

Schistosomal Myeloradiculopathy: case report in a pediatric patient

Lívia Helena Gomes de Barros Brandão¹ , Camila Neves de Melo Cavalcanti¹ , Laércio Pol Fachin¹ ,
Adriana Ávila Moura² , Ana Carolina de Carvalho Ruela Pires¹ ,

ABSTRACT

Schistosomiasis is a parasitic endemic typical of the Americas, Asia and Africa. Schistosomal Myeloradiculopathy is a severe evolution of schistosomiasis infection and, although very common, the prevalence in endemic areas has been underestimated. Objective: to report Schistosomal Myeloradiculopathy case in a pediatric patient. Methodology: descriptive study of the type Case Report retrospective, submitted and approved by the Research Ethics Committee of the CESMAC University Center, CAAE: 28835220.0.0000.0039, Opinion N.º: 3.898.292. Case report: a previously healthy 11-year-old boy, started with a history of acute pain in lower limbs that worsened during the night accompanied of fever. Evolved with low back pain, dysuria, oliguria, subsequent anuria, vesical globe formation and lower limbs paresis. The investigation resulted in positive stool examination for schistosomiasis and magnetic resonance imaging of lumbosacral spine that corroborated the diagnostic hypothesis. The treatment included Albendazol, Praziquantel and pulsetherapy with Methylprednisolone during hospitalization. The patient was discharged from the hospital with improved neurological status, using prednisone 40 mg/day. Conclusion: Schistosomal Myeloradiculopathy is the most severe form of the ectopic manifestations of schistosomiasis. The difficulty in recognizing the clinical condition and the limitation of access to complementary diagnostic methods contributes to the underdiagnosis of the disease, causing severe sequels for patients with disease and hiding its epidemiological importance, especially in pediatric and young patients.

Keywords: Schistosomal Myelopathy, Neuroschistosomiasis, Ectopic Schistosomiasis, Neglected Diseases, Pediatrics.

1. Centro Universitário CESMAC, Maceió (AL), Brasil.

2. Universidade Federal de Alagoas (UFAL), Maceió (AL), Brasil.



INTRODUCTION

Schistosomiasis is a typical parasitic endemic in the Americas, Africa and the eastern Mediterranean. It arrived in Brazil with African slaves brought by the Portuguese colony and, although the slaves were infected by two species of schistosome, *Schistosoma mansoni* and *Schistosoma haematobium*, only the first one developed in Brazil.¹

The World Health Organization (WHO) estimates that schistosomiasis affects 200 million people in 54 countries. In Brazil, it is believed that there are about 12 million infected people, mainly in the states of Northeast and Southeast regions, being considered an endemic condition and affecting all of population's life stages.^{2,3,4}

The spinal cord is the main ectopic focus of *S. mansoni* implantation and represents a severe evolution of the schistosomal infection, whether symptomatic or not. Schistosomal Myeloradiculopathy (SMR) is highly suggestive in individuals with an epidemiological history of the infection.⁴

SMR is an underreported disease, with an estimated prevalence between 1% and 5% among all cases of diagnosed schistosomiasis and has an underestimated morbidity. In Brazil, it is believed that spinal cord injuries prevalence due to schistosomiasis is around 5%.⁴ The difficulty in recognizing its clinical picture and the limited access to complementary diagnostic methods contribute to SRM underdiagnosis, causing serious sequelae for the disease carriers and hiding its epidemiological importance, especially in pediatric and young patients.⁵

The following case report aims to describe a case of SMR that occurred in a pediatric patient in the scope of Brazilian Unified Health System (Sistema Único de Saúde, SUS), during Schistosomiasis endemic in the state of Alagoas. It is a public health problem that can lead to disabling complications, and proper management is essential for a good prognosis, avoiding disabilities that can extend into adulthood.

CASE REPORT

Patient, male, 11 years old, 34 kg, previously healthy, born and resident in Barra de Santo Antônio,

Alagoas, admitted in 2017 to the HGE pediatric sector, complaining of pain in the lower limbs that worsened at night, accompanied by not measured fever. Reports improvement with massage and oral dipyrone.

After a week, he developed severe low back pain, thus looking for a health service, where he was medicated with intravenous and oral dipyrone. After 01 day, he started dysuria and oliguria, being transferred to HGE where he remained for 20 days in pediatric hospitalization.

He denied comorbidities or previous surgeries, allergies, alcoholism or smoking. He presented a positive epidemiology for schistosomiasis and Chagas disease, as he reported bathing in rivers in an endemic region and having already lived in a mud house. His vaccination calendar was updated, according to maternal report.

On physical examination, asymmetry of motor strength in the lower limbs was evidenced: grade III strength in the right lower limb and grade II in the left lower limb, in addition to low back pain and difficulty walking. There was no evidence of change in strength in the upper limbs. During hospitalization, he evolved with anuria and bladder globe formation.

Complementary exams were performed, including lumbosacral spine MRI, cerebrospinal fluid collection (CSF), lumbar spine Computed Tomography (CT), contrast-enhanced abdominal CT, rapid tests for syphilis and HIV, in addition to parasitological examination of feces (PEF).

Rapid tests showed negative results for both investigated pathologies. The PEF sample had 15 *S. mansoni* eggs per gram of feces. Research for poliovirus was carried out in an isolated sample of the collected stools, with a negative result. Contrast-enhanced abdominal CT showed a mild and homogeneous enlargement of the liver parenchyma in the left lobe. Lumbosacral spine CT, performed on the 4th day of hospitalization, showed only a small disc protrusion between L4-L5, requiring MRI for diagnostic complementation.

Lumbosacral spine MRI, performed on the 11th day of hospitalization, showed extensive myeloradiculitis, with edematous appearance, affecting the spinal cord in the segment between C3 and the medullary cone, showing irregular uptake by the contrast medium, involving the anterior cords in the segment between T7 and the

medullary cone, associated with the impregnation of the roots of the cauda equina. The findings corroborate the hypothesis of spinal implantation of the schistosomiasis parasite (Figure 1).

By correlating the clinical and epidemiological data and the findings obtained in complementary exams, therapy was started with Albendazol for 05 days, Praziquantel 1800 mg, in a single dose,

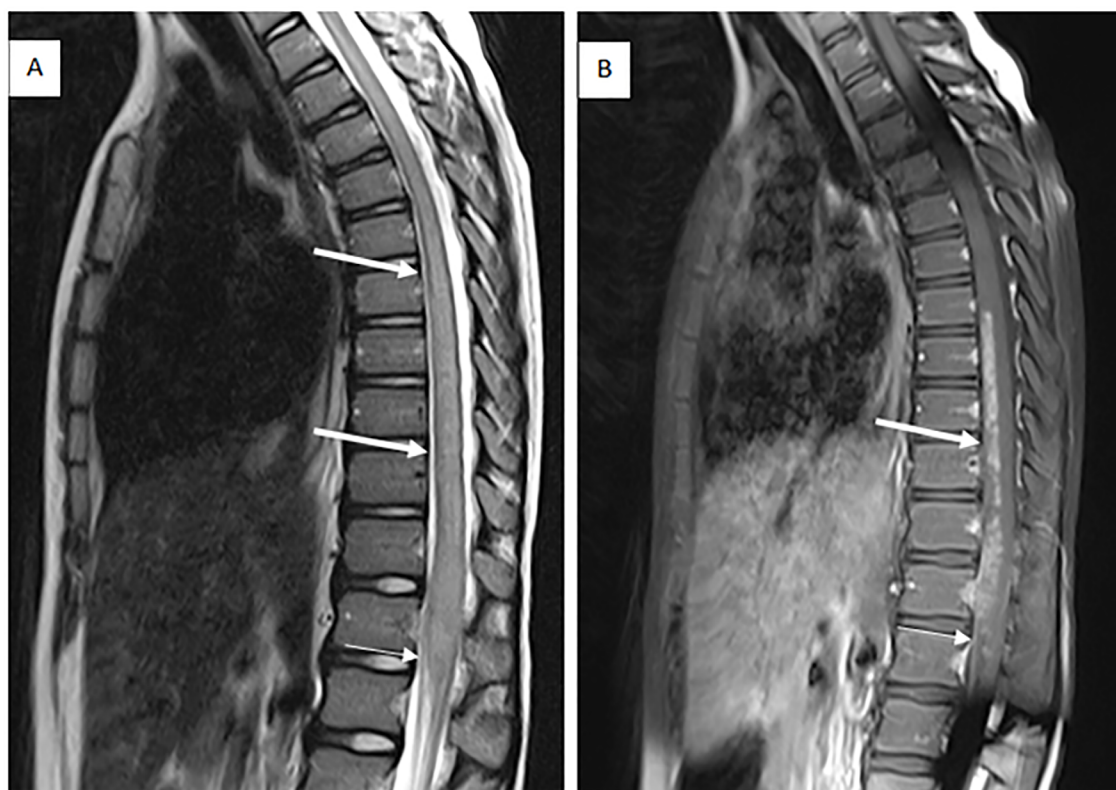


Figure 1: (A) Dorsal column magnetic resonance at T2-weighted shows diffuse spinal cord involvement, with hypersignal and spinal cord thickening, more evident in the cone. (B) T1-weighted magnetic resonance imaging of the dorsal column with fat suppression showing heterogeneous paramagnetic contrast uptake, predominantly in the anterior cords.

