Case Report

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Spinal schistosomiasis mimickingspinal tumour: a case report

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Introduction and importance: Schistosomiasis, a parasitic disease, is caused by blood flukes from the schistosoma genus. Neuroschistosomiasis is the most severe form of schistosomiasis, which occurs when the host's brain and spinal cord react to the deposition of eggs, leading to neurological symptoms. Neuroschistosomiasis causes various signs and symptoms, such as myelopathy, radiculopathy, and elevated intracranial pressure.

Case presentation: A 12-year-old child from Ethiopia who presented with progressive weakness in his lower extremities that has been ongoing for 2 months. Alongside the weakness, the patient also experienced tingling sensations and numbness in his lower extremities. Additionally, he had bladder and bowel incontinence. Spinal MRI showed signs suggestive of myxopapillary ependymoma, but the histopathology result showed schistosomiasis. Postoperatively, the patient had a slight improvement in terms of lower extremity weakness (flickering of the digits). However, there was no improvement in his continence ability.

Clinical discussion: The most common neurological manifestation of Schistosoma mansoni infection is myelopathy, which includes subacute myeloradiculopathy and acute transverse myelitis. The cauda equina and conus medullaris are the areas most frequently affected.

Conclusion: When spinal schistosomiasis presents itself as a mimicking spinal tumour, it poses a complex clinical challenge that necessitates a comprehensive interdisciplinary approach to ensure accurate diagnosis and effective treatment. It is imperative for healthcare practitioners to enhance their knowledge and awareness of this uncommon parasitic infection, particularly in regions where it is prevalent.

Keywords: child, mimicking, schistosomiasis, spinal, surgery, tumour, weakness

Introduction

The parasite genus schistosoma has demonstrated an ability to survive for millions of years while maintaining its characteristics intact. Recent DNA sequencing studies have revealed that it originated as a parasite of hippos during the Cenozoic epoch[1]. Although relatively rare, neuroschistosomiasis can have serious consequences for the central nervous system, affecting the brain and/or spinal cord. In communities where the disease is prevalent, the estimated prevalence of spinal schistosomiasis cases, which range from 1 to 4%, may be overestimated[2]. One reason for misdiagnosis is the similarity between spinal schistosomiasis and spinal malignancies. Therefore, it is crucial to consider parasite infections as a potential cause when evaluating patients with spinal cord diseases. It is believed that when mature worms become lodged in the spinal cord or brain microcirculation, they release eggs, triggering a severe granulomatous reaction that leads to the destruction and scarring of local tissue[3].

Symptoms of spinal schistosomiasis can include double incontinence, discomfort and weakness in the lower limbs, and low back pain[4]. To accurately diagnose parasitic spinal infections, various imaging techniques, laboratory tests, and a thorough medical history are essential[5]. In addition, certain surgical procedures, such as decompressive laminectomy and debulking of intramedullary lesions, may be necessary[6]. Attaining complete remission requires the use of anti-schistosomal medications as part of medical therapy. Taking all factors into consideration, a high level of suspicion, thorough evaluation, and coordinated

HIGHLIGHTS

- Neuroschistosomiasis is the most severe form of schistosomiasis, which occurs when the host's brain and spinal cord react to the deposition of eggs.
- Symptoms of spinal schistosomiasis can include double incontinence, discomfort and weakness in the lower limbs, and low back pain.
- When spinal schistosomiasis presents itself as a mimicking spinal tumour, it poses a complex clinical challenge.

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care from multiple disciplines are imperative for an accurate diagnosis and successful treatment of spinal schistosomiasis[7]. We hereby present a case of spinal schistosomiasis in a paediatric patient who initially exhibited characteristics resembling a tumour.

'This case report has been reported in line with the SCARE Criteria'[8].

Clinical presentation

The patient is a 12-year-old child from Ethiopia who presented with a progressive weakness in his lower extremities that has been ongoing for 2 months. The weakness initially started in the distal muscles and has gradually spread to involve the proximal muscles. Alongside the weakness, the patient also experienced tingling sensations and numbness in his lower extremities. Additionally, he had bladder and bowel incontinence. The patient denied experiencing any headache, nausea, vomiting, fevers, chills, cough, rhinorrhea, diarrhoea, or recent contact with sick individuals. He had no history of hemoptysis, right upper quadrant pain, yellowish discoloration of the eye, seizure or focal neurologic deficit. Although he frequently came into contact with river water, he had no history of bloody diarrhoea or previous treatment for schistosomiasis.

During the examination, the patient appeared well-nourished and in good overall health. His vital signs were stable, and he showed no signs of acute distress. Neurologically, the patient was awake, alert, and oriented to person, place, and date. His cranial nerves were functioning normally. While his upper extremities exhibited full strength bilaterally with no pronator drift, his lower extremities displayed zero strength throughout all muscle groups and were hypotonic. There was no specific sensory level, and sensation was intact for pain, light touch, vibration, and proprioception. However, the patient experienced heightened sensitivity (hyperaesthesia) throughout his lower extremities. Biceps tendon reflexes were normal bilaterally, but reflexes were absent at the knees and ankles on both sides.

The laboratory investigation revealed a normal complete blood count, with no eosinophilia on the differential. The patient's organ function tests were within the normal limits, and stool and urine examinations did not provide any significant findings. However, an MRI scan showed a $2.4 \times 1.1 \times 1.6$ cm eccentrically located intramedullary mass at the conus medullaris. The mass appeared T1 isointense and T2 hyperintense, with avid post-contrast homogenous enhancement. Cord expansion with effacement of cerebrospinal fluid and cord oedema at the superior aspect of the mass were also observed (Figs. 1–3). Based on the above finding, the diagnosis of myxopapillary ependymoma was made, but no abnormalities were detected in the bones or intervertebral discs.

Based on the preoperative diagnosis of myxopapillary ependymoma, the patient underwent T1–L1 laminectomy, myelotomy and mass excision. The excised mass was sent for histopathologic analysis. Following the surgery, the patient was started on ceftriaxone 800 mg IV twice a day, dexamethasone 4 mg IV four times a day (followed by prednisolone 40 mg po daily for 3 weeks), diclofenac 50 mg IM twice a day, and received continuous bladder and bowel care. Unfortunately, the patient developed a urinary tract infection and the ceftriaxone 1 gm IV was continued for 7 days. To our surprise, the pathology result revealed spinal schistosomiasis (Figs. 4–6). After that the patient was treated with praziquantel 40 mg/kg for 7 days.

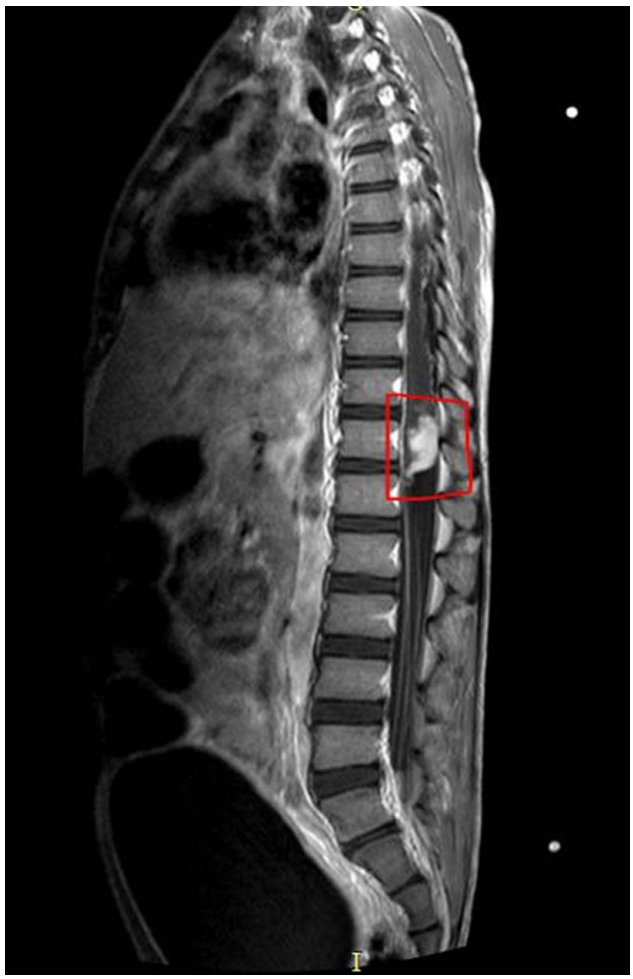

Figure 1. MRI of the patient (sagittal section). Red arrow showed the lesions in the spinal cord.

Postoperatively, the patient had a slight improvement in terms of lower extremity weakness (flickering of the digits). However, there was no improvement in his continence ability. The postoperative MRI images showed below (Fig. 7).

