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Case Report

A case report of conus intramedullary mansoni neuroschistosomiasis

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ABSTRACT

Background: Conus schistosomiasis is an extremely rare entity in which patients present with a wide range of neurological symptoms such as low back pain, paresthesia, fever, myalgia, paraparesis, and paraplegia. This neglected tropical parasitic disease causes significant neurological morbidity. The migration of parasitic worms and their eggs into the central nervous system can lead to profound and life-altering disabilities. Early, accurate diagnosis, and treatment can lead to the complete resolution of symptoms.

Case Description: A 5-year-old boy from South Sudan presented with complaints of sudden onset, progressive bilateral lower limb weakness for the past month, inability to walk or stand without assistance, and urinary incontinence. Magnetic resonance imaging spine showed an irregular enhancing lesion within the conus. The various possibilities, such as Astrocytoma, and granulomatous disorders were considered. However, a biopsy revealed the lesion to be Conus schistosomiasis, a rare condition with only a few cases reported in children. Typical clinicoradiological presentation and the treatment paradigm have been discussed in this manuscript. Appropriate management of this lesion can avert surgical intervention needed for either a diagnosis or treatment.

Conclusion: This case report aims to emphasize the importance of considering schistosomiasis as an important differential diagnosis of a conus intramedullary lesion, especially in patients from tropical endemic countries. The neurological recovery in this infestation is directly related to early diagnosis and treatment. Therefore, it is essential to recognize this entity, as early detection and management would result in significant neurological improvement without undergoing surgery.

Keywords: Conus intramedullary schistosomiasis, *Schistosoma mansoni*, Schistosomal myeloradiculopathy, Schistosomiasis, Spinal neuroschistosomiasis

INTRODUCTION

Neuroschistosomiasis is an unacknowledged, rare disease affecting between 1% and 4% of the estimated 200–300 million people with systemic schistosomal infections.[11] Schistosomiasis has been listed as one of the neglected tropical diseases. Schistosomiasis or Bilharziasis is a trematode platyhelminthic parasite and is the second most commonly diagnosed parasitic disease causing the most significant economic impact.[4] Neuroschistosomiasis can be classified into three clinical forms: Spinal, myeloradiculopathy, and conus/cauda equine syndrome.[11] Neuroschistosomiasis can potentially develop into irreversible scarring in the central nervous system without proper

treatment.[3] Most cerebral forms of schistosomiasis are asymptomatic, while medullary neuroschistosomiasis are symptomatic. Approximately, 40.3% of medullary schistosomiasis involves the lower thoracic spinal cord, 15.8% involves the lumbar region, and 0.7% involves the lumbosacral region. The presence of lateral spicule prevents the migration of the eggs through the smaller caliber blood vessels, which impact the eggs in the lower medullary canal. Symptoms of neuroschistosomiasis are due to the destruction of the nervous tissue, the mass effect produced by many eggs, and granulomas forming space-occupying lesion in the nervous system.[6] Classical magnetic resonance imaging (MRI) finding of spinal cord schistosomiasis is scarce, and pathologically, confirmed cases are limited.[10] MRI of the conus medullaris mass is often highly suspicious of the tumor.[5] Here, we describe our experience with a child from an endemic region with neuroschistosomiasis with a classical radiological imaging finding.

CASE PRESENTATION

A 5-year-old boy from South Sudan presented with complaints of sudden onset, progressive bilateral lower limb weakness for the past month, and inability to walk or stand without assistance. Before the onset of symptoms, he could walk and run without support. He has had urinary incontinence since the onset of weakness. He had experienced a few episodes of fever and cough for the past month. His prior medical history did not reveal any other clinical comorbidity. The family was from a peri-urban area of South Sudan known for its endemic intestinal and urogenital schistosomiasis, particularly with *Schistosoma mansoni* species.

On examination, he was noted to have grade 2 weakness of the lower limbs, right more than left. He could not stand and sit without support but could roll over in bed and had areflexia, which involved absent knee, ankle and plantar reflexes. He was also observed to have no cremasteric reflex, lax anal tone, and neurogenic bladder.

