Case Reports in Neurology

Case Rep Neurol 2016;8:47–52

DOI: 10.1159/000443787
Published online: February 12, 2016

1662–680X/16/0081–0047$39.50/0
www.karger.com/crn

Spinal Cord Schistosomiasis: Two Different Outcomes

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Key Words
Spinal cord schistosomiasis · Saudi Arabia · Myelopathy

Abstract
Spinal cord schistosomiasis is difficult to diagnose in nonendemic areas. We report the clinical profile of 2 young Saudi males who presented with myelopathy. The first patient arrived at our hospital relatively late, i.e. 3 months following the presentation of initial symptoms, and had received both pulse steroid therapy and a plasma exchange. Praziquantel was administered late and the patient did not recover. The second case presented early, i.e. within around 8 weeks of initial symptoms. This patient received praziquantel without any kind of steroid and had a complete recovery. We concluded that prompt recognition and early treatment with praziquantel is crucial for a better outcome. The role of steroids in these cases still needs to be proven.

Introduction

Schistosomiasis is a parasitic infection caused by different species of schistosomal parasites. It includes *Schistosoma haematobium* (endemic in Africa and Mediterranean countries), *S. mansoni* (endemic in Africa and the Middle East) and *S. japonicum* (endemic in Japan and China). It is estimated that 200 million people are infected with this parasite worldwide [1]. The infection occurs following contact with fresh water containing snails, which act as intermediate hosts, harboring the eggs of parasites [1].

Schistosomal infection can affect any part of the body including the bladder, intestine, skin, liver, brain, and spinal cord. The incubation period usually lasts from weeks to months. Schistosoma parasite eggs reach the spinal cord through the Batson plexus that connects the pelvic plexus to the spinal cord [2].

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Neuroschistosomiasis refers to a schistosomal infection of the brain or spinal cord. It is considered an underestimated cause of myelopathy, especially in nonendemic areas. If the infection becomes symptomatic and involves the central nervous system, it may cause disabling disease if not treated promptly [2].

We are presenting 2 cases of myelopathy caused by schistosomal infection with totally different clinical outcomes.

Case Presentation

Case 1

The patient was a 24-year-old male from the southern part of Saudi Arabia. He presented with a 3-month history of acute-onset, progressive-course severe lower back pain. Prior to this, he was healthy. The lower back pain was followed by progressive lower limb weakness, urinary incontinence, and a loss of sensation below the umbilicus. The weakness was severe enough to make him wheelchair bound. The patient gave a remote history of swimming in contaminated wells (around 8 months before the onset of his symptoms). He had initially gone to another hospital where he was diagnosed with transverse myelitis and was given pulse steroid therapy. He improved initially for 2 weeks but then worsened.

The patient had no history of fever, blurred vision, loss of consciousness, or seizure; no associated history of trauma, use of blood thinners, or illicit drugs; no history of exposure to a tuberculosis (TB) patient or ingestion of raw milk; no history of traveling or sexual abuse, and no history suggestive of an autoimmune disease or malignancy.

Upon examination, the patient presented as vitally stable, conscious, and alert. He had normal higher mental function and cranial nerves, and a visual field examination showed no meningeal signs. Examination of both upper limbs was normal. On examination of his lower limbs, the legs showed normal bulk and were not atrophied; there was increased tone, power was 0/5 bilaterally, he was hyperreflexic, had clonus, and exhibited an increasing plantar response.

The initial MRI (fig. 1) showed an extensive lesion extending from T_6 through T_9 with edema. The workup of his acute myelopathy was completed. Cerebrospinal fluid (CSF) analysis showed high protein of 1.08 g/dl. His cell count and glucose were normal, and he was negative for infection and cytology. Autoimmune, paraneoplastic, and serological testing of HIV, cytomegalovirus, and Epstein-Barr virus were negative. His Schistosoma titer was high at a ratio of 1:64. A spinal cord biopsy was completed and confirmed the diagnosis of spinal cord schistosomiasis.

The patient had been treated earlier (before the biopsy) with a plasma exchange, an extended course of intravenous dexamethasone, and had been enrolled in an extensive rehabilitation program. The patient had received praziquantel and continued on steroids. His rehabilitation was without significant improvement. He was assessed 11 months later and was found to have improved sensory symptoms, but he remained wheelchair bound. He was able to make minimal movements with both lower limbs but not against gravity. The patient exhibited urinary incontinence, yet he had no erectile dysfunction.

Case 2

A previously healthy, 29-year-old Saudi male presented with a 2-month history of bilateral lower limb weakness and urinary incontinence. The weakness gradually progressed with time. He also had pain in his right thigh accompanying the weakness. He had no history of other sensory symptoms, nor of a trauma or a fall. There was no history of fever or similar

illness in the past and no weakness in the upper limbs accompanied his symptoms, and there was no history of headache or loss of consciousness. Though he did travel to Egypt, he had no history of contact with TB in the past, nor did he have a history of cough, loss of weight, or night sweating. Upon examination, he was vitally stable and afebrile. The patient's higher mental function and cranial examination were completely normal, upper limb motor and sensory examination was also normal, and lower limb examination showed no atrophy and normal tone. The power was 4/5 with bilateral hyperreflexia and plantar response was equivocal. He could walk independently, but with difficulty due to the pain.