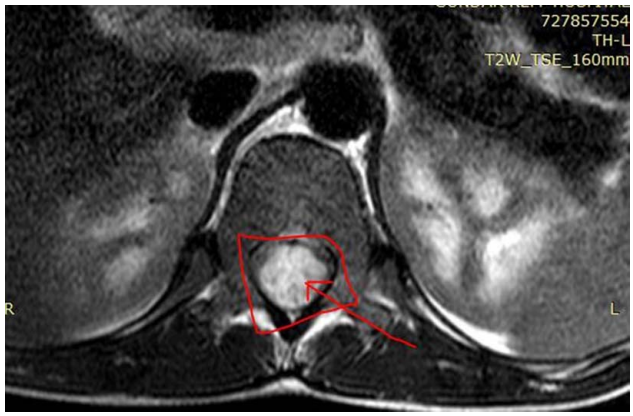

Figure 2. MRI of the patient (axial section). Red arrow showed the lesions in the spinal cord.

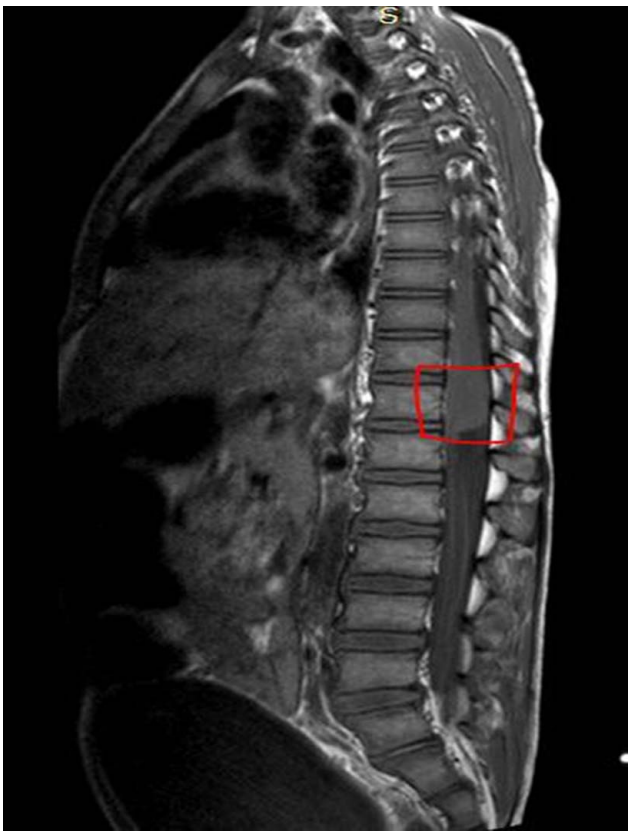

Figure 3. MRI scan showing a 2.4 × 1.1 × 1.6 cm eccentrically located intramedullary mass at the conus medullaris. The mass appeared T1 isointense and T2 hyperintense, with avid post-contrast homogenous enhancement. Red arrow showed the lesions in the spinal cord.

Discussion

Schistosomiasis, a parasitic disease, is caused by blood flukes from the schistosoma genus. Currently, over 200 million individuals worldwide are affected by this disease[9]. One severe form of schistosomiasis is neuroschistosomiasis, which occurs when the host's brain and spinal cord react to the deposition of eggs, leading to neurological symptoms. These symptoms can include delirium, loss of consciousness, seizures, difficulty speaking, impaired vision, motor impairments, and unsteady movements, among others, in cases of cerebral schistosomiasis. The most common neurological manifastation of Schistosoma mansoni infection is myelopathy, which includes subacute myeloradiculopathy and acute transverse myelitis. Compared to cerebral schistosomiasis, schistosomal myelopathy is more likely to cause symptoms and develop soon after infection. The cauda equina and conus medullaris are the areas most frequently affected. Severe cases of schistosomal myelopathy can result in complete paralysis of the lower limbs, accompanied by loss of reflexes, dysfunction of the sphincter muscles, and sensory abnormalities like this patient[10,11].

