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Case Report EAJNS 3(1):22-26

Conus Medularis Neuroschistosomiasis In A 12-Year-Old Boy

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Abstract

Introduction: Neuroschistosomiasis, an infrequent consequence of schistosomal infection, typically presents with deteriorating motor and sensory function coupled with sphincter dysfunction. Diagnosis involves a clinical history, physical examination, cerebrospinal fluid studies, and imaging. **Case report:** We detail the case of a 12-year-old boy hailing from Machakos County, who manifested lower limb weakness and urine retention for two weeks. Examination unveiled diminished lower limb tone and areflexia, with a power grade of 0 in distal muscles below the knee and normal muscle bulk. Magnetic resonance imaging revealed a cauda equina region tumor. Subsequent histopathology after tumor-debulking surgery unveiled schistosomal-associated necrotizing granulomatous inflammation. Following tumor resection at T1-L1 and receipt of histopathology results, the patient underwent optimum pain management and praziquantel therapy. Clinical improvement ensued, albeit with distal paralysis. **Conclusion:** Our case highlights the necessity of heightened suspicion for cauda equina tumors in young patients from schistosoma-endemic regions, advocating early diagnosis and management involving praziquantel treatment.

Keywords: Neuroschistosomiasis, conus medularis

Introduction

Neuroschistosomiasis is the involvement of the CNS by parasites of the genus Schistosoma. Schistosoma parasites are transmitted through a cycle involving water contamination by specific freshwater snails and subsequent contact of the contaminated water by humans (1). The main disease-causing species are *Schistosoma mansoni*, *Schistosoma haematobium*, and *Schistosoma japonicum*. The most prevalent schistosomal species in Kenya are Schistosoma haematobium and Schistosoma mansoni (2). The WHO has recommended using praziquantel as the treatment of choice (3).

Socio-economic factors and regional endemicity highly influence the distribution of schistosomiasis. In their cross-sectional survey, Chadeka et al., (2017) demonstrated that the low socio-economic status of the 368 study participants played a crucial role in the high prevalence of the parasite (4). According to the WHO Kenya carries a substantial risk of schistosomiasis with more than 3.5 million people at risk of infection (5). This shows the high prevalence of the disease in Kenya.

The pathophysiology of schistosomiasis in the CNS has been explained in previous

studies. According to Domingues et al., 2020, the eggs of Schistosoma reach the CNS by a retrograde venous flow through Batson's venous plexus (7). Alternatively, the spread through emboli from the liver or pulmonary clinical forms has been explained in another study (6). The presence of parasite eggs in the central nervous system (CNS) can form large granulomas, which in turn can cause a mass effect and give rise to the clinical manifestations.

In neuroschistosomiasis most patients present with cauda equina syndrome features of progressively worsening motor and sensory functions. An MRI is useful in characterizing the lesion. Microscopic and immunohistochemistry assay demonstration of the parasites while excluding other causes of transverse myelitis is required for diagnosis (8).

To the best of our knowledge, only one report has hitherto described a patient with neuroschistosomiasis in the Kenyan setting (8). In this case report, we describe the second case of a 12-year-old male with neuroschistosomiasis who developed paraplegia at Machakos Level 5 Hospital. We aim to update the medical and research community regarding the occurrence, clinical presentation, severity, and outcome of the disease. The patient's guardian provided an oral consent to write the case study.

Case Report

A 12-year-old male patient from Machakos County presented with a 2-week history of Lower Limb weakness, hard stools, and inability to pass urine. The patient was well until two weeks before presenting to the facility when he developed the above symptoms. He had no history of trauma and did not report weight loss or cough with no history of cancer risk. He did not have a headache or any lateralizing signs or recall history of skin irritation. His vaccination was up to date.

He was in good general condition on examination, without pallor, jaundice, cyanosis, lymphadenopathy or oedema. His vital signs were within normal ranges. He was well oriented to time, place, and person. A CNS examination revealed normal speech and intelligence. All cranial nerves were intact, and normal muscle bulk on motor examination; he had a flaccid tone, and power was zero below the knee joint level. He had normal coordination (upper limbs).

