Schistosomiasis of the spinal cord: case report

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Summary

Schistosomiasis of the central nervous system is a rare condition and affection of the spinal cord is even less common than that of the brain, the conus medullaris being the primary site of spinal involvement. We describe the case of a 42-year-old caucasian male with initial sphincter dysfunction, followed by sudden paraparesis caused by schistosoma haematobium infection of the spinal cord. There was no previous history of bilharziasial infestation. Neurological deficits improved markedly after surgery and medical treatment.


Case report

A 42-year-old patient was admitted to our Hospital due to severe paraparesis established during the previous night. He went to bed with normal strength in both legs and woke up the next morning incapable of walking. About a week before he referred progressive constipation, associated in the last two days with urine retention.

Before the present illness he had always enjoyed good health, exception made for a surgery for two herniated lumbar discs (L3 - L4 and L4 - L5) in 1980. By that time his complaints included bilateral pain irradiating from the lumbar region to both ankles. Although we have no available clinical or surgical records, the fact is that the pain disappeared after surgery. He had always lived in Portugal except for 1.5 years (1978-79) in Mozambique during the colonial war in this former Portuguese colony.

On examination, his mental state, cranial nerve functions and neurological tests for the upper limbs were all normal. A severe flaccid paraparesis was present (grade 2 muscular strength in both legs), with normal cutaneous abdominal reflexes, diminution of patellar and ankle reflexes, absence of cutaneous plantar response and deficit of all sensory modalities below the L2 dermatome. A urinary catheter was necessary to relieve bladder retention.

Laboratory investigations including tests for syphillis and tuberculosis were normal, but a search for Schistosoma eggs in stool or urine was not performed. Spine roentgenograms were also normal but M.R.I. showed an intramedullary lesion at the level of the conus medullaris (Fig. 1). Meanwhile, his clinical condition improved and when surgery was performed (March 1994), with the pre-operative probable diagnosis of intramedullary tumor, he already had attained grade 4 muscular strength in both legs.
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Fig. 1. - TI weighted images of M.R.I. after gadolineum injection showing a intramedullary lesion at conus level with heterogeneous contrast enhancement.

After T12 - L3 laminectomy and opening of normal dura we could see a slightly enlarged spinal cord just above the conus medullaris. A midline mielotomy was performed at that level and we found a friable grayish low vascular tissue with ill-defined margins. A biopsy of that tissue was accomplished and histological examination revealed typical schistosoma granulomata with ova of schistosoma haematobium surrounded by eosinophils and macrophages (Fig. 2).

Post-operative search of schistosoma eggs in faeces and urine was negative and a cistoscopy with biopsy disclosed no lesions. Medical treatment with praziquantel (single dose of 40 mg/kg), dexametasone and physiotherapy was then performed. Two months later the patient recovered bladder control and now, two years after surgery, still remains with some weakness of both legs but has an independent life. M.R.I. seven months after treatment showed a smaller lesion (Fig. 3), being these images essentially the same as in a more recent one.

Discussion

Schistosomiasis or bilharziasis has humans as primary hosts, but schistosoma presents a complex life cycle involving a specific secondary host, a water snail. The disease is acquired by men while swimming, bathing, wading or working in infected fresh water (rarely by drinking) and, after skin penetration, the worms reach the circulatory system in 4 to 5 days. Schistosoma mature and couple in the liver and then migrate through the portal system against the blood stream into the mesenteric or vesical veins to initiate egg-laying. The ovae can then reach any organ migrating through thin blood vessels. Part of the eggs escape into stool or urine and the life cycle of the worm is completed in an appropriate snail living in contaminated water.6,8,10,12. Humans can be infected by one of the five species of schistosoma (haematobium, mansoni, japonicum, intercalatum, mekongi), each of them with specific geographic areas of distribution around the world in accordance with the specific water snail intermediate.13