There are four primary types of spinal schistosomiasis, each characterized by its own distinct set of clinical and anatomical symptoms. The most common presentation of the disease in the lower thoracic and lumbar cord is transverse myelitis, which can be used to differentiate spinal schistosomiasis from other forms of

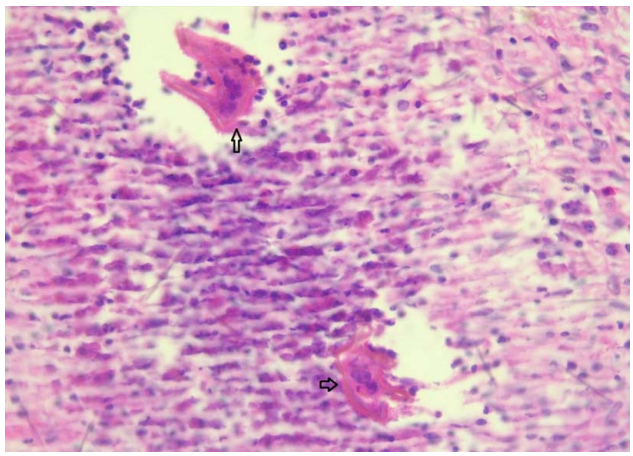

Figure 5. Histopathology showing schistosoma eggs surrounded by eosinophils and epithelioid histiocytes. black arrow shows the schistosoma parasite.

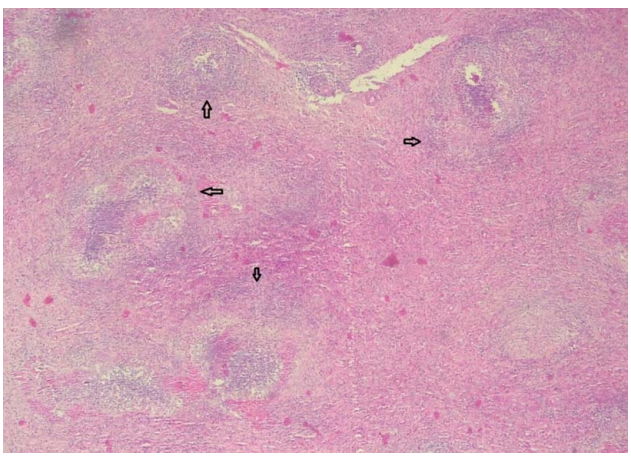

Figure 4. Histopathology section of the excised sample showing multiple epithelioid granulomas (arrows). black arrow shows the schistosoma parasite.

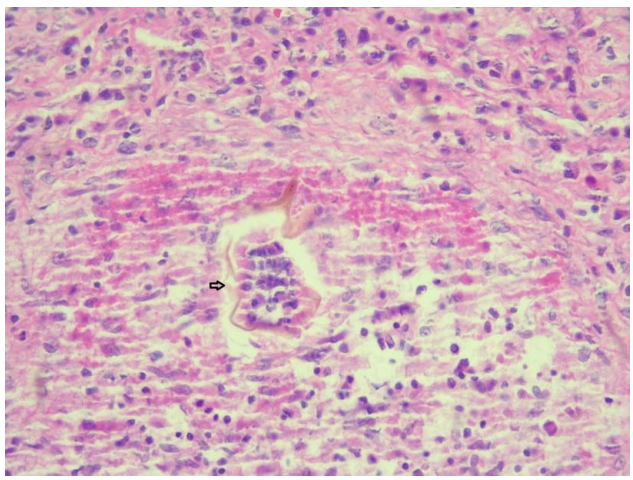

Figure 6. Histopathology showing schistosoma eggs surrounded by eosinophils and epithelioid histiocytes. black arrow shows the schistosoma parasite.

