Rare disease

Myeloradiculopathy associated to *Schistosoma mansoni*

Patrícia Pita Lobo,¹ Miguel Coelho,¹ Ruth Geraldles,¹ Carolina Santos,² Maria Grácio,³ Mário Miguel Rosa,¹ João Lobo Antunes⁴

¹Neurology Department, Hospital de Santa Maria, Lisbon, Portugal;

²Neurological Imaging Department, Hospital de Santa Maria, Lisbon, Portugal;

³Unit of Medical Parasitology and Microbiology, Instituto de Higiene e Medicina Tropical, Lisboa, Portugal;

⁴Neurosurgical Department, Hospital de Santa Maria, Lisbon, Portugal

Correspondence to Miss Patrícia Pita Lobo, anapa.pitalobo@gmail.com

Summary

Neuroschistosomiasis caused by *Schistosoma mansoni* (Sm) is a rare and severe condition potentially leading to permanent neurological deficit. An 18-year-old Brazilian female was admitted due to a severe conus medullaris and cauda equina syndrome. MRI of thoracic/lumbar spine showed an expanded conus medullaris with patchy gadolinium-enhancement, needle electromyography revealed acute bilateral radiculopathy (L5-S1-S2), cerebrospinal fluid (CSF) showed lymphocytosis and increased proteins and lesion surgical biopsy documented a lymphocyte infiltrate. Immunodiagnosis with cercariae hullen reaction using Sm cercariae in CSF and serum and immunoelectrodiffusion for circulating antigens detection using anti-Sm antibodies were positive. No schistosoma parasites were found. The patient was treated with praziquantel and corticotherapy for 6 months. At 1 month, partial clinical improvement was noticed, and MRI showed a normal size conus medullaris. At 6 months, there was complete clinical recovery. This case shows that a severe neurological deficit by Sm may have a clinical full recovery after treatment.

BACKGROUND

Schistosomiasis (or bilharziasis) is an endemic parasitic disease caused by Trematoda platyhelminthes of the genus schistosoma.¹ Its incubation period typically varies from weeks to years.¹ The reproduction of the parasite occurs in host snails that live in fresh water, and large numbers of larval schistosomes are then released to the infested water. Schistosomes enter the human body through the skin in contact with the infested surface water, and migrate through the bloodstream to reach the liver; after differentiation in the liver, they finally settle in the intestine or bladder. In rare instances, the parasite may reach the nervous system (neuroschistosomiasis NS), probably through

perispinal vascular network,² causing an exuberant host inflammatory response to the eggs laid in the nervous tissue.³ The WHO estimated that 200 million people are infected worldwide and that 650 million live in an endemic area.⁴ People living near fresh water are particularly at risk. The infection is widely distributed in Africa, South America and the Caribbean Islands, and is also endemic in some regions of Brazil such as Minas Gerais, where the most common species is *Schistosoma mansoni* (Sm). Nevertheless, population migration and travelling may lead to the spread of the disease to low or non-endemic areas.

NS is a severe disease whose prognosis largely depends on early diagnosis and treatment.² Usual manifestations

Figure 1 Thoracic and lumbar spine MRI sagittal imaging showed an expanded conus medullaris lesion, hyperintense in T2-weighted (A), light hyperintense in T1-weighted image (B), with patchy pathological gadolinium-enhancement (C). Thoracic and lumbar spine MRI sagittal imaging performed 1 month after corticosteroid therapy showed no conus medullaris expansion in T2-weighted (D) and in T1-weighted image (E) and less gadolinium-enhancement (F).

are due to low thoracic/upper lumbar spine involvement. The prevalence of schistosomal myeloradiculopathy is unknown, but it is probably under-diagnosed.⁵ We report a case of a myeloradiculopathy secondary to Sm in a Brazilian female living in Portugal.