MRI of the brain with the whole spine showed an intramedullary lesion extending from D11 to L1 with irregular margins in conus medullaris and perilesional edema [Figure 1]. Differential diagnoses of astrocytoma, ependymoma, and granulomatous conditions were suspected, and he was investigated to rule out the differentials.

Urine and stool routine examinations were done to observe viable Schistosoma eggs, which were reported negative. Serum angiotensin converting enzyme (ACE) level was done to rule out Sarcoidosis, which was noted to be within normal limits. Serum neuromyelitis optica (NMO) and myelin oligodendrocyte glycoprotein (MOG) levels were done to rule out immune-mediated disorders, Brucella antibodies IgG/IgM to rule out

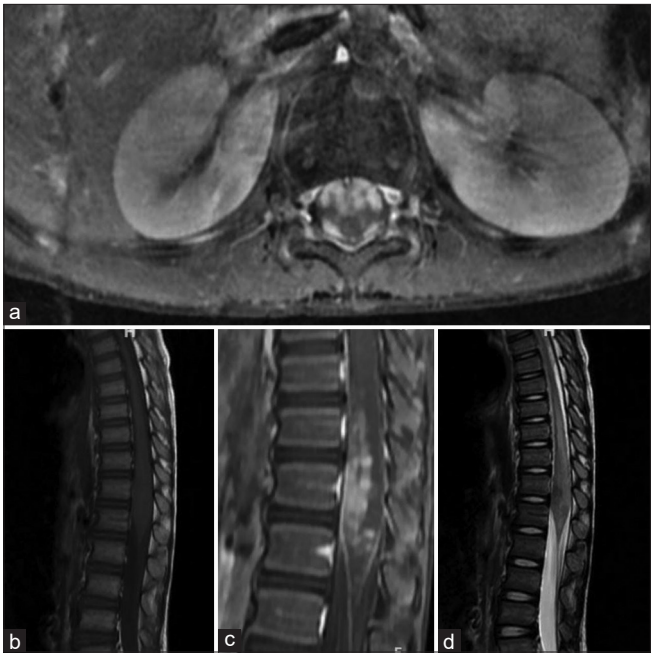

Figure 1: Preoperative magnetic resonance images (a). Axial section T1-weighted, (b). sagittal T1 sequence, (c). sagittal T2-weighted (T2W), and (d). post-contrast sagittal with IV contrast of Gadovist showing heterogeneously enhancing intramedullary lesion D11–L1 level with poorly defined margins in the conus medullaris with expansion and abnormal enhancement of the dorsal and ventral rami at L1 level with perilesional edema. (c) Sagittal T2W showed a typical "String of Bead" appearance due to multiple enhancing nodules within the conus associated with interlacing and matting.

brucellosis and immunoblot antinuclear antibody (ANA) profile to rule out antinuclear antibody-positive disease were reported negative. Systemic involvement of schistosomiasis was ruled out as well. Since there was no definitive diagnosis, a surgical biopsy was contemplated, and therefore, he underwent a laminoplasty proceed biopsy under intraoperative neuromonitoring, C-arm, and fluorescein guidance. Intraoperatively, the edematous and inflamed nerve roots were noted, along with an intramedullary conus lesion [Figure 2]. The culture of tissues showed no growth of organisms after 48 h of incubation. Gene X-pert and acid-fast bacilli were done to rule out tuberculosis, which was reported to be negative.

Histopathological examination of the spinal cord lesion showed necrotising granulomatous chronic inflammation with Schistosoma eggs and no evidence of malignancy [Figure 3]. As per the clinical, laboratory, and imaging evidence of neuroschistosomiasis, he was treated with steroids and Praziquantel 300 mg. He was initially treated with a high dose of parenteral steroid, tapered down, and converted to the oral route along with the oral anti-parasitic agent Praziquantel as recommended by the World Health Organization protocol.[7]

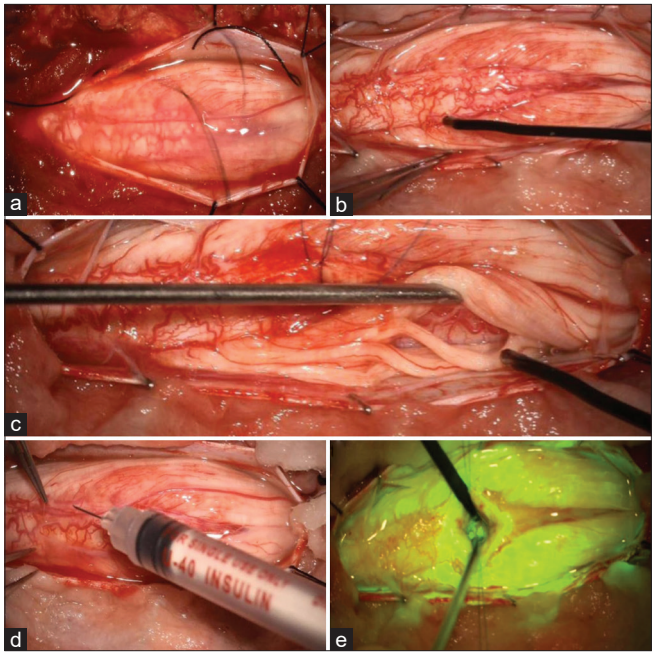

Figure 2: Intraoperative images (a) edematous and inflamed root, (b) electrode placed after sectioning, (c) exploration of the nerve roots on the left side, (d) dorsal myelotomy performed, and (e) microscopic vision under fluorescein dye. Copyright 2022 by MIOT International Hospital, Chennai. Copyright 2022 by MIOT Internatinoal Hospital, Chennai.

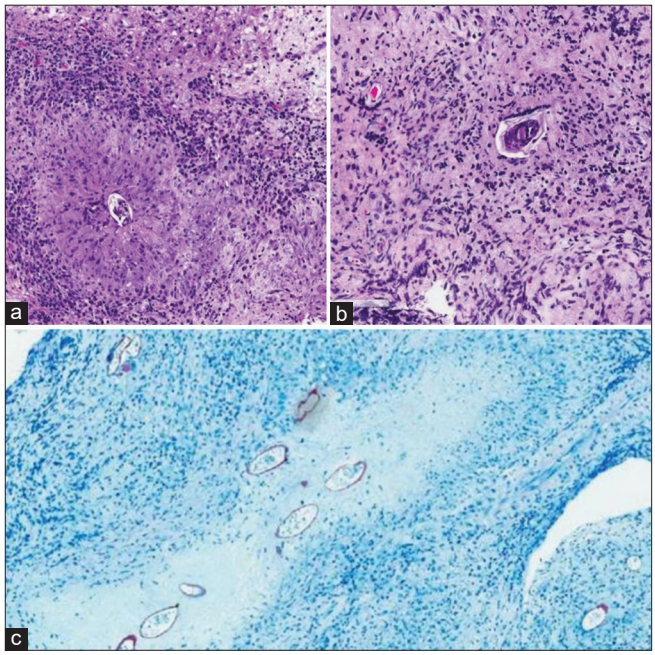

Figure 3: Histopathological images. (a) The high-power section of *Schistosoma* eggs, (b) low-power *Schistosoma* eggs with multinucleated cells showing inflammatory reaction to the egg, and (c) modified Ziehl–Neelsen staining showing *Schistosoma* eggs. Copyright 2022 by MIOT International Hospital, Chennai.

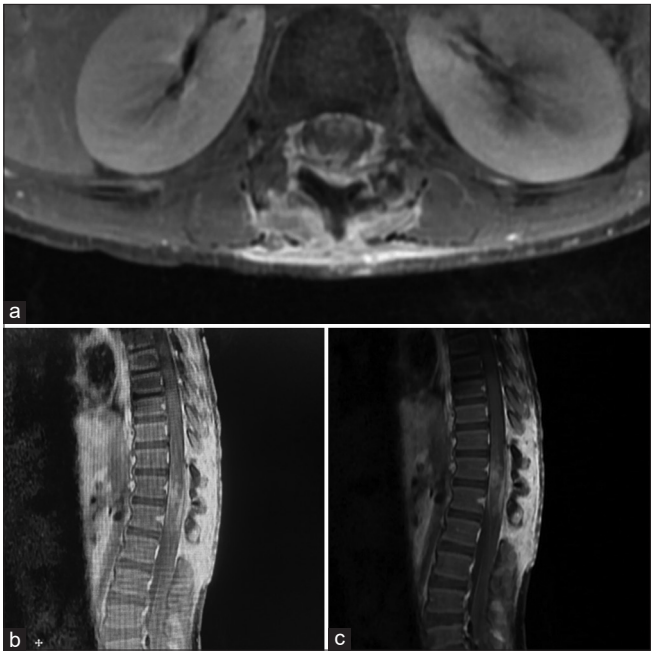

Figure 4: Postoperative magnetic resonance images (a). Axial T2-weighted, (b). sagittal T1-weighted (T1W), and (c). sagittal T1W shows a decrease in conus swelling and decreased enhancement in the residual conus medullaris lesion with edema cranial to the conus medullaris completely resolved. Copyright 2022 by MIOT International Hospital, Chennai.

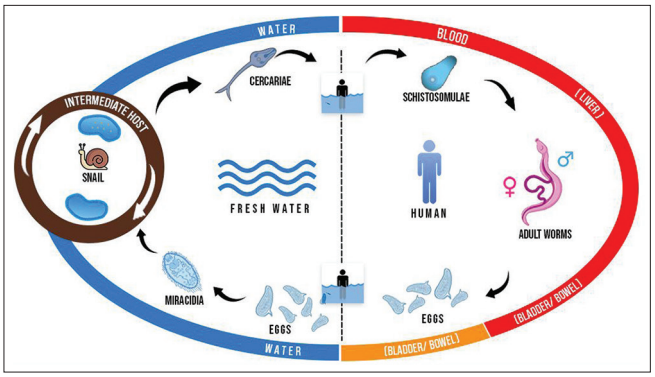

Figure 5: Life cycle of Schistosomiasis. Copyright 2022 by MIOT International Hospitals, Chennai.

Postoperatively, he was given regular intensive in-house rehabilitation and physiotherapy. Before his discharge, he became well ambulant and had gross improvement in the power of his lower extremities, the tone of the anal sphincter improved, and he was voiding well by himself. The postoperative MRI scan showed decreased conus swelling and decreased enhancement in the residual conus medullaris lesion with edema cranial to the conus medullaris completely resolved [Figure 4].

Table 1: Literature review of spinal schistosomiasis in children. Copyright 2022 by MIOT International Hospitals. Copyright 2022 by MIOT International Hospitals

Reference articles	Age (Years)	Sex	Place	Symptoms	Organism	Imaging finding	Treatment	Outcomes
Dastoli *et al.*[3]	13	Female	T4–T10	Lumbar pain and progressive lower limb weakness	*S. mansoni*	Tumefactive spinal cord	Praziquantel with corticosteroid	Partial neurological recovery under rehabilitation
Kollapen *et al.*[4]	9	Male	T10–L1	Progressive paraparesis	*Schistosoma haematobium*	Isointense intramedullary lesion	Praziquantel with corticosteroid after spinal biopsy	Improved
Al-Abdulwahhab *et al.*[2]	13	Male	Thoracolumbar	Back pain, paraparesis and paresthesia	Not isolated	Hyperintensive intramedullary lesion	Praziquantel with corticosteroids	Complete recover
Salgado *et al.*[11]	6	Male	Caudal cord	Abdominal pain, diarrhea, holocranial headache, fever, back pain paresthesia, paraparesis and neurogenic bladder	*S. mansoni*	Caudal thickening with contrast enhancement	Praziquantel with corticosteroids after lumbar puncture	Partial neurological recovery with residual paraparesis mobilises with wheelchair
Rodrigues *et al.*[9]	4	Male	T12–L2	Progressive paraparesis with fecal incontinence and urinary retention	S. masoni	Isointense conus lesion	Surgery followed by Praziquantel	Near complete neurological recovery
Labeodan and Sur[5]	10	Female	Conus medullaris	Ascending progressive paraparesis and fecal incontinence	*S. mansoni*	Isointense conus lesion enhancing with contrast	Surgery followed by Praziquantel with steroids	Residual areflexic detrusor
Lighter *et al.*[6]	14	Male	T11–L1	Bilaterally radiating low back pain	*S. mansoni*	Circumferential enlargement of caudal aspect of the spinal cord and conus with contrast enhancement	Praziquantel with corticosteroids	Improved
Saleem *et al.*[10]	7	Male	T12–L2	Bilateral paraparesis with leg pain	*S. mansoni*	Isointense lesion with contrast enhancement	Total surgical resection with Praziquantel and corticosteroids	Improved
	9	Male	T12–L1	Autonomic sphincteric dysfunction, bilateral paraparesis,	S. hematobium	Isointense lesion with contrast enhancement	Total surgical resection with Praziquantel and corticosteroids	Improved