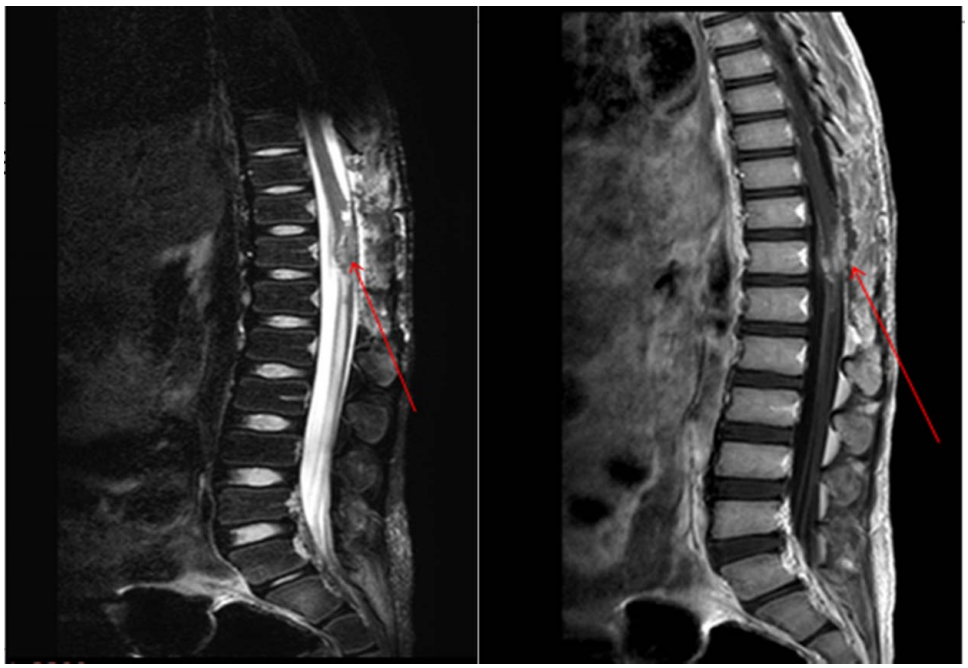

Figure 7. Postoperative MRI revealing T1 isointensity and T2 and SPIR hyperintensity, along with post-contrast peripheral and patchy enhancement at the T11 vertebral level of the spinal cord and T9–T12 vertebral level paraspinal soft tissue. Notably, there is an absence of the T9–T12 vertebral spinous process, indicative of postoperative haematoma and paravertebral soft tissue oedema. Red arrow showed the lesions in the spinal cord.

myelitis[12]. In our case, we observed a granulomatous tumour in the conus and cauda equina, which is the second most prevalent symptom. The positioning of these lesions in the lower cord is likely due to the anatomical pathway of the pelvic and intra-abdominal veins, as well as their connections with the peri-vertebral venous plexus[13]. The third clinical sign of spinal schistosomiasis is the presence of extra-axial granuloma followed by cord compression[14]. Lastly, although rare, spinal schistosomiasis can manifest as acute anterior spinal artery blockage and cord infarction due to localized granuloma extension and arteritis[15,16].

Neuroschistosomiasis causes various signs and symptoms, such as myelopathy, radiculopathy, and elevated intracranial pressure. If left untreated, these lesions can lead to permanent glial scars. Spinal symptoms may include lumbar discomfort, lower limb radicular pain, muscular weakness, sensory loss, and bladder dysfunction[17]. The connection between the internal vertebral venous plexus and the lumbar veins, which are tributaries of the inferior vena cava, allows ectopic ova to enter the central nervous system. These ova can then lodge in the nearby spinal cord, leading to the formation of granulomas[10]. Alternatively, they can travel upwards towards the brain tissue during coughing or straining. The preference of S. japonicum in causing cerebral schistosomiasis can be attributed to the small size of its eggs, which makes it easier for them to enter the brain. On the other hand, S. mansoni and S. haematobium infections are more commonly associated with myelopathy in the lumbosacral area[18].

The differential diagnosis of a progressive spinal cord illness encompasses various conditions, such as demyelinating, inflammatory, infectious, and tumours affecting the spinal cord[19]. In ~60% of cases, stool testing fails to detect S. mansoni eggs, even when multiple samples are examined on different days like this patient. Rectal biopsies are considered the most reliable method for confirming the presence of an active schistosomal infection due to their high sensitivity in identifying eggs. However, the presence of eggs in the stool or a positive serology only provides suggestive, not definitive, evidence of schistosomal myelopathy in areas where the disease is prevalent like Ethiopia. Serological detection of schistosomiasis is limited in endemic regions due to potential cross-reactions with other antigens, making it challenging to distinguish between an ongoing infection and a past one, even after successful treatment. For individuals travelling to endemic areas, serologic testing may be more beneficial in diagnosing schistosomiasis. Additionally, blood eosinophilia, although not always present, is another suggestive but non-specific sign[20]. Our patient did not have eosinophilia, but rectal biopsy and serologic tests were not done.

On MRI, the signs of spinal schistosomiasis include the involvement of long segments, patchy enhancements, and poorly defined intramedullary lesions with cord extension[3]. Other possible findings on MRI include a mass effect, necrosis, cystic degeneration, intralesional bleeding, and perilesional oedema. When using T1-weighted imaging (T1WI), nodular and mass lesions may appear hyperintense or isointense, while with T2-weighted imaging (T2WI), they appear hyperintense. Lesions may exhibit homogenous or ring-shaped enhancement, as well as clustered nodules. Additionally, gyral enhancement, leptomeningeal enhancement, and central linear enhancement may be observed[21]. MRI findings may also include spinal cord oedema, enhancement in the cauda equina area, and longitudinally widespread transverse myelitis[22]. However, for a definitive diagnosis of spinal schistosomiasis, histological testing of a spinal cord lesion would be necessary[19].