and pulse therapy with Methylprednisolone 30 mg/kg/day, for 05 days. The patient evolved with an improvement in his condition, being discharged from the hospital with prescription of corticosteroid therapy maintained with 40 mg/day of prednisone and forwarding to neurology and infectiology specialized services.

The feasibility of performing CSF was only possible on the last day of the patient's hospital stay, with unspecific cellularity and biochemistry results and no eosinophil findings. The neoplastic cell search was negative, as well as the bacteriology tests for Gram, China ink and blue methylene. Regarding to pathological investigation, the research showed non-reactive VDRL, in addition to the

research for cysticercosis IgG, toxoplasmosis IgG, cytomegalovirus IgG, herpes simplex IgG, Epstein-Barr IgG, which showed negative results. The investigation for HIV-1, HIV-2, HTLV-1 and HTLV-2 also came up with non-reactive results.

During outpatient follow-up, about 01 month after the initial condition, the patient was still using prednisone, but at a dose of 20 mg/day and showed an unaltered neurological physical examination, grade V strength in both lower and upper limbs, adequate tone and trophism, no pain or urinary complaints. After new PEF and CSF results, both within the normal range, the patient was released from follow-up at an infectiology clinic.

DISCUSSION

Schistosomiasis is a serious and highly prevalent parasitic disease among worldwide diseases within this classification. It is predominant in tropical and subtropical areas, especially in houses with precarious conditions of basic sanitation.⁶ In the Americas, Brazil is the country with the highest number of cases, reaching endemic rates in states such as Pernambuco, Alagoas, Bahia and Sergipe.⁷

In Alagoas, according to data published by the Ministry of Health (MS), schistosomiasis would have a positivity rate of 8.54%. A study carried out between 2010 and 2014 showed that among 102 surveyed counties in Alagoas, 70 had reported cases of schistosomiasis. Prevalence greater than 15% was observed in seven counties, being considered by the MS as areas of high endemicity. The largest proportion of those infected was made up of individuals between 15 and 49 years old (58.63%).⁷

The central nervous system (CNS) can be affected by *S. mansoni* infections, with SMR being the most common form of the disease in the nervous system, with a preference for males. Although frequent, the prevalence of SMR in endemic areas is not known, as its diagnosis is neglected.⁵

It is believed that eggs and worms can move, even during the acute or chronic intestinal phase of verminosis, reaching Baston's vertebral venous plexus, which makes the connection between the portal system and vena cava to spinal canal veins, a fact which justifies the higher incidence of myelopathy in the lumbosacral region.⁵ In addition to oviposition or embolization, there is an intense inflammatory reaction to foreign bodies present in the nervous system, which can vary among infected individuals from minimal reactions, without clinical expression, to granulomas or expansive masses.^{8,9}

In the prodromal phase, SMR is frequently manifested as a triad of symptoms, characterized by low back pain (97.5% of cases), changes in lower limb sensitivity (97.5%) and urinary dysfunction (96.2%). There is not always clinical evidence for systemic schistosomiasis existence. In general, patients will present an acute or subacute condition of lower medullary involvement. Typically, low back pain will be part of the condition and precedes the appearance of other neurological symptoms by hours

to 03 weeks.¹⁰ In the described case, the patient showed two of the main prodromal symptoms manifested by infected individuals.

