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Case Report

Medullary schistosomiasis

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Abstract

Background: Schistosomal infestation of the central nervous system is a rare cause of cord compression, although a predominant one in endemic areas.

Case Description: A 38-year-old male, native of Ivory Coast, with a history of 1 month of progressive paraparesis, neurogenic bladder, diminished deep tendon reflexes of the lower limbs, and sensory level. The magnetic resonance imaging (MRI) showed a medullary lesion at D4-D5 level, suggestive of an intramedullary tumor. Laminotomy of D3 to D5 and excision of a grayish white lesion according to a preliminary histopathologic review suggestive of a high grade glioma. Definitive histopathology review established the diagnosis of medullary schistosomiasis.

Conclusion: Schistosomal myeloradiculopathy should be considered in patients presenting with cord compression or features of transverse myelitis, especially in patients from endemic areas or low social economic settlements.

Key Words: Differential diagnosis, medullary schistosomiasis, outcome

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INTRODUCTION

According to the World Health Organization (WHO) about 200 million people are infected with schistosomiasis worldwide.[5,12] Schistosomal infestation of the central nervous system is known since 1899, when Yamagiva identified a brain granuloma in autopsy of a patient with Katayama disease,[13] later identified in 1904 by Katsurada to be caused by *Schistosoma japonicum*.[6] In 1911, the first case of transverse myelitis to schistossoma was described by Day, with autopsy finding of *Schistosoma haematobium* eggs in the medullary cord. The prevalence of egg deposition in the medullary cord varies among studies, ranging from 0.3% to 13%.[3] In 1940, Muller and Stender reported the first infestation by *Schistosoma mansoni* in the medullary cord, and in 1945 Gama and Marques de Sá described for the first time a tumoral schistosomiasis of the medullary cord.[13]

CASE REPORT

We report a case of a 38-year-old male, native of Ivory Coast, living in Portugal for the past 6 years, presenting with a history of 1 month of deterioration of motor function in his lower limbs with symptoms of neurogenic bladder (urinary retention and overflow incontinence). The patient was admitted to our emergency room, with assymetric paraparesis with motor strength grade 3 (right lower limb) and 4 (left lower limb), with diminished deep tendon reflexes of the lower limbs, sensory level at D10, and urinary retention ensuring the need to bladder catheterization, with otherwise unremarkable neurologic examination.

The magnetic resonance imaging (MRI) documented a moderate and diffuse enlargement of medullary cord, in the D4-D5 region, with a central nonhomogeneous T2

hypersignal as well as heterogeneous enhancement after intravenous injection of gadolinium, with a granular pattern (disruption of the blood-brain barrier) [Figure 1]. Laminotomy of D3 to D5 was carried out, and the durotomy revealed an expanded cord at the level of D4 and D5. Posterior midline myelotomy was performed using microsurgical dissection [Figure 2]. A grayish coal-like lesion was found at the center of the cord, which was gently excised with micro dissectors and micro pituitary rongeurs, piecemeal, until the macroscopic tumor was removed. Preliminary histopathologic review found the lesion to be suggestive of a high grade glioma. That result being known, after gross total removal, the dura was closed with 3/0 vycril suture, and the *laminae* were not put in place as previously planned in order to allow lesion growth without cord compression.

The postoperative period was uneventful, with slight decrease in lower limb proprioception and suspended sensory level between D10 and D12 in the immediate postoperative period, which progressively disappeared [Figure 3].

The diagnosis of schistosomiasis was based solely on the demonstration of eggs and adult worm in the granuloma supplied for histopathology review [Figure 4], and later confirmed by positivity in serological tests hemagglutination, with all other work-ups inconclusive (no eggs found on stool or urine or rectal snipes, no eosinophilia in peripheral blood, no augmented VS). The CSF biochemical analysis was normal, with no pleiocytosis, no eosinophils, normal protein, and glucose levels.

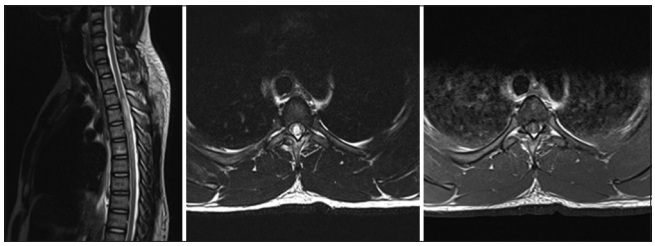

Figure 1: Preoperative MRI, revealing a hyperintense signal of the cord in the D4-D5 region, with a slight and diffuse thickening (Sagittal T2), cetromedullary (axial T2), showing a modest and diffuse enhancement with Gadolinium

On histopathological examination, the spinal cord tissue showed edema, demyelination, and a chronic inflammatory cellular infiltrate with some eosinophils. Numerous eggs were observed, some containing *miracidia* with intact nuclei and a few showed calcifications. Occasional giant cells were seen around the eggs.

As described by Salomão *et al.*,[13] the vascular alterations seen in the medullary tissue surrounding the granuloma, with vascular proliferation (presumably veins), with enlarged lumina, parietal thickening and tortuous, may very well justify the edematous enlargement of the medullary cord surrounding the granuloma (due to venous stasis), suggesting that in these forms, the best treatment should entail surgical removal of the lesion.

