

Schistosomal Cervical Myelopathy

Mielopatia esquistossomótica cervical

Ronald Farias¹ Kléver Forte de Oliveira¹ George de Albuquerque Cavalcanti Mendes¹
Ussânio Mororó Meira¹

¹ Center of Medical Science, Universidade Federal da Paraíba,
João Pessoa, PB, Brazil

Address for correspondence Ronald Farias, MSc, Centro de Ciências
Médicas, Universidade Federal da Paraíba, João Pessoa, PB, Brazil
(e-mail: ronalfarias@uol.com.br).

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Abstract

Schistosomiasis is an infectious disease caused by trematode platyhelminths of the genus *Schistosoma*. The involvement of the cervical spinal cord is rare, with few cases reported in the literature. The management of such patients is particularly challenging, since clinical and radiological findings may be confounded with other inflammatory diseases and/or spinal cord tumors. We describe a 20-year old male with a history of swimming outdoors. He first presented pain in the back of the neck extending to shoulders and upper limbs paresis associated with four limbs hyperreflexia. The magnetic resonance imaging (MRI) showed a hypointense T1-weighted lesion in the cervical spinal cord, which was hyperintense on T2 images. The serologic testing was negative for schistosomiasis. A cervical cord biopsy at the C5-C6 level showed *Schistosoma* eggs in the histopathological examination. The treatment was performed using a single dose of praziquantel 50 mg/kg, with prednisone 40 mg/day for 3 weeks. On the follow-up, 1 year later, the patient presented mild reduction of the vibratory sensitivity in the distal third of both legs. Our illustrative case strengthens that, in endemic regions, *Schistosoma mansoni* infestation should be included in the differential diagnosis of intramedullary expansive lesions.

Keywords

- ▶ schistosoma mansoni
- ▶ schistosomiasis
- ▶ neuroschistosomiasis
- ▶ cervical cord

Resumo

A esquistossomose é uma doença infecciosa causada por platelmintos trematódeos do gênero *Schistosoma*. O acometimento da medula espinhal cervical é raro, com poucos casos apresentados na literatura. O manejo desses pacientes é particularmente difícil, uma vez que os achados clínicos e radiológicos podem ser confundidos com outras doenças inflamatórias e/ou tumores da medula espinhal. Descrevemos um homem de 20 anos de idade com história de natação ao ar livre. Primeiramente, ele apresentou cervicálgia que se estendeu até os ombros e paresia dos membros superiores, associada à hiperreflexia de quatro membros. A ressonância magnética (RM) mostrou lesão hipointensa em T1 na medula espinhal cervical, a qual foi hiperintensa nas imagens em T2. O teste sorológico foi negativo para esquistossomose. Uma biópsia da medula cervical ao nível C5-C6 evidenciou ovos de *Schistosoma* no exame histopatológico. O tratamento foi realizado com dose única de praziquantel 50 mg/kg, com prednisona 40 mg/dia por 3 semanas. No seguimento de 1 ano, o paciente apresentou discreta redução da sensibilidade vibratória no terço distal de ambas as pernas. Nosso caso ilustrativo reforça que, em regiões endêmicas, a infestação pelo *Schistosoma mansoni* deve ser incluída no diagnóstico diferencial de lesões expansivas intramedulares.

Palavras-Chave

- ▶ schistosoma mansoni
- ▶ esquistossomose
- ▶ neuroesquistossomose
- ▶ medula cervical

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Introduction

Schistosomiasis is an infectious disease caused by trematode platyhelminths of the genus *Schistosoma*. Among the species of this genus, the most important for humans are *Schistosoma mansoni*, *Schistosoma haematobium* e *Schistosoma japonicum*.¹ It is estimated that at least 200 million people are affected by it worldwide, and 200 thousand deaths due to this condition are registered annually, being endemic in Africa, South America and most of Asia.¹⁻³ *S. mansoni* is the only species existing in Brazil. Around 26 million Brazilians are at risk of contracting it.⁴

The typical manifestations of schistosomiasis include the urogenital, gastrointestinal and hepatolienal systems. The central nervous system involvement by *S. mansoni* is an atypical presentation, occurring by anomalous migration of the schistosome or unusual egg deposition from the portal venous system, through the venous plexus of Batson.^{3,4} This explains the higher prevalence of conus medullaris and thoracolumbar medulla envelopment. The involvement of the cervical spinal cord is rare, with few cases being reported in literature. The management of such patients is particularly challenging, since clinical and radiological findings may be confounded with other inflammatory diseases and/ or spinal cord tumors.³