Central nervous system schistosoma infections are rare. Herskowitz2 in 1972 collected 104 cases, with 25 of them limited to the spinal cord. Scrimgeour and Gajdusek13 in 1985 reviewed 64 patients with mielopathy caused by schistosoma, the majority caused by s. mansoni and only in 12 cases by s. haematobium. This incidence is probably due to the fact that s. mansoni ovae are larger and with a lateral spin that may prevent their progression along blood vessels, causing them to be lodged in the region of the vertebral venous plexus12; s. haematobium ovae are smaller but nevertheless responsible for some cases of mielopathy14-16. This species is highly endemic in the entire Nile Valley and has spread over practically all of Africa16. Spinal cord involvement is most frequently described in men between 8 and 55 years-old13, being the predominance of male cases probably because men in rural areas are more likely to contact with infected water during childhood and in the course of their work13.
As schistosoma haematobium lies essentially in the venous supply of the genitourinary tract, the reflux from here to spinal cord venous plexus by way of valveless anastomotic veins, facilitated by repeated Valsalva manoeuvres, is the mechanism most authors propose to explain attainment of the spinal cord. However, ova are not found frequently enough inside the spinal cord, and so other factors are probably involved. Alternatively, arterial embolization of the ova through the great anterior radicular artery could also explain why conus medullaris is the primary site of spinal lesions. Because of the absence of susceptible snails, s. haematobium's life cycle cannot be established in Portugal, and so we think that our patient contacted the parasite during his stay in Africa. In infected individuals who migrate to non-endemic areas, the parasite's lifespan is usually in the range of 3 to 10 years, but, as happened with our patient, may be longer.

There are no clinical features specific to schistosome mielopathy and most patients have no other clinical evidence of bilharziasis. Spinal lesions may manifest themselves either as acute necrotizing myelitis or as a mass producing lesions and, in some cases, patients may be asymptomatic. Low back pain and leg pain, weakness with sphincter disturbance and sensory loss in the lower limbs are early symptoms in the majority of patients. Most frequently they are caused by an intramedullary granuloma of the conus medullaris, in some cases with extension to the cauda equina. Symptoms may begin suddenly or progress rapidly in spite of ova present within the spinal cord for months or years. There is no obvious reason (occasionally diminished defences?) for the beginning of symptoms, that is usually ascribed to an immunological reaction to the ova (of a delayed hypersensitivity type) with rapidly evolving tissue destruction or mass effect. Although there are no reported cases where lesions were confined to cauda equina, we think that it is possible that in our patient the initial episode of pain in both legs can be related with the involvement of cauda equina by the schistosoma.

Frequently, in non endemic areas, diagnosis is considered only after biopsy, as what happened in our case.
Our patient had no blood eosinophilia, as can occur in some long standing cases and we did not examined cerebrospinal fluid, as the most probable pre-operative diagnosis was intramedullary tumor. Anyway, patients with symptoms of a rapidly progressing transverse myelitis or of an intrinsic spinal cord tumor and that have stayed in an endemic area, even without any past history of active intestinal or urinary tract infections, should be evaluated for schistosomiasis by identifying ova in faeces or urine or by intracutaneous and serologic tests.

Surgery has been used more frequently for diagnosis than for treatment. Laminectomy and biopsy are useful in decompressing obstructive lesions and establishing a diagnosis. These remain indications for surgery when a spinal cord mass is evident in M.R.I. However, in endemic areas, patients with appropriate clinical and laboratorial findings should be given a therapeutic trial of praziquantel before considering surgery. When diagnosis is made just after biopsy, surgery should be followed by a single dose of praziquantel (40 mg/kg) and steroids for one or two weeks. The change in images of pre and post-operative M.R.I. is probably due to diminishing in size of lesions after treatment. Our patient improved markedly after that treatment. Residual deficits may be the result of ischaemic changes caused by vascular occlusion by eggs.

In conclusion, we think that beyond endemic areas, where schistosomiasis is one of the main health problems, the increase in world travel and immigration either by civilian or military personal are likely to produce sporadic cases and probably an increase in numbers. So, even in non endemic areas this uncommon spinal cord disease should be considered in the differential diagnosis of intramedullary lesions, namely at the conus medullaris level.

References