastic paraparesis and minimal sensory abnormalities in patients with cirrhosis, particularly those with portosystemic shunts that have been created surgically or have occurred spontaneously.

Materials and methods: We described a patient who presented with spastic paraparesis and diagnosed as hepatic myelopathy when all the other possible diagnoses were ruled out with a review of literature in the goal of promoting prompt recognition and enhancing understanding of HM.

Results: We reported a patient aged 51 years who presented with spastic paraparesis with insidious onset and progressive course and diagnosed as hepatic myelopathy.

Conclusion: Unlike hepatic encephalopathy, hepatic myelopathy is usually considered irreversible.

Keywords: Hepatic myelopathy, Cirrhosis, Spastic paraparesis.

INTRODUCTION

Hepatic myelopathy (HM) is characterized by progressive spastic paraparesis and minimal sensory abnormalities in patients with chronic liver disease, particularly those with portosystemic shunts forming surgically or spontaneously. It may have a significant impact on mobility and quality of life. Neuropathological studies of the patients with hepatic myelopathy have demonstrated demyelination of the lateral corticospinal tracts with various degree of axonal loss.

MATERIALS AND METHODS

Early and accurate diagnosis of HM is important because patients with early stages of the disease can fully recover following liver transplantation. In this study, we described a patient who presented with spastic paraparesis and diagnosed as hepatic myelopathy when all the other possible diagnoses were ruled out with a review of literature in the goal of promoting prompt recognition and enhancing understanding of HM.

RESULTS

Clinical Examination

This is a 51-year-old patient who was followed for 2 years in the service of gastroenterology for post-hepatitis B cirrhosis discovered as a result of inaugural edema and ascitic decompensation. One year later, he was taken in intensive care unit for hepatic encephalopathy with a favorable outcome.

His actual history dates back about 6 months before admission, marked by the installation of low back pain without radicular symptoms, followed four months later by the appearance of gait disorders evolving progressively into a bedridden state and associated with vesico-anal incontinence.

On examination, he had a well general condition without clinical signs of decompensation of the cirrhosis, but he had a sub-conjunctival jaundice, moderate hepatomegaly with firm lower border. On the neurological examination, he was conscious with preserved higher functions, walking was impossible and he had a four-pyramidal syndrome with complete spastic paraplegia without superficial or deep sensory disturbances, there was no atrophy or fasciculation’s.

In view of its clinical and biological data, cirrhosis was classified as Child-Pugh B.

Radiological Findings

The MRI showed no spinal cord compression or signal abnormality of the spinal, but noted a lumbar epidural lipomatosis (Figs 1A to F).

Treatment Plan

The history of liver cirrhosis, the clinical context and the negativity of the MRI allowed us to make the diagnosis of hepatic myelopathy and refer the patient to an appropriate medical care.

DISCUSSION

Hepatic myelopathy was first described in 1949 by Leigh and Card as an encephalo-myelopathy, but due to its progressive...
and permanent character and nonaccessibility to medication, she was differentiated from hepatic encephalopathy as a separate entity in 1974 by Fukuda and Hirayama.\textsuperscript{1-3}

The pathogenesis is not well understood, two mechanisms have been proposed:

- The alteration of hepatic metabolism results in a deficiency of essential elements in the nervous system.
- The overproduction of ammonia, fatty acids, idols and mercaptan, which are unusual metabolites of the liver, resulting in axonal degeneration and demyelization.\textsuperscript{4}

The damage is localized preferentially at the dorsal spinal cord with preference for the cortico spinal pathway.

Lesions in the brain are responsible for Alzheimer’s disease type II.\textsuperscript{1}

The clinical manifestations of hepatic myelopathy are gait difficulty with insidious onset and progressive course. The upper extremities are rarely affected. Examination reveals symmetric spastic paralysis with hyperactive muscle stretch reflex of the lower extremities and extensor toe sign. Sensory findings are usually normal, and sphincter function is almost always preserved.\textsuperscript{1}

In the pathological study, as in typical hepatic myelopathy, myelin loss of the pyramidal tract was detected only in the spinal cord, and was particularly marked in the thoracic segments.

Our patient presented with exactly the same except that the clinical sphincter disturbances were, however, and unusually, too pronounced in our case. On the other hand, most of the myelopathy livers are preceded by episodes of encephalopathy, which was also the case in our patient. A completely normal MRI of our patient’s spinal cord excluded such etiologies, including epidural cord compression, intrinsic cord tumor, or infarction.

Faced with this clinical presentation, other etiologies should be mentioned: Amyotrophic lateral sclerosis, multiple sclerosis, post-radiation myelopathy, the association reached an HTLV-1 or vascular disease of the spinal...,\textsuperscript{2} but the age of presentation, rapidly developing disability, sparing of upper limbs, no sensory signs and imaging in a patient of chronic liver disease with features of hepatic decompensation should lead us to consider this diagnosis, with the knowledge that an association between cirrhosis and one of those diagnosis is always possible.

As we know, in the literature, there are no studies who spoke of imaging, particularly MRI, in this type of pathology.

Early diagnosis is important in case of chronic liver failure; reducing ammonia treatment can delay neurological manifestations. Liver transplantation demonstrates its effectiveness in improving paraplegia especially in the case of the less developed myelopathy.\textsuperscript{5-8}

\textbf{Figs 1A to F:} Spinal MRI sagittal T1-weighted (A and B) and T2 (C and D) revealed no abnormalities. The same MRI axial T1 (E) and T2 (F) showing the epidural lipomatosis.
CONCLUSION

Hepatic encephalopathy has long been the best-known neurological complication and best studied among those of cirrhosis. Recent studies have improved knowledge about other neurological complications, such as demyelinating myelopathy. It must henceforth come to mind of all neurosurgeon deal with an evocative picture.

REFERENCES