

SYRINGOHYDROMYELIA OR HTLV-I-ASSOCIATED MYELOPATHY/TROPICAL SPASTIC PARAPARESIS

A DIAGNOSTIC CHALLENGE

Case Report

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ABSTRACT – Human T-cell lymphotropic virus type I (HTLV-I) associated myelopathy / tropical spastic paraparesis (HAM/TSP) is the most common chronic myelopathy in Brazil. We present the case of a 53 year old man that fulfilled the diagnostic criteria for HAM/TSP but had at the magnetic resonance imaging (MRI) of the spinal cord evidences of syringohydromyelia at the C6-C7 and D2-D7 levels along with Chiari type I malformation. The clinical picture was more typical of HAM/TSP than of syringohydromyelia, which was probably asymptomatic. The present case clearly demonstrates that serology and neuroimaging should be always used together. We conclude that, specially in places where HTLV-I is endemic, every patient with a spastic paraparesis, even with a radiological picture suggestive of a structural spinal cord lesion, should have a screening test for HTLV-I. The clinical picture must dictate the final direction of the diagnosis.

KEY WORDS: HTLV-I, tropical spastic paraparesis, syringomyelia, hydromyelia, magnetic resonance imaging, differential diagnosis.

Siringo-hidromielia ou mielopatia associada ao HTLV-I: um desafio diagnóstico (relato de caso)

RESUMO – Mielopatia associada ao HTLV-I / paraparesia espástica tropical (MAH/PET) é possivelmente a mielopatia crônica progressiva mais comum no Brasil. Apresentamos o caso de um homem de 53 anos que, a despeito de preencher critérios clínicos e sorológicos para MAH/PET, exibia à ressonância magnética de coluna evidências de siringo-hidromielia de níveis C6-C7 e D2-D7, além de malformação de Chiari tipo I. Acreditamos que o quadro clínico seja devido à MAH/PET e não à siringo-hidromielia, esta, possivelmente, assintomática. Este caso demonstra que dados sorológicos e de neuroimagem devem sempre ser utilizados judiciosamente para o diagnóstico final de pacientes com suspeita de ambas as enfermidades. Concluímos que, particularmente em locais onde o HTLV-I seja endêmico, todo paciente com paraparesia espástica, mesmo com evidências radiológicas de lesão estrutural medular, deva ser submetido a avaliação sorológica para o HTLV-I. O quadro clínico deverá ser prioritário no estabelecimento do diagnóstico final.

PALAVRAS-CHAVE: HTLV-I, paraparesia espástica tropical, siringomielia, hidromielia, ressonância magnética, diagnóstico diferencial.

Human T-cell lymphotropic virus type I (HTLV-I) associated myelopathy / tropical spastic paraparesis (HAM/TSP) is probably the most common chronic myelopathy in certain regions of the world, such as in Brazil¹. Its diagnosis is based on a typical clinical picture coupled with laboratory

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Figs 1a and 1b. Spinal MRI (T2 ws) showing (a) Chiari type 1 malformation and syrinx cavities at C6-C7 and D2-D7 levels.

confirmation of HTLV-I infection in both sera and cerebrospinal fluid (CSF). In addition, neuroimaging should exclude structural causes for the syndrome².

We present a case that fulfilled the diagnostic criteria for HAM/TSP but had at the magnetic resonance imaging (MRI) of the spinal cord evidences of syringohydromyelia.

CASE REPORT

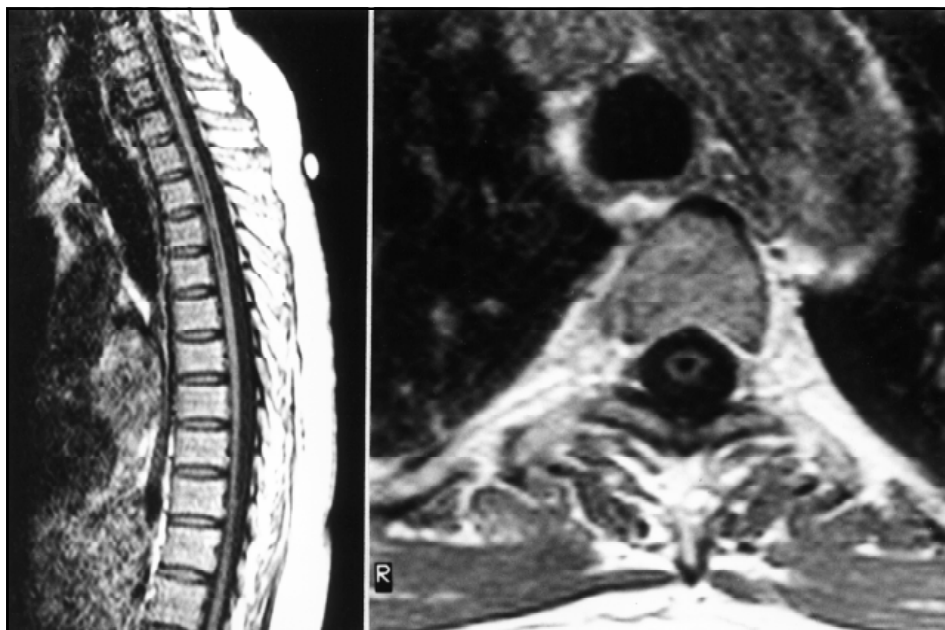
RPS, a 53 year old Brazilian white man presented a five year history of progressive walking and urinary difficulties. The neurological examination disclosed a spastic paraparesis, a tetrapyramidal syndrome and a neurogenic bladder. There was no pain or amyotrophy. The sensory examination was unremarkable. CSF examination was normal except for the presence of HTLV-I antibodies. An HTLV-I positive ELISA and Western blot analysis in serum were followed by PCR amplification of proviral sequences in peripheral mononuclear cells in blood and CSF. The brain MRI was normal; however an MRI of the spine revealed Chiari type 1 malformation, atrophy of the thoracic cord and syrinx cavities from C6-C7 and D2-D7 (Figs 1a, 1b, 2a and 2b).