Case Report

A male patient, 20 years old, white, with previous history of swimming outdoors, presented pain in the back of the neck extending to the shoulders. The neurological examination showed upper limbs paresis 4/5 associated with 4 limbs hyperreflexia. Sensory changes were not found. The magnetic resonance imaging (MRI) showed a hypointense T1-weighted lesion in cervical spinal cord, which was hyperintense on T2 images (→ Fig. 1). The biochemical and cytological evaluation of the cerebrospinal fluid (CSF) showed no alterations. Indirect immunofluorescence of CSF for schistosomiasis was negative.

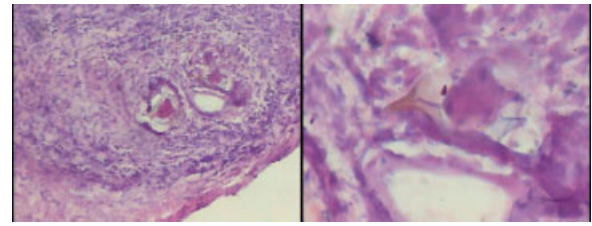


Fig. 2 Histopathological examination of the cervical spinal cord evidencing fibroconjunctive stroma containing remains of *Schistosoma mansoni* eggs (birefringent material) in the middle of a granulomatous-like inflammatory infiltrate, compatible with the diagnosis of schistosomiasis.

The laboratory tests revealed mild leukocytosis (11,900/mm³) associated with moderate eosinophilia (1,035/mm³). Feces samples (Kato-Katz method) showed no *Schistosoma* eggs.

A cervical cord biopsy at the C5-C6 level showed a chronic granulomatous inflammatory process with multinucleated giant cells and birefringent material, suggestive of *S. mansoni* eggs (→ Fig. 2).

The treatment was performed using a single dose of praziquantel 50 mg/kg, with prednisone 40 mg/day for 3 weeks, performing a gradual withdrawal.

At the one-year postoperative follow-up, the patient presented mild reduction of the vibratory sensitivity in the distal third of both legs. However, he preserved reflexes in the four limbs, with normalization of muscular strength in the upper limbs.

Discussion

Vascular obstruction occurs both through venous migration of the eggs or direct deposition of these by the parasite in the perimedullary plexus, with formation of granulomas and intense local inflammatory reaction, followed by necrotizing myelitis. There are two clinical forms of schistosomal involvement of the spinal cord: granuloma formation and transverse

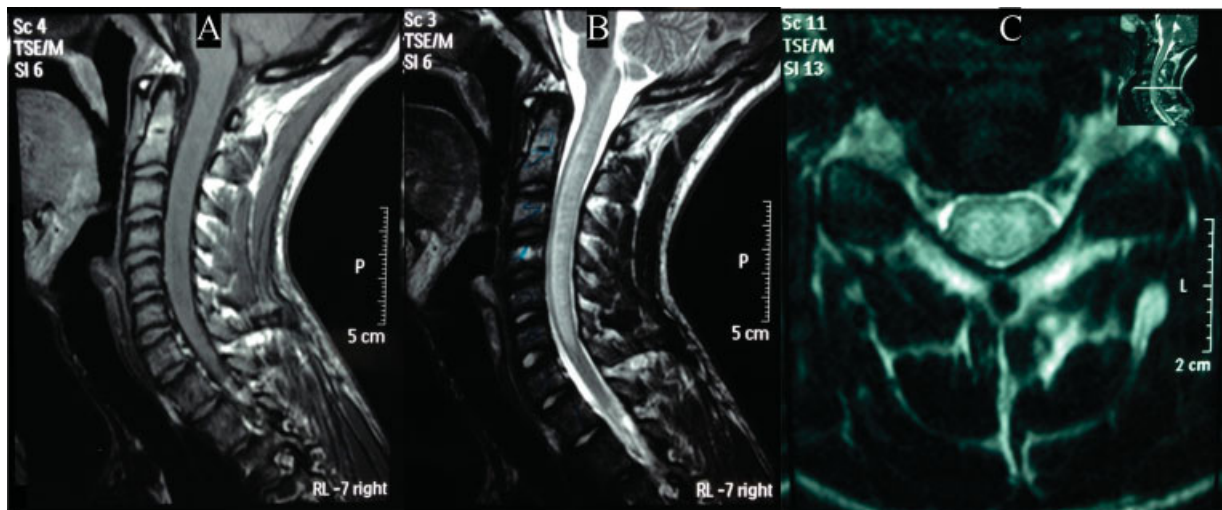


Fig. 1 Preoperative magnetic resonance imaging (MRI). (A) T1-weighted sagittal view, showing hypointense signal in the C4-C6 spinal segments; (B) in the T2-weighted sagittal view, hyperintense signal extending through almost the entire cervical spinal cord. (C) T2 axial view shows hyperintense signal in the cervical spine at the C4-C5 transition level.