After diagnosis, the patient was treated medically with praziquantel at a dose of 40 mg/kg/day in two doses 6 h apart for two consecutive days and steroids (prednisolone 5 mg/day for 6 weeks).

The postoperative MRI, under steroid and antihelminthic, showed progressive reduction in the edema and enlargement of the cord, with concurrent regression of neurologic deficits, allowing deambulation with support of a clutch at his 2-month follow-up. At 6 months, deambulation was independent, maintaining urinary catheterization, however.

DISCUSSION

S. mansoni (as well as *S. haematobium*) is the commonest species in the East African and Middle East Region, it also is present in some parts of South America and the Caribbean.[3] The principal intermediate host is the *Biomphalariaglabrata* snail.

The adult worm form resides in the mesenteric or pelvic venules (*S. mansoni* is more common in the superior mesenteric veins), where the female releases the eggs, which move toward the lumen of the intestines. When in contact with water, the eggs swell due to osmotic pressure and the shell breaks open freeing the motile, ciliated *larvae (miracidia*), which penetrate the slow-moving snails that serve as intermediate hosts. Here, they transform into sporocysts, which mature into cercaria, which in turn

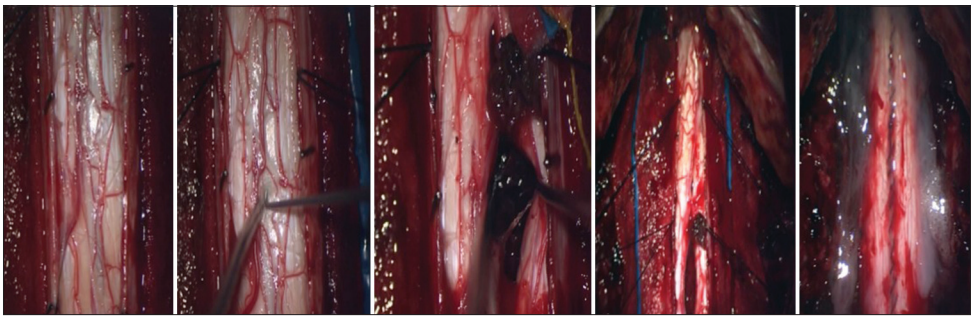

Figure 2: Intraoperative imaging of the midline myelotomy revealing the grayish coal-like intramedullary tumor

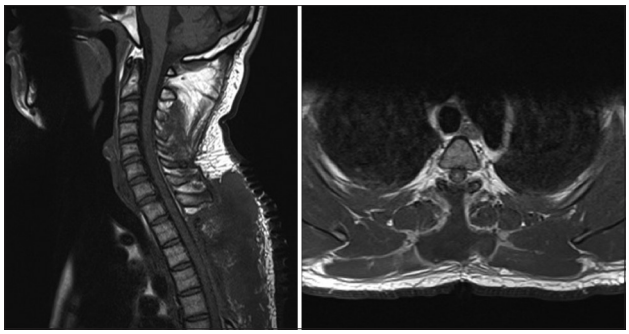

Figure 3: Immediate postoperative MRI, showing laminectomy of D3 to D5, with apparent gross total removal of the medullary granuloma (T1-weighed, sagittal, and axial)

swim and penetrate the skin of human host. The cercaria mature in the bloodstream and eventually set up in the aforementioned venules.[1,3,11]

The most accepted hypothesis of dissemination to the medullary cord is based on Batson's studies,[2] in which he showed the presence of valveless anastomosis between the pelvic veins and the perimedullary plexus, allowing the spontaneous and free blood flow upon increments of intrabdominal pressure (the presence of Schistosomal eggs linearly displayed along the medullary vessels supports the theory that the eggs may be deposited directly in the medullary circulation).[3]

The lateral spike of the egg of the *Schistosoma mansoni* may impede its further progress in embolizing the brain (the lower mass and absence of lateral spike in *S. japonicum*, make it the species responsible for cerebral infestation).[3] It also secretes a fibroblastic stimulating factor, which results in a higher probability of forming a microgranuloma. Histopathology examination unvariably reveals a tumor-like mass formed by microgranulomata containing live or dead (calcified) eggs of Schistosoma.[10]

Schistosomal myelopathy tend to occur early after infection and is more likely to be symptomatic than cerebral schistosomiasis. It may present as an acute or subacute myelopathy that may be accompanied by polyradiculitis, bowel or bladder disfunction, paraparesis, paraplegia, lumbosacral pain, and sensory disturbances.[3,9] The damage to the cord is attributable to the presence of the eggs (which may lead to vascular obstruction, granuloma formation, and hypersensitivity reactions)[13] and transverse myelitis as evidenced by the demyelination in the cord, which was associated with a chronic inflammatory cellular infiltrate around the eggs. Living adult worms cause no inflammatory response, the main pathology is caused by a granulomatous inflammation around eggs trapped in the tissues;[7,14] a delayed hypersensitivity reaction by the glial cells to the *S. mansoni* eggs.[3,16] The presence of eggs always triggers a foreign body like reaction with encapsulation in granulomata and is always accompanied by significant