Laboratory investigations

A complete blood count showed: White Blood Cell count $13x10^9$ cells/l, Hemoglobin16.3g/dl, and Platelets 246 $x10^9$ cells/l. UECs (Electrolytes, Urea, and Creatinine) were within normal range, urinalysis results were normal, stool examination was not done, and a head CT scan showed no abnormalities.

Imaging findings

The lumbosacral MRI revealed several findings shown in the figures above. There is an intramedullary intradural syrinx present, causing an expansion of the cauda equina as shown by the arrow in figure 1 above. Notably in figure 2, the syrinx wall exhibits moderate enhancement after the administration of intravenous contrast. However, the spinal canal itself does not display significant expansion. No changes or erosions are observed in the vertebral bodies, which maintain their normal shape in figure 1. The overall spinal curvature is preserved, and the vertebral bodies shows no abnormalities in terms of bone marrow signal intensity as shown in figure 1. The intervertebral discs appear normal, with no facet joint irregularities or nerve root exit involvement. The paraspinal soft tissues are normal as well.

Imaging: Lumbosacral MRI images showing the tumor

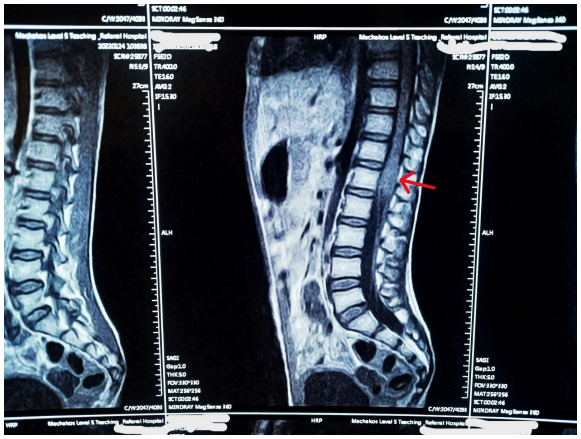

Figure 1. A lumbosacral sagittal MRI showing a cauda equina mass

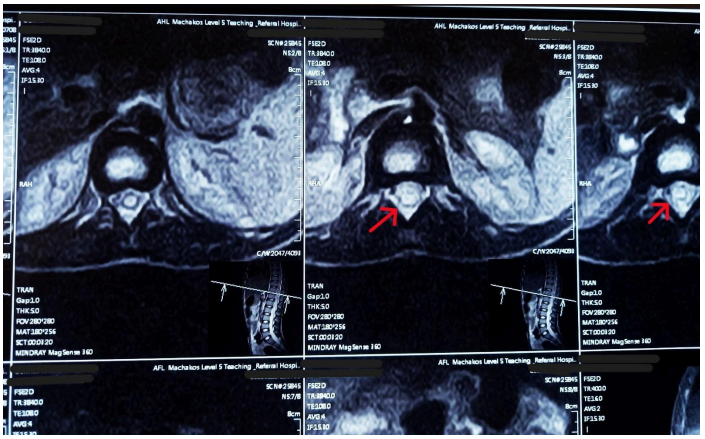

Figure 2. An axial lumbar MRI showing an intramedullary intradural syrinx

Differentials

Several possibilities were considered from the MRI findings. There was a suspicion of a syrinx involving the cauda equina. Additionally, other potential diagnoses under consideration included intramedullary astrocytoma, ependymoma, cystic schwannoma, and neurocytoma located at the cauda equina.

Intervention.

Based on the clinical presentation, the worsening neurological functions, and MRI findings of a possible tumor, the patient was admitted to the surgical ward and scheduled to undergo tumor debulking surgery. The patient underwent a debulking surgery of the tumor, and samples for histopathology were taken in the immediate postoperative period.

Histopathology results a week later showed schistosomal-associated necrotizing granulomatous inflammation but the exact schistosomal species was not isolated. Upon receiving histopathology results, praziquantel was added to the patient's treatment regimen.

Outcome.

The patient was discharged and was on follow-up at the neurosurgical clinic. Significant improvement of his neurological deficit, i.e., mainly hip flexion and sensation around the L1 and L2 region was observed for 2 weeks. Distal to this region, there was flaccid paralysis with no sensation. He has since then resumed school with assistance 1 month after discharge.

Case Discussion

We present the case of a 12-year-old boy with neuroschistosomiasis. The patient initially presented with progressively worsening neurological symptoms over a two-week period. The diagnosis of neuroschistosomiasis was confirmed post-operatively based on histopathology results of the cauda equina tumor. The patient's young age and residence in a schsitosomal endemic zone might have increased his risk of schistosoma infection (12).

In cases of spinal schistosomiasis, the transportation of eggs to the spinal veins can occur through the valveless venous plexus of Batson (11). This intricate network links the deep iliac veins and the inferior vena cava with the veins found within the spinal cord. The introduction of Schistosoma ova into these veins might

coincide with instances of heightened intraabdominal pressure, such as during episodes of coughing or defecation. This phenomenon could potentially elucidate the elevated occurrence of myelopathy specifically within the lumbosacral regions. Adult worms can migrate via leptomeningeal veins (11).

The lower thoracic or upper lumbar regions are the most typical sites of spinal cord schistosomiasis (10). This was evident in our patient based on the MRI findings (*figures 1 and 2*). Similarly, cauda equina syndrome resembling features have been seen in patients with neurological schistosomiasis with progressively worsening sensory and motor symptoms and sphincter dysfunctions. Our patient presented with areflexia and paraplegia with urine retention. A previous study showed bladder dysfunction was a constant finding in all patients (100%) (10). In the same study, most patients had radicular pain in the lower limbs and lower back pain.

The clinical diagnosis of neuroschistosomiasis relies on clinical presentation and results of different tests, including serological assays (9), immunological and biochemical assays of the CSF, parasitological confirmation of infection, imaging findings and exclusion of other causes of transverse myelitis. It is essential to conduct CSF studies and serological assays to aid in diagnosis (9). Unfortunately, in our case these were not performed due to lack of the studies in our facility and inadequate finances from the family to outsource the services.
MRI on our patient showed a tumor in the cauda equina region with the differentials of syrinx at the cauda equine, intramedullary astrocytoma, ependymoma, cystic schwannomas, and neurocytoma at the cauda equine. Later, the excised lesion's histopathologic findings were consistent with those of schistosomiasis.

Early treatment and follow-up are highly recommended to manage the disease. In their study, Domingues et al. reported MRI studies of two brothers with spinal schistosomiasis. Following recognition of the cauda equine mass, corticosteroids were the primary treatment, coupled with pain management by pregabalin. The patients improved their clinical outcomes with medical therapy (7). In contrast, a follow-up of 69 patients with neuroschistosomiasis reported no consensus in managing the disease (11). In the case study reported by Wanyoike and Qureshi, laminectomy of T11-L1 was done with posterior midline myotomy, and the lesion was excised (8). The role of surgery has been scarcely reported in the different cases of the same condition. Moreover, most studies show the crucial role of steroids in managing the patients' symptoms. Furthermore, treatment with anti-helminthic drugs is paramount in eliminating the infection.

Based on these findings, it should be noted that patients presenting with paraplegia and MRI findings of a cauda equine tumor might have neuroschistosomiasis, and these patients might be missed or misdiagnosed. Clinicians should anticipate neuroschistosomiasis based on the clinical presentation, the recent or history of travel to endemic areas, microscopic and immunohistochemistry assay demonstration of the parasites while excluding other causes of transverse myelitis (8). If diagnosed and managed early, the outcomes of patients can be improved.

Conclusion

Our case underscores the importance of having a high index of suspicion in cauda equine tumors in patients from Schistosoma endemic areas. CSF studies should be considered to evaluate antibody titers. The use of steroids and treatment with anti-helminthic drugs is paramount in the management of suspected cases.

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