(Contd...)

Table 1: *(Continued).*

Reference articles	Age (Years)	Sex	Place	Symptoms	Organism	Imaging finding	Treatment	Outcomes
	6	Male	T12–L3	Autonomic sphincteric dysfunction, bilateral paraparesis	Non-spined ova	Isointense lesion with contrast enhancement	Total surgical resection with Praziquantel and corticosteroids	Improved
	11	Male	T11–T12	Autonomic sphincteric dysfunction, bilateral paraparesis and low back pain	*S. mansoni*	Isointense lesion with no contrast enhancement	Total surgical resection with Praziquantel and corticosteroids	Stabilised
	15	Male	T12–L1	Autonomic sphincter dysfunction, bilateral paraparesis	Non-spined ova	Iso-hypointensity with no contrast enhancement	Total surgical resection with Praziquantel and corticosteroids	Improved
	16	Male	T11–T12	Bilateral paraparesis and low back pain	Deformed ova	Isointense and no contrast enhancement	Partial surgical resection with Praziquantel and corticosteroids	Improved
Da Paz *et al.* [8]	3	Female	Thoracolumbar	Fever, diarrhea, abdominal colicky pain, non-ambulatory, asymmetrical paraparesis	*S. mansoni*	Spinal enlargement with contrast enhancement	Oxaminiquine and corticosteroid	Partial neurological recover
	6	Male	Thoracolumbar	Leg pain, symmetrical flaccid crural paraparesis, areflexia	*S. mansoni*	Spinal enlargement with contrast enhancement	Oxaminiquine and corticosteroid	Recovery data unavailable
	12	Male	Thoracolumbar	Asymmetrical flaccid crural paraparesis, left lower limb areflexia of achilles and popliteal tendon, neurogenic bladder	*S. mansoni*	Spinal enlargement with contrast enhancement	Laminectomy with biopsy followed by oxaminiquine and corticosteroid	Residual achillean areflexia of the left deep tenson
	11	Male	Cauda equina	Urinary and fecal incontinence, flaccid asymmetrical areflexic crural paraparesis	*S. mansoni*	Spinal enlargement with contrast enhancement	Oxaminiquine and corticosteroid	Complete neurological recovery

(Contd...)

Table 1: *(Continued).*

Reference articles	Age (Years)	Sex	Place	Symptoms	Organism	Imaging finding	Treatment	Outcomes
	2.5	Male	Thoracolumbar	Neurogenic bladder, fecal incontinence, non-ambulatory, flaccid areflexic crural symmetrical paraparesis and paresthesia below D10	*S. mansoni*	Spinal enlargement with contrast enhancement	Laminectomy followed by oxaminiquine and corticosteroids	Residual sphyncteral disturbance
	14	Male	Cauda equina	Sudden onset pain and paraparesis in the left lower limb, neurogenic bladder, fecal incontinence, flaccid right crural monoparesis and achillean areflexia	*S. mansoni*	No abnormalities	Oxaminiquine and corticosteroids	Complete neurological recovery
	10	Male	Thoracolumbar	Acute onset pain and paraparesis in bilateral lower limbs, neurogenic bladder, flaccid asymmetric crural areflexia and paraparesis	*S. mansoni*	Spinal enlargement with contrast enhancement	Oxaminiquine and corticosteroid	Residual achilles areflexia

S. mansoni: *Schistosoma mansoni*

DISCUSSION

Neursochistosmiasis is a neglected and unacknowledged complication of Schistosomiasis infesting the central nervous system by the *Schistosoma* species, which can provoke severe neurological disability due to the consequence of an immune reaction.

The parasite's ovum is passed in infected individuals' urine or feces, contaminating fresh water, where ciliated miracidium is liberated and enters the intermediate host, a species of freshwater snails, where it multiplies [Figure 5]. Many fork-tailed cercariae are released into the water, which may survive for 2–3 days. These cercariae can penetrate the skin or mucous membranes of the mouth of humans and can transform into schistosomula and molt as they pass through the bloodstream.[9] They can be further carried to the lungs, heart and liver by the bloodstream, where they can mature in the portal vein. Acute infection can cause dermatitis, also known as cercarial dermatitis and Katayama fever. Within days, chronic disease can cause intestinal, hepatosplenic, pulmonary, urogenital, and neuroschistosomiasis.[8] Identifying eggs in excreta and urine, antibody detection, molecular diagnostic study, and tissue biopsy are diagnostic methods used to measure the intensity of infestation and imaging of the lesion is useful for indicating schistosomiasis infection.[5] The migration of the eggs and parasites occurs through the retrograde valveless venous flow of Batson's venous plexus located between T11 and L1, which can manifest as cerebral or neuroschistosomiasis.[8] The infestation may lead to an immunological response, leading to granuloma formation with an intense inflammatory reaction and ischemic necrosis.[8] It affects between 1% and 4% of systemic schistosomal infections. Schistosomiasis commonly manifest with symptoms such as sensory-motor deficits, ataxia, paresis and bladder dysfunction. Our extensive literature review of children with spinal

Schistosomiasis shows that this is a relatively rare condition with significant neurological morbidity, which is often permanent. The low number of documented pediatric spinal neuroschistosomiasis is probably attributed to less awareness and diagnosis of this disease.[2] Since this disease comes from a severely resource-depleted part of the world, diagnosis and initiation of appropriate management remain challenging.

LITERATURE REVIEW OF SPINAL SCHISTOSOMIASIS IN CHILDREN

As noted with the children reported in the literature, our patient also belonged to this particular endemic region and demonstrated classical clinicoradiological findings [Table 1]. The average age of neuroschistosomiasis prevalence in the literature review is 11.8 years. The prevalence and severity curves show a peak in children aged 6–15 years, and the incidence declines gradually as the age increases. The preponderance of this infection is higher in the males, as noted in all the case studies except for cases Dastoli *et al*.[3] and Da Paz *et al*.[8] The male predominance is attributed to the males' increased exposure to infected freshwater compared to the females. The most common symptom is profound paraparesis which was recorded in 95% of cases, followed by neurogenic bladder in 55% of cases, back pain in 45% of patients, areflexia, and fecal incontinence in 20% of cases, paresthesia in 15% of cases, and diarrhea and fever in 10% children. Monoparesis, abdominal pain and headache were noted in 5% of patients. Our patient presented with symptoms, such as paraparesis, neurogenic bladder, areflexia, and non-ambulation, which correlated with the literature. As noted in our patient, the most common site of neuroschistosomiasis was in the thoracolumbar region. About 55% of the patients were treated surgically in the study mentioned above for diagnostic or therapeutic purposes, and all the patients were treated medically with steroid and anti-helminthic medications. Partial recovery was noted in about 85% of the cases, while the complete recovery was reported in only 15%. This highlights the importance of prompt initiation of treatment to avoid permanent deficits. About 5% of the patients have no post-treatment prognosis data available in the literature review. About 70% of the case study in the literature review were noted to have isolated species of *S. mansoni*. About 15% were observed to have non-specified ova, and 10% were reported with *Schistosoma hematobium*. The most common species of schistosomiasis is *S. mansoni*, as noted in the literature review and our case study.