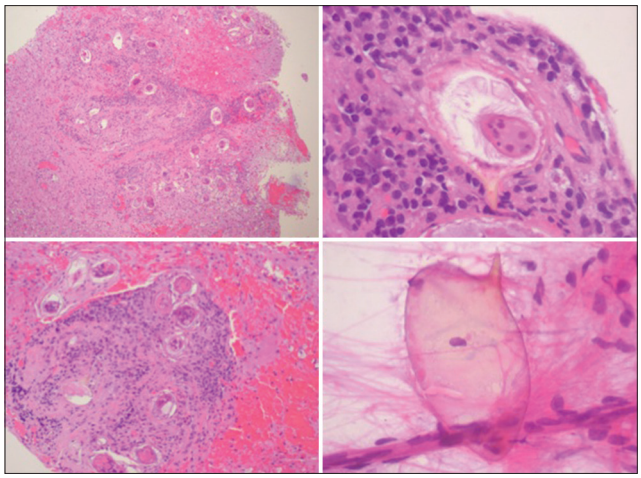

Figure 4: H and E stain, showing numerous granulomata, enclosing parasites with the characteristic spicula of *Schistossoma mansoni*

eosinophilia. There may also be some auto-immune induced lesion produced by the schistosomal infection, causing vasculitis and ischemia, but data lacks to confirm this hypothesis.

There are two described forms of medullary disease to *Schistosoma spp*: Transverse myelitis (typically to *S. haematobium*; microscopic inflammatory response, with necrosis, vacuolization, and atrophy of the nervous tissue) and granulomatous or tumoral form (typically to *S. mansoni*) due to the inflammatory response surrounding the egg, frequently in the medullary cone. Spinal schistosomiasis can also present as a progressive or acute paraparesis or tetraparesis, albeit a rare form of presentation.[3] Patients typically present with motor deficit to the lower limbs, urinary retention, and sometimes fecal retention, typically within days to months of primary infection. Its prevalence in endemic areas has been underestimated. The diagnosis relies on the presence of low thoracic/upper lumbar neurological symptoms, demonstration of the *S. mansoni* infection by microscopic or serologic techniques, and exclusion of other causes of transverse myelitis.[15]

Treatment with antischistosomal drugs interrupts egg production and fibroblastic stimulating factors by destroying the adult worm, thus limiting the inflammatory reaction of the central nervous system and granuloma formation.[10] It has been speculated that it might have an associated egg destruction action and immune response modulating effect, in light of the speedy and important recovery after its institution. The steroids reduce inflammatory reaction and as a consequence reduce the compression and destruction of the nervous tissue. It is in fact recommended to initially treat patients with acute neuroschistosomiasis with corticosteroids, in order to suppress the inflammatory response and granuloma formation, thereby preventing further tissue destruction, and reducing egg deposition.[3] Even though there is no

definitive consensual therapy for neuroschistosomiasis, consistently, the association of antischistosomal drugs and steroids produces speedy recovery of motor and sensory deficits, as well as sphincter control.[3,4,8]

Schistosomal myeloradiculopathy should be considered in patients presenting with cord compression or features of transverse myelitis, especially in patients from endemic areas or low social economic settlements. The typical presentation, with acute or subacute myelopathy was not present in this case, but instead a delayed presentation, with the late onset of myelopathy of rapid progression of parapresis with sensory disturbances and bladder dysfunction, which were not directly attributable to any specific immunological or individual variation.

It is our assumption that in light of not being previously diagnosed with Schistosomal infestation and therefore not receiving treatment for eradication, this allowed for the continuous production of viable eggs, which eventually disseminated to the medullary cord.[3]

The imaging pattern in the MRI, although not present in every case, when observed should suggest the diagnosis, which is characterized by linear central enhancement with gadolinium, surrounded by multiple punctiform nodules in a tree-like pattern.[3,11]

Diagnosis from CSF studies and stool/urine/rectal biopsy might obviate need for surgery, especially in transverse myelitis. However, in deteriorating neurological conditions, urgent surgical intervention followed by antischistosomal drug therapy remains the treatment of choice, ranging from simple decompressive laminectomy to mass exeresis.[3]

In this patient, the diagnosis of neuroschistosomiasis was not considered initially, being that there was no history suggesting primary infection, the long period (6 years) between presumed exposure and development of symptoms (with myelopathy occurring early after infection being the norm)[3,9] and also the atypical localization (thoracic medullary cord), without significant improvement with corticosteroid therapy. Therefore, surgery was proposed with diagnostic and therapeutic intent, the latter supported by the information conveyed by the preliminary histopathological exam of a high grade glioma.

It is our belief, as is of other authors,[11] that it is beneficial to entertain surgery in the initial approach of these patients, especially those with progressive neurological deficit. It allows for decompression and granuloma mass reduction, which followed by the standard treatment with antischistosomal drugs and steroids, leads to a rapid recovery of function, without morbidity related to the intervention, as was the case in this patient.

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