With regard to the treatment of spinal schistosomiasis, active lesions readily respond to anti-schistosomal chemotherapy. The drug of choice in current practice is Praziquantel (PZQ), a pyrazinoisoquinoline derivative[23]. With an impressive 80% cure rate, PZQ effectively combats all species of human pathogenic schistosome. However, it is important to note that PZQ only targets adult worms and therefore cannot be used for chemoprophylaxis. For the treatment of S. haematobium and S. mansoni, a single morning dosage of 40 mg/kg body weight is recommended as the therapeutic dose. In the case of S. japonicum, two separate doses of 60 mg/kg body weight should be administered on the same day, effectively doubling the dosage. In the case of S. mekongi, the same dosage is administered, ideally split into three doses and given on the same day.

While PZQ is generally safe, a few adverse effects have been observed, including headaches, dizziness, skin rash, and abdominal discomfort. It is worth noting that human resistance to PZQ is still uncommon in S. mansoni and has not been documented in S. haematobium. Meteifonate, an organophosphorus preparation, is specifically effective against S. haematobium infections and is considered safe for widespread use. To achieve a high cure rate, a single dosage of 10 mg/kg body weight is recommended, which can be repeated every two weeks. Oxamniquine, a derivative of quinoline (2-aminomethyl-tetrahydro-quinoline), is effective against S. mansoni infections. A single-dose medication of 20 mg per kg of body weight is advised for optimal results[24].

Depending on the specific species of the parasite, the organ or organs that are affected, the duration of the infection, and the severity of the damage already inflicted, it is possible that established lesions may or may not be reversible after successful treatment with anti-schistosomal drugs. For instance, in cases of S. mansoni infections, early lesions in the colon or liver, or early lesions in the bladder or back pressure in S. hematobium infections, or even early brain lesions in S. japonicum illness tend to have the most favourable outcomes when treated promptly[17]. In our particular case, the patient exhibited a significant degree of weakness and experienced double incontinence upon initial assessment. Regrettably, there was only a dismal observed improvement in the patient's condition. Finally, the patient claimed he was satisfied with the care he was provided.

In the realm of schistosomiasis prevention, it is imperative to acknowledge that schistosoma is frequently transmitted through tainted freshwater sources. Various preventive measures must be implemented to combat this disease, including refraining from contact with contaminated water (may not be possible in a set-up like ours), utilizing safe water sources, maintaining proper sanitation and hygiene practices, donning protective clothing, disseminating health education and awareness, and undergoing medical screening and treatment. These strategies are crucial in safeguarding individuals from the debilitating effects of schistosomiasis.

Limitations

Diagnostic challenges: The similarity between spinal schistosomiasis and spinal tumours leading misdiagnosis, requiring a high level of suspicion and thorough evaluation.

Treatment efficacy: The paper discusses the effectiveness of Praziquantel treatment but acknowledges that established lesions may not be reversible even after successful treatment

Case specificity: The findings are based on a single paediatric case, which may limit the generalizability of the results and conclusions.

Conclusion

When spinal schistosomiasis presents itself as a mimicking spinal tumour, it poses a complex clinical challenge that necessitates a comprehensive interdisciplinary approach to ensure accurate diagnosis and effective treatment. It is imperative for healthcare practitioners to enhance their knowledge and awareness of this uncommon parasitic infection, particularly in regions where it is prevalent. This heightened awareness will facilitate timely diagnosis and treatment, ultimately leading to enhanced patient outcomes.

Ethical clearance

The case report has been submitted for Ethical Board Review and approved as an ethically sound report.

Consent

Written informed consent was taken from the patient's father for publication of this case report and any accompanying images. A copy of the written consent is available for review for the editor-in-chief of this journal on request.

Source of funding

Not applicable.

Author contribution

All authors contributed to the conception, writing and editing of the case report. All authors are agreed to be accountable for all aspects of it.

Conflicts of interest disclosure

No potential conflict of interest relevant to this article was reported.

Research registration unique identifying number (UIN)

Not applicable.

Guarantor

Dr Yohannis Derbew Molla, Dr Esayas Adefris Tefera, and Dr Almaz Enku Selamawi.

Availability of data and materials

The authors of this manuscript are willing to provide any additional information regarding the case report.

Provenance and peer review

Not invited.

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