Table 1 Cases of schistosomal myelopathy reported in the literature

Case	Clinical Manifestations	Treatment	Outcome
Junker et al ³ (2001)	Pain in the four limbs; tetraparesis associated with the occurrence of clonus in the legs and hypoesthesia under the T4 level on the left, and the T8 level on the right	Decompressive laminectomy, followed by praziquantel, high doses of corticoid and physiotherapy	Distal spastic tetraparesis in the legs and hypoesthesia from the C6 level down
Silva et al ⁵ (2002)	Neck pain, tetraparesis, paresthesia in the four limbs and hypoesthesia at the C5 level	Oxamniquine (15 mg/kg in a single dose) and prednisone 60 mg/day for 2 months	Muscle strength improvement and hypoesthesia resolution; persistence of left hemi-body paresthesia
Laureys et al ² (2007)	Pain, tactile hypoesthesia, hypoalgesia, hypopalesthesia and decreased distal proprioception in the legs, associated with flexion and extension paresis (strength grade 4); mild gait instability	Praziquantel (60 mg/kg/day for 3 days) and methylprednisolone (64 mg/day)	Resolution of pain and sensory symptoms, as well as marked motor improvement
Our case	Paresis of the upper limbs (strength grade 4) associated with hyperreflexia in the 4 limbs	Single dose of Praziquantel (50 mg/kg) associated with prednisone (40 mg/day for 3 weeks, performing a gradual withdrawal)	Hypopalesthesia in the distal third of both legs; preserved reflexes in the 4 limbs, with normalization of muscular strength (grade 5) in the upper limbs

myelitis.³ The schistosomal myelopathy (SM) prevalence is unknown. However, strong evidences indicate it is an underdiagnosed condition.^{1,4} The typical patient of this disease is young, male, presenting low back or lower limbs pain, usually of radicular pattern, followed by weakness and progressive sensorial loss in the lower limbs, associated with autonomic dysfunction, mainly of the bladder.¹

The diagnosis is primarily based on clinical examination. The MRI usually presents hyperintense T2-weighted lesions, which are isointense on T1 images.² The laboratory tests of CSF reveal nonspecific findings.^{1,4} Our patient presented normal protein and glucose levels; pleocytosis was not evidenced.

The SM treatment is based on the association of an anthelmintic (praziquantel or oxamniquine) and a corticosteroid. In cases of granuloma formation, a surgical approach with decompression is indicated.^{1,2}

Reports of cervical spinal cord schistosomiasis have been previously published. Junker et al³ and Laureys et al² reported cases of neuroschistosomiasis presenting as transverse myelitis in men in their fourth decade. A decompressive laminectomy was performed in the first case due to acute incomplete tetraparesis and evidence of spinal stenosis at MRI. Silva et al⁵ described a case of granuloma formation case in a 34 years old male. In the three cases, the patients presented sensory and motor impairment, in addition to pain. The proposed pharmacological treatment was similar to our case (anthelmintic and corticosteroid), as well as the clinical outcome (sensory and motor improvement and pain resolution) (► **Table 1**).

Conclusion

Cervical myelopathy is an extremely rare presentation of Schistosomiasis. Our illustrative case strengthens that, in

endemic regions, *S. mansoni* infestation should be included in the differential diagnosis of intramedullary expansive lesions. The diagnosis is based on clinical and radiological findings, and pharmacologic therapy is the primary treatment approach. Surgical decompression and biopsy are reserved to patients in whom no clinical improvement is observed after institution of antischistosomal medications and/or in case of neurological deterioration. Only in such cases, surgery should be indicated. The SM prognosis is excellent when the diagnosis is early, and the treatment is appropriate.^{1,3}

Conflicts of Interest

The authors declare that there are no conflicts of interest.

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