There is a progressive and accumulative worsening of signs and symptoms, with the complete neurological picture usually taking place within 15 days.¹⁰ The isolated analysis of symptoms is nonspecific and makes diagnosis difficult, since the clinical picture is also characteristic of other pathologies, including tertiary syphilis disease, spinal cord abscesses, tuberculosis, B12 deficiency myelopathy, neurocysticercosis, HIV and HBV myelitis, among others.⁸

In the case of schistosomiasis, diagnostic methods can be direct or indirect; the direct tests can identify the parasite or parts of it, while the indirect tests depend on biochemical or immunological markers for proof. The MS recommends the direct investigation of *S. mansoni* eggs in the feces, using the Kato-Katz technique, which configures a simple, quantitative, and highly sensitive way for diagnostic investigation.¹¹

Regarding indirect methods, immunological tests are the most used, but limited since they do not define the intensity of infection and can remain positive even after established treatment. Among the indirect methods, the immunoenzymatic technique (ELISA) is economically useful and is currently the most used method, even though it has lower predictive value and sensitivity when compared to indirect immunofluorescence.⁹

The diagnosis of SMR is in most cases presumptive, and based on clinical and epidemiological findings, as well as in pathogen detection by microscopic or serological techniques. It is crucial to exclude differential diagnoses. Histological study by medullary laminectomy with finding of eggs in the nervous tissue represents the only confirmation of *S. mansoni* medullary involvement, but it is an invasive and risky procedure. The possibility of using complementary methods that include CSF reactions or spinal cord radiological imaging has been avoiding routine biopsies.⁵

A case series study of seven SMR pediatric patients in Brazil evaluated, through the PEF, the presence of worm eggs, which was confirmed in five of the investigated patients. The presence of eggs corroborates the hypothesis of SMR, but the absence

of the finding does not exclude the diagnosis. Rectal biopsy appears as a more sensitive method option for research and should be performed in the absence eggs in the PEF.¹²

The CSF analysis does not show a pattern, finding nonspecific alterations, such as lymphocytic pleocytosis with eosinophils, high protein level and low or normal glycemic levels. Obtaining positive serology for the parasite in CSF can be considered evidence of neurological infection. Thus, SMR can be presumed if pleocytosis findings with the presence of eosinophils and hyperproteinorrachia combined with positive CSF serology are present.^{5,10}

As for imaging exams, CT with myelo-CT protocol may reveal thickening with irregular spinal cord contours to the point of determining a reduction in the intraspinal cerebrospinal fluid space with or without spinal canal block, in addition to thickening of the nerve radicles of the cauda equina.⁵

MRI stands out, showing alterations in all the cases in which it was used, being a highly sensitive method, with thickening of the medulla, as well as of the nerve radicles of the tail and corner, alteration of the medullary intrasubstantial signal in the T2 weighting, mainly in the cone, and heterogeneous uptake by paramagnetic contrast, usually assuming a granular pattern. With the instituted treatment, as the patient's condition improves, the MRI images also return to the normal pattern.⁵

Recognition of SMR is important so that therapy can be started early, as the acute and chronic forms can seriously compromise affected people. Treatment includes antiparasitic drugs, mainly Praziquantel. According to the SMR manual of MS, the praziquantel treatment recommended for children is given with 60mg/kg in two doses, in association with prolonged corticosteroid therapy. In specific cases of clinical worsening even when using conservative treatment or acute paraplegia, surgery may be indicated.^{5,11}

Evidence in the literature affirms that corticosteroid therapy should be instituted for more than 2 months, as if it is discontinued early, there is an increased risk of recurrence.^{5,11,13} The treatment observed in literature is consistent with that found in the above-mentioned case, with continued corticosteroid therapy for the patient on

an outpatient basis, in weaning, without recurrence of symptoms.

The implementation of SMR epidemiological surveillance throughout the country is justified, even in non-endemic states, since the movement of people in the national territory is free, and there are states with high levels of endemicity of the disease. The recognition and early institution of treatment play a fundamental role in the prevention of serious and irreversible injuries, as well as in the recovery of affected people, in general young people in full production.

CONCLUSION

The schistosomiasis endemic in Brazil, and particularly in the state of Alagoas, is a public health problem. SMR represents a severe ectopic form since the CNS involvement can lead to disabling complications. The difficulty in recognizing the clinical picture and the limited access to complementary diagnostic methods contribute to the disease underdiagnosis, hiding its epidemiological importance.

Considering the case description in a pediatric patient with SMR in the scope of SUS, it is known that an early approach can determine the child's life quality, with adequate management being essential for a good patient prognosis, and for the prevention of permanent disabilities.

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Corresponding Author:

Lívia Helena Gomes de Barros Brandão
livinha_brandao@hotmail.com

Editor:

Prof. Dr. Felipe Villela Gomes

Received in: jul 24, 2020

Approved in: oct 26, 2020
