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Case Report

Medullary neuroschistosomiasis in a pediatric patient: a case report

Kumeshnie Kollapen, FcRad Diag(SA)^{a,*}, Farhana Ebrahim Suleman, FcRad Diag(SA)^b, Izelle Smuts, BSc, MBChB, MMed, FC(Paed)SA, PhD^c, Lebohang Siwela, MBChB^d

^a Department of Radiology, Faculty of Health Sciences, University of Pretoria, Steve Biko Academic Hospital, c/o Steve Biko and Malan Street, Private Bag X169, Pretoria, 0001, South Africa

^b Department of Radiology, Faculty of Health Sciences, University of Pretoria, Steve Biko Academic Hospital, Private Bag X20, Pretoria, 0001, South Africa

^c Department of Radiology, Faculty of Health Sciences, University of Pretoria, Steve Biko Academic Hospital, Private Bag X20, Pretoria, 0001, South Africa

^d Department of Radiology, Faculty of Health Sciences, University of Pretoria, Steve Biko Academic Hospital, Private Bag X20, Pretoria, 0001, South Africa

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ABSTRACT

A pediatric patient with neurological deficit was examined using magnetic resonance imaging (MRI). The images revealed abnormal signal intensity and enhancement of the spinal cord, indicating myelopathy. Identifying the cause of the myelopathy required a differential diagnosis. Images from MRI included a pre-contrast T1 weighted sagittal sequence, which revealed expansion of the distal lumbar spinal cord and conus medullaris from T10-L1. The T2 weighted sagittal sequence revealed patchy areas of hyperintense signal. We did not notice any chronic hemorrhagic products or cysts. Within the field of view, we saw multifocal areas of bladder wall thickening. Sagittal and axial T1 weighted post gadolinium images demonstrated mixed linear and nodular patchy enhancement of the conus medullaris predominantly anteriorly and along the anterior surface of the meninges. On the 18 day of hospitalization, a spinal biopsy revealed the presence of granuloma with non-viable bilharzia ova, and schistosomiasis of the spinal cord was diagnosed. Although uncommon, when it does occur, schistosomiasis has significant implications. Using MRI, the medical team noticed abnormal features that called for a biopsy, and were thus able to differentiate between medullary schistosomiasis and other infective/inflammatory conditions. A prompt diagnosis is vital for initiating early treatment, and avoiding complications and invasive surgery.

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Introduction

Schistosomiasis has a high incidence worldwide, and affects mostly children [1]. In 2018, the World Health Organization

* Corresponding author.

E-mail address: Kumeshnie.kollapen@yahoo.com (K. Kollapen).

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