The patient's erythrocyte sedimentation rate was normal. His white blood cell count was 4.9 with no eosinophilia. His hemoglobin was measured at 14.6 g/dl, the liver function test was normal as were the CT results of his abdomen and pelvis. The CSF analysis showed a white blood cell count of 157 (mainly mononuclear cells at 98% and polymorphonuclear leukocytes at 2%) and a red blood cell count of 3. His protein and glucose levels were at 0.79 (normal: 0.15–0.45) and 3.7 (normal: 2.2–3.9), respectively. The oligoclonal band in the CSF was positive in this patient, and his Schistosoma antibody titer was very high on 2 occasions: 512 and 4,096 (normal <16 titers). His results for brucellosis, HIV, leishmaniasis, amebiasis, echinococcosis, and cytomegalovirus serology were all negative; furthermore, a rectal snip biopsy was negative for ova, and no schistosomal ova were detected in the patient's urine or stool. An MRI of the patient's brain showed normal results and an MRI of the whole spine presented high signals in the conus medullaris (fig. 2).

The patient was given praziquantel and no steroids. He was seen in the clinic 6 months later with complete recovery of motor functions and exhibited no more pain, although he did, however, still have mild urinary symptoms. We also contacted him recently (5 years after the event), and he is completely independent and plays football with no difficulty. He is married and has a 2-year-old daughter.

Discussion

Neuroschistosomiasis is considered an uncommon disease worldwide [3]. This is due, in part, to the fact that it is not usually symptomatic and if it is, it may be difficult to diagnose based on the clinical background or serological testing. Therefore, a low threshold for diagnosing myeloschistosomiasis should be attained during the assessment of nontraumatic back pain of patients in their adolescence or early adulthood presenting with transverse myelitis symptoms and signs, particularly if there is a history of traveling to endemic areas [1].

Myeloschistosomiasis presents with nontraumatic lower back pain that usually has a predilection for the lower spine, involving especially the lower thoracic (T_6), like in our case, to the upper lumber spinal cord segments. It is generally associated with sensory level, bladder, and bowel dysfunction. The onset of this presentation varies from a few months to a year after exposure to contaminated water. Neuroschistosomiasis can present up to 3 years after exposure [4]. The first patient did swim in a contaminated well around 8 months before the start of the symptoms. We believe that this could be the source of infection. Patients can also present with a prolonged onset of intermittent back pain until the diagnosis [5]. Spinal cord schistosomiasis can mimic spinal cord tumor presentation and necessitate biopsy as in our case and as reported in the literature [6].

Diagnosis of myeloschistosomiasis is a challenging process. During evaluation, stool ova and parasites should be tested. However, it has a low sensitivity profile. Rectal biopsy can detect the Schistosoma eggs only in 42% of cases, while stool examination for the eggs is

successful in 57% [1]. A CSF analysis will usually show pleocytosis and high protein content [1].

Another diagnostic modality is an MRI, which usually shows intramedullary expansion signals. The definitive diagnosis requires a spinal cord biopsy to show egg granuloma in the specimen [2].

Treatment for myeloschistosomiasis includes the anthelmintic praziquantel 40–60 mg/kg in divided doses for 3 days. It is recommended to be preceded by a steroid therapy in order to avoid the immune response that usually occurs during starting praziquantel [1–3]. The earlier the steroid dose, the better the associated outcome. The usual dose to administer is methylprednisone 0.5 g for 3–5 days followed by prednisone 1 mg/kg for a total duration of 3–6 months. Unfortunately, our first case did not improve whatsoever with steroids or praziquantel. This is probably due to the delayed diagnosis and the use of steroids 1 month before the correct diagnosis was made.

Most reported cases showed improvement after commencing therapy promptly. In a large cohort of 63 cases, a favorable outcome was present in 38 (60.3%), partial recovery with functional limitation in 16 (25.4%), and an absence of recovery in 9 (14.3%) [3].

In conclusion, spinal cord schistosomiasis can cause significant disability if not treated promptly with praziquantel or, especially, if treated with steroids alone in the beginning. Although the use of steroids is highly recommended in the literature, our second case had complete recovery without steroids. The use of steroids is still unproven and will be better judged by well-constructed randomized controlled trials.

Statement of Ethics

The authors have no ethical conflict to disclose.

Disclosure Statement

The authors would like to confirm that there is no conflict of interest, and no funding was received for this report.

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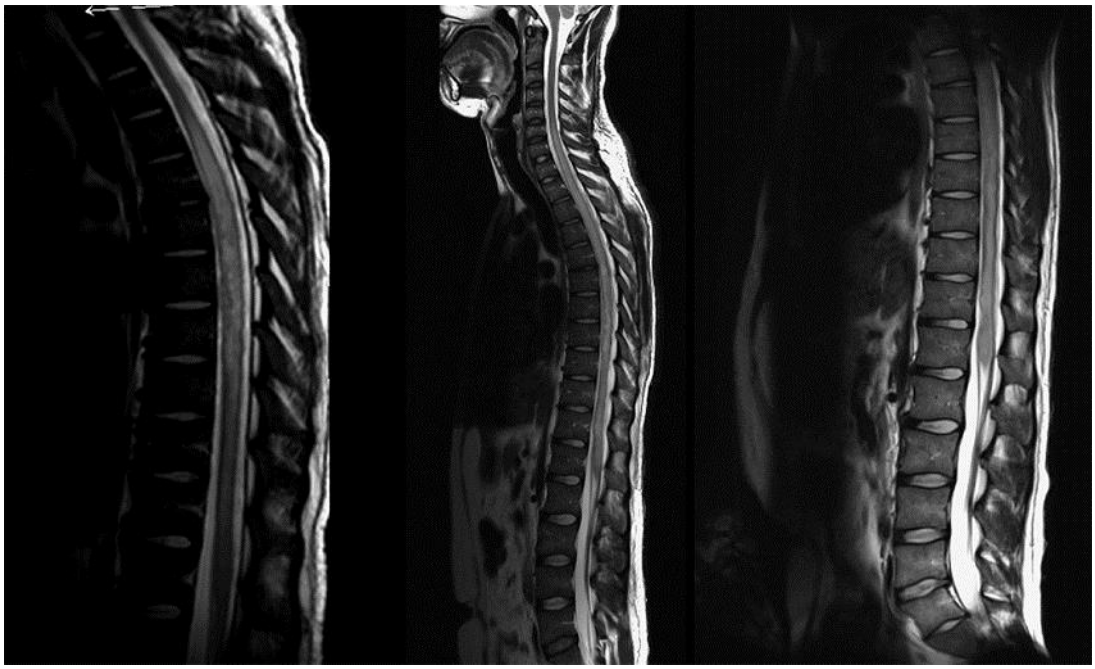

Fig. 1. Sagittal T2 MRI of the spinal cord shows a long-segment intramedullary high signal intensity involving most of the thoracolumbar spinal cord with minimal cord expansion (case 1).

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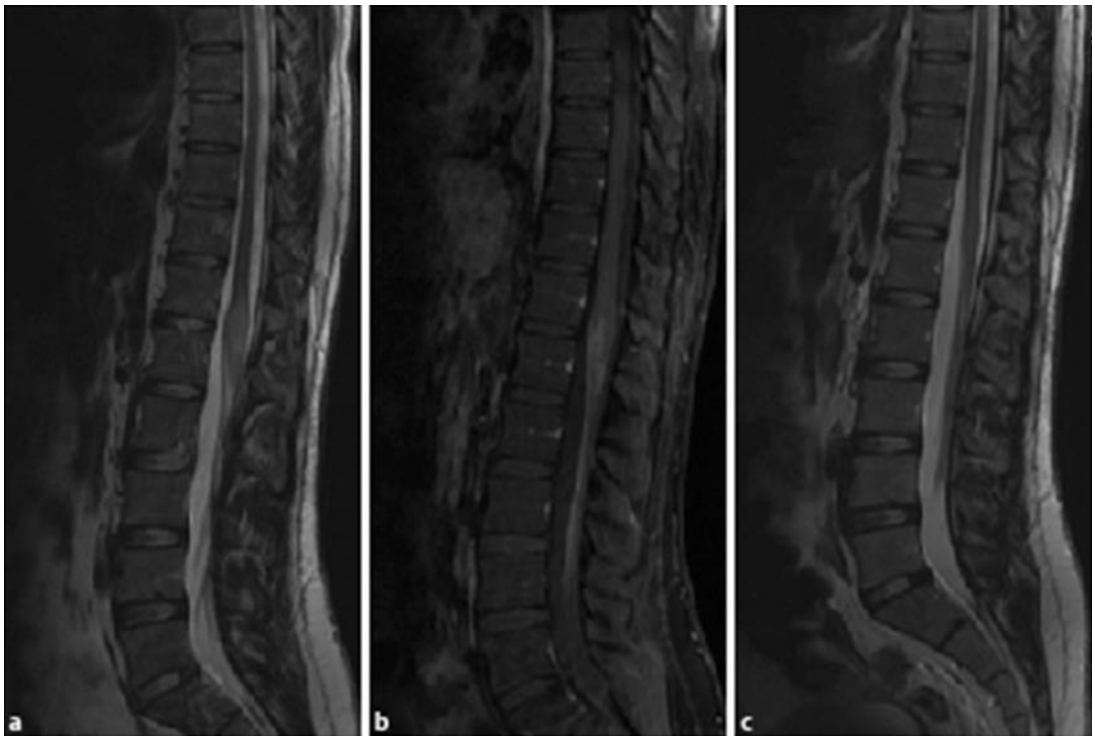

Fig. 2. **a** T2 sagittal MRI of the thoracolumbar spine at the time of admission demonstrates swelling of the conus medullaris which enhances heterogeneously after contrast (**b**). **c** T2 sagittal MRI of the spine of the same patient after treatment, showing marked interval reduction in the size of the swelling, denoting treatment response (case 2).