CASE PRESENTATION

An 18-year-old Brazilian woman, born in Minas Gerais, who arrived to Portugal 2 years before the presentation of neurological symptoms. She was admitted to our hospital in February 2008, and her symptoms had begun 1 week before. She reported, in the following order, urinary retention, numbness in her legs, diminished muscle strength in the lower extremities and finally walk incapability and fecal incontinence 1 day before hospital admission. She reported no systemic symptoms, no weight loss (body mass index 24), no recent trauma or exposure to fresh water. Her medical history and family history were unremarkable for systemic or neurological diseases. She denied recent travel to endemic areas of schistosomiasis. General physical examination was normal, namely no fever or hepatosplenomegaly were observed. The neurological examination disclosed a distal flaccid paraparesis (crural/hip flexion, abduction and adduction strength scored four on the medical research council scale; leg flexion and extension strength scored two and foot dorsiflexion and extension strength scored zero, that unable ambulation; knee jerks were preserved but ankle jerks were weak, and plantar responses were absent. There was a saddle pinprick and touch hyposthesia and L4-L5-S1-S2 pinprick and touch hyposthesia bilaterally (no sensory dissociation), and the sense of vibration and position were diminished in the feet. During hospital stay, she developed severe dysesthesia and hyperesthesia of the lower limbs in the radicular territory described above, relieved by amitriptyline and carbamazepine. The haemogram was unremarkable. The cerebrospinal fluid (CSF) analysis disclosed lymphocytosis (174.4 cell/mm³), high protein content (2.96 g/l) and a positive Pandy reaction (+++). Immunophenotypic analysis of CSF cells showed eosinophils (4%) and T cell predominance. Oligoclonal bands were positive. CSF serology and/or PCR for tuberculosis, herpes simplex virus, cytomegalovirus, Epstein-Barr virus, Human T-lymphotropic virus-1 and syphilis were negative, as was HIV serology in blood. Angiotensin converting enzyme test was also negative. On admission, the thoracic and lumbar spine CT scan was normal. A MRI of thoracic and lumbar spine showed an expanded conus medullaris (figure 1A,B), with patchy abnormal gadolinium-enhancement (figure 1C). Needle electromyography study showed fibrillation and slow potentials of denervation at rest in both anterior tibialis muscles and long extensor of the left hallux, and there are some polyphasic units in attempts to contraction representing re-innervation. Motor conduction velocities in the left external popliteal sciatic nerve were decreased and absent in the pedal uptake in the right external popliteal sciatic nerve, potentials' amplitude was decreased in both posterior tibialis nerve, and the sensory action potentials were normal; these findings are consistent with acute axonal damage of roots L5-S1-S2 bilaterally. Additionally, cranial and cervical spine MRI, and thoracic CT scan were normal, considering the hypothesis of Chagas or lung schistosoma disease. To exclude an

intraspinal tumour of the conus, a surgical biopsy of the spinal lesion was performed, which showed a lymphocyte infiltrate without observed parasites or schistosoma eggs and no neoplastic cells. Subsequent immunodiagnosis with cercariae hullen reaction (CHR) was positive in CSF (figure 2A) and serum (figure 2B), using Sm cercariae as antigen. Immuno-electrodiffusion for circulating antigens detection using anti-Sm antibodies was also positive in serum (table 1). No schistosoma eggs were found in urine or stool samples. One day after admission, the patient was started on intravenous methylprednisolone (1 g/day, for 3 days), followed by dexametasone for 12 days. After diagnosis of schistosomal myeloradiculopathy by CHR, praziquantel (one single oral dose of 50 mg/kg), prednisolone (1 mg/kg/day, for 6 months) and physiotherapy were started. One month later, the patient significantly improved, reporting partial remission of sensory dysfunction and paraparesis (crural/hip flexion, extension, abduction and adduction strength scored 4+, leg flexion and extension strength scored 3; foot extension and dorsiflexion scored 2), was able to walk with help and recovered partially from sphincters dysfunction (moderate urinary retention and no fecal incontinence). The MRI performed 1 month after steroid therapy showed no conus medullaris expansion and diminished gadolinium-enhancement (figure 1D,E,F). We documented a decline in the positivity of CHR after the treatment with praziquantel (table 1). Six months later, she was able to walk independently, and had no neuropathic pain or sphincter dysfunction.