DISCUSSION

The present case report is unusual since it demonstrates the coexistence of two diseases of the spinal cord in the same patient. As far as we know this coincidence has never been described and might have important implications in the diagnostic assessment of future cases of patients with chronic progressive spastic paraparesis.

HAM/TSP is histopathologically characterized as a chronic progressive inflammatory process with marked parenchymal exudation of lymphocytes and monocytes into both the gray and white matter of the spinal cord resulting in severe degeneration of the last one accompanied by striking glio-mesenchymal tissue reaction. Both the inflammation and the white matter degeneration are most conspicuous in the lower thoracic cord³.

MRI in HAM/TSP reveals mainly lesions in the periventricular white matter in addition to atrophy of the thoracic spinal cord⁴. Some authors have reported a correlation between histopathologic



Figs 2a and 2b. Spinal MRI (T1 ws) showing the syrinx cavities and cord atrophy.

features and MRI of spinal cord lesions. Abnormally high T2-weighted image signal intensities appeared nonspecifically in mildly altered lesions or areas with edema. In the gray matter, a low T1-weighted image in addition to a high T2-weighted image signal intensity appeared in severely altered lesions with necrosis, myelomalacia, or spongiform change. In the white matter, abnormally high T1-weighted image intensities appeared in severely altered lesions⁵.

Syringomyelia is a cystic cavitation within the spinal cord but not within the central canal. The cavity walls are not lined by ependyma. Hydromyelia is dilatation of the central canal of the spinal cord. Its walls are lined with ependyma. Syringohydromyelia can be either asymptomatic⁶ or associated with a variety of pathologic conditions, most notably Chiari 1 and 2 malformations, other posterior fossa developmental and acquired abnormalities, tumors of the spinal cord, inflammatory changes around the spinal cord, trauma, and almost any external spinal cord compressive lesion⁷.

Syrinx from isolated spinal arachnoiditis is a relatively rare occurrence. It has been associated with meningitis, subarachnoid hemorrhage, arteriovenous malformation, prior spinal surgery, prior spinal anesthesia, or oil-based myelography. The onset of symptoms tends to be over several years and can occur years or even several decades after the original insult. The syrinx associated with arachnoiditis tends to be progressive and recalcitrant to treatment⁸⁻¹⁰. In the present case, one could speculate that a severe necrotizing myelitis could lead to residual spinal cord cavitation. However, this hypothesis is weakened by the cervical and thoracic localisation of the lesions within the spinal cord coupled with the concomitance of a Chiari malformation.

Adult onset syringomyelia has been classically described as a progressive disease presenting in the second or third decade of life with amyotrophy, dissociated analgesia, paraparesis with scoliosis that can lead to neurogenic arthropathies and other trophic lesions. Generally symptoms develop gradually over 2-5 years. Adults with syringomyelia present with an assortment of complaints and neurologic findings that frequently consist of pain and point to dysfunction of the cervical spinal cord and/or the craniovertebral junction. It is worth noting that only 49% of patients with syrinx are

found to have dissociated sensory loss. Outside sensory and motor loss, pain is the other most predominant clinical feature of syringomyelia¹¹. We think that the clinical picture presented by our patient is more typical of HAM/TSP than of syringohydromyelia, which was probably asymptomatic in the present case. This is reinforced by the presence of a severe thoracic cord atrophy as shown in the MRI scan.

In HTLV-I endemic areas HAM/TSP is usually the first diagnosis to be considered when facing a patient with a chronic progressive spastic paraparesis. This frequently leads the physician to order HTLV serological tests for confirmation. In the presence of a positive serology most physicians would not ask for a neuroimaging study. The widely unavailability of MRI scans in less developed centers reinforces this conduct. On the other hand, in more developed non HTLV-I endemic areas the opposite might be true, being neuroimaging the first diagnostic tool to be used. The present case clearly demonstrates that serology and neuroimaging should be always used together.

We conclude that, specially in places where HTLV-I is endemic, every patient with a spastic paraparesis and a clinical and radiological picture suggestive of a structural spinal cord lesion should have a screening test for HTLV-I. The clinical picture must dictate the final direction of the diagnosis.

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