The ideal way to manage this disease would need a high index of suspicion and early institution of anti-helminthic therapy.[7] The characteristic "String of Beads" was noted in the MRI of our manuscript case study and is a characteristic feature of this infestation due to the nodular enhancement by the schistosome eggs.[11] However, the diagnosis can be easily missed due to limited resources such as MRI availability.[2] Even with characteristic MRI scan findings, schistosomiasis could not be suspected due to the lack of awareness of this disease entity, especially in a non-endemic area. Therefore, following a protocol suggested by Adeel is strongly recommended.[1] Hence, an appropriate diagnosis of this condition would obviate the need for surgery, as seen in our case.[4] Our patient had classical MRI features, which were observed retrospectively. If the patient had been started on empirical praziquantel-based therapy, the patient would not have needed surgery. Even though the deficits were severe enough to make the child completely bed-bound, the medicines and intensive rehabilitation led to significant improvement. This demonstrates that even in the presence of poor neurological grade and abysmal improvement rate, the efforts such as neurorehabilitation for a prolonged period can result in significant improvement, as demonstrated in our case. Evaluation of family members and close relatives should also be screened for asymptomatic individuals to plan for prompt and early treatment to avoid disease progression.

The available evidence and our case can further help advance the knowledge about the spinal manifestation of neuroschistosomiasis, emphasizing its early diagnosis and intervention. As observed in our case, the typical clinical manifestations and radiological findings of "String of Beads" may avoid diagnostic surgical intervention. This will reduce the time for initiation of appropriate anti-parasitic medications and steroids, leading to favorable outcomes.

CONCLUSION

This manuscript provides additional insights into the rare diagnosis of spinal neuroschistosomiasis, which is associated with significant neurological morbidity. Early diagnosis and treatment can improve neurological outcomes. In the presence of characteristic imaging findings, empirical treatment may be warranted to avoid surgical intervention and provide favorable results. This article adds to the importance of awareness of this rare entity in the list of differential diagnoses of conus lesions.

Ethics approval and consent to participate

This manuscript has been reported post approval from the ethics committee of MIOT international and consent from the child's parents.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Adeel AA. Spinal cord schistosomiasis. Sudan J Paediatr 2015;15:23-8.
2. Al-Abdulwahhab AH, Al-Sharydah AM, Al-Suhibani SS, Al-Jubran SA, Al-Hailey AK, Al-Hifzi AI, *et al*. Neuroschistosomiasis mimicking lower back pain: Case report of a rare differential diagnosis in a pediatric patient. Patient Saf Surg 2018;12:28.
3. Dastoli PA, Leite AL, da Costa MD, Nicácio JM, Pinho RS, Ferrarini MA, *et al*. Medullary neuroschistosomiasis in adolescence: Case report and literature review. Childs Nerv Syst 2021;37:2735-41.
4. Kollapen K, Suleman FE, Smuts I, Siwela L. Medullary neuroschistosomiasis in a pediatric patient: A case report. Radiol Case Rep 2021;17:462-6.
5. Labeodan OA, Sur M. Intramedullary schistosomiasis. Pediatr Neurosurg 2003;39:14-6.
6. Lighter J, Kim M, Krasinski K. Intramedullary schistosomiasis presenting in an adolescent with prolonged intermittent back pain. Pediatr Neurol 2008;39:44-7.
7. Lo NC, Bezerra FS, Colley DG, Fleming FM, Homeida M, Kabatereine N, *et al*. Review of 2022 WHO guidelines on the control and elimination of schistosomiasis. Lancet Infect Dis 2022;22:e327-35.
8. Paz JA, Valente M, Casella EB, Marques-Dias MJ. Spinal cord schistosomiasis in children: Analysis of seven cases. Arq Neuropsiquiatr 2002;60:224-30.
9. Rodrigues DB, Rodrigues AB, Queiroz JW, Braga MC, Kita WS, Netto RH, *et al*. Intramedullary spinal schistosomiasis in a child with acute myelopathy: A case report. Surg Neurol Int 2020;11:371.
10. Saleem S, Belal AI, El-Ghandour NM. Spinal cord schistosomiasis: MR imaging appearance with surgical and pathologic correlation. AJNR Am J Neuroradiol 2005;26:1646-54.
11. Salgado JV, da Silva Cruz Salgado IA, Braga Júnior LL, Serra SC, de Carvalho Barros VM, Silva MJ, *et al*. Myeloradicular form of neuroschistosomiasis in a six-year-old boy infected with *Schistosoma mansoni*. Pediatr Infect Dis J 2015;34:1409-11.

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