DISCUSSION

We report the case of a Brazilian patient with an acute (1-week duration) schistosomal myeloradiculopathy of the conus medullaris and cauda equina, who had a clinically full recovery after long duration treatment with corticosteroids (6 months) and praziquantel (a single dose). It was documented an almost complete recovery in spinal MRI too (figure 1D,E,F) and a reduction in the CHR positivity (table 1) 1 month after treatment. The diagnosis of NS is based on the presence of symptoms from low thoracic/upper lumbar spine involvement, evidence of schistosomal infection (either microscopic observation of the parasite or eggs, or by serologic techniques) and exclusion of other causes of transverse myelitis.⁶ In our case, the diagnosis of NS was based on a suggestive clinical picture, proper epidemiologic history, evidence of exposure to schistosoma by immunodiagnostic techniques, characteristic MRI findings and response to specific therapy.²⁻³ The response to praziquantel and steroids was documented in both clinical and imagiological terms. Furthermore, a decline in the positivity of CHR was observed after treatment with praziquantel.⁷⁻⁸ Actually, serologic diagnosis like CHR has been superseded by techniques such as ELISA. Despite this, the CHR and the immuno-electrodiffusion results were concordant to the diagnosis of a Sm infection. As our patient was of Brazilian origin, Sm was the expected species of schistosoma because *Schistosoma haematobium* is not found in Brazil. The availability of a spinal biopsy further excluded other causes for the clinical picture, such as a spinal tumour. The differential diagnosis with tumours is of paramount importance, as 10% of all intraspinal tumours affect the conus.⁹ The presence of inflammatory infiltrate in the spinal cord biopsy leads us to believe that the injury

Figure 2 In the cercariae hullen reaction (CHR) technique, the cercariae of *Schistosoma mansoni* are placed in the patient serum or cerebrospinal fluid (CSF) for 24 to 48 h. If there are serum or CSF antibodies to the parasite, they react with the cercariae, manifesting itself by cercariae swelling, and the surface becomes wrinkled and with bubbles, that could be located in different areas. Cercariae Hullen Reaction (CHR) positive in CSF (arrows show wrinkled surface) (A). Cercariae Hullen Reaction (CHR) positive in serum (arrows show bubbles in the anterior and tail end of cercariae) (B).

in this case is due to the exuberant inflammatory reaction to possible parasite antigens present in spinal cord, despite not having found parasites or parasite eggs (possibly due to the small size of biopsy). The need to perform a biopsy was raised because the MRI findings did not allow a precise differential diagnosis, and the possibility of a granulomatous / inflammatory response inside a swollen conus medullaris was considered. The presence of severe neuropathic pain and of an acute radicular damage documented by electromyography, supported a radicular involvement in our patient, pointing away from a pure conus medullaris lesion.⁷ Our patient presented thus with a myeloradicul form of NS, but other forms of spinal cord schistosomiasis are possible, such as myelitic (the most frequent) and granulomatous or pseudotumoural.² The typical patient with NS is a young male, with no other complaints of schistosomal infection, presenting with a flaccid paraplegia secondary to a conus medullaris or cauda equine lesion or a transverse myelitis,³ that in 70% of the cases unable ambulation.⁴ Although the onset of the disease may vary from sudden to progressive, it arises in most instances in a subacute or chronic manner, with progressive worsening of the deficits.⁵ Our case is exceptional in that, the clinical progression was acute of 1-week duration. Moreno-Carvalho *et al*,¹⁰ suggested that a patient is considered to be at high risk for schistosomiasis when the CSF count shows white blood cells $> 4/\text{mm}^3$, protein $> 40/\text{mm}^3$ and an increase in the eosinophilic granulocytes, as was the case in our patient. The finding of oligoclonal bands, consistent with intrathecal synthesis of antibodies, also supported the diagnosis.^{11 12} Available data clearly show that the prognosis of NS depends on factors

Table 1 Immunodiagnostic tests results before and after treatment.

Immunodiagnostic Tests	
Immunodiagnostic Techniques	Results
Before treatment	
Cercariae hullen reaction (CHR) in CSF	Positive (++)
CHR in serum	Positive (++++)
Immunoelectrodiffusion (anti-Sm antibodies) for circulating antigens detection in serum	Positive - two traces
After treatment	
CHR in serum	Positive (+)
CSF, cerebrospinal fluid	

related to the disease itself and its treatment.² They also indicate that improvement is usually progressive and more rapid at the beginning of corticosteroid and antischistosomal therapy.² In fact, in our patient significant improvement was noted 1 month after the onset of treatment. Although schistosoma infection affects 200 million people worldwide,⁴ NS is not commonly diagnosed, probably because it is still under-recognised.³ It is estimated that in sub-Saharan Africa schistosomiasis causes 1–5% of non-traumatic spinal cord lesions.¹ NS is a disease that can lead to permanent neurological disability, but which is potentially treatable, especially if adequate therapy is instituted early on. A high level of suspicion should be considered in those patients presenting with a spinal cord syndrome, in the appropriate epidemiological setting, for a promptly treatment even before definite diagnosis.

Learning points

- ▶ NS, namely myeloradiculopathy, is a disease that can lead to permanent and serious neurological disability.
- ▶ NS is potentially treatable if adequate therapy is instituted early on, by that, the recognition of the disease by physicians is very important.
- ▶ Schistosomiasis is an endemic disease in Africa, South America like in Brazil and the Caribbean Islands, but not in Europe. Therefore, it is rarely diagnosed in Europe, but should be considered in travelers returning or emigrants from endemic areas.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Chen AW**, Alam MH, Williamson JM, *et al*. An unusually late presentation of neuroschistosomiasis. *J Infect* 2006;**53**:e155–8.
2. **Yu-Gang Jiang**, Zhang MM, Xiang J. Spinal cord schistosomiasis japonica: a report of 4 cases. *Surgical Neurology* 2008;**69**:392–7.
3. **Nobre V**, Silva LC, Ribas JG, *et al*. Schistosomal myeloradiculopathy due to *Schistosoma mansoni*: report on 23 cases. *Mem Inst Oswaldo Cruz* 2001;**96** Suppl:137–41.
4. WHO Expert Committee. Prevention and control of schistosomiasis and soil-transmitted helminthiasis. Geneva, 8–14 October 2001. *World Health Organ Tech Rep Ser.* 2002;**912**:1–57.
5. **Ferrari TC**, Moreira PR, Cunha AS. Spinal cord schistosomiasis: a prospective study of 63 cases emphasizing clinical and therapeutic aspects. *J Clin Neurosci* 2004;**11**:246–53.
6. **Moreno-Carvalho OA**, Nascimento-Carvalho CM, Bacelar AL, *et al*. Clinical and cerebrospinal fluid (CSF) profile and CSF criteria for the diagnosis of spinal cord schistosomiasis. *Arq Neuropsiquiatr* 2003;**61**:353–8.
7. **Samandouras G**, King A, Kellerman AJ. *Schistosoma haematobium* presenting as an intrinsic conus tumour. *Br J Neurosurg* 2002;**16**:296–300.
8. **Wan F**, Li L, Chen J, *et al*. Conus medullaris schistosomiasis. *J Neurosurg Spine* 2006;**5**:146–9.
9. **Lambertucci JR**, Silva LC, do Amaral RS. Guidelines for the diagnosis and treatment of schistosomal myeloradiculopathy. *Rev Soc Bras Med Trop* 2007;**40**:574–81.
10. **Sheta ESA**, Saadany S. Schistosomiasis. *Tanta Medical Sciences Journal* 2006;**1**:1–10.
11. **Murinello A**, Germano N, Mendonça P, *et al*. Liver disease due to *Schistosoma guineensis* – a review. *J Port Gastroenterol* 2006;**13**:97–104.
12. **Sandalcioglu IE**, Gasser T, Asgari S, *et al*. Functional outcome after surgical treatment of intramedullary spinal cord tumors: experience with 78 patients. *Spinal Cord* 2005;**43**:34–41.

This pdf has been created automatically from the final edited text and images.

Copyright 2011 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Pita Lobo P, Coelho M, Gerales R, Santos C, Grácio M, Rosa MM, Antunes JL. Myeloradiculopathy associated to *Schistosoma mansoni*. *BMJ Case Reports* 2011;**10**.1136/bcr.12.2010.3631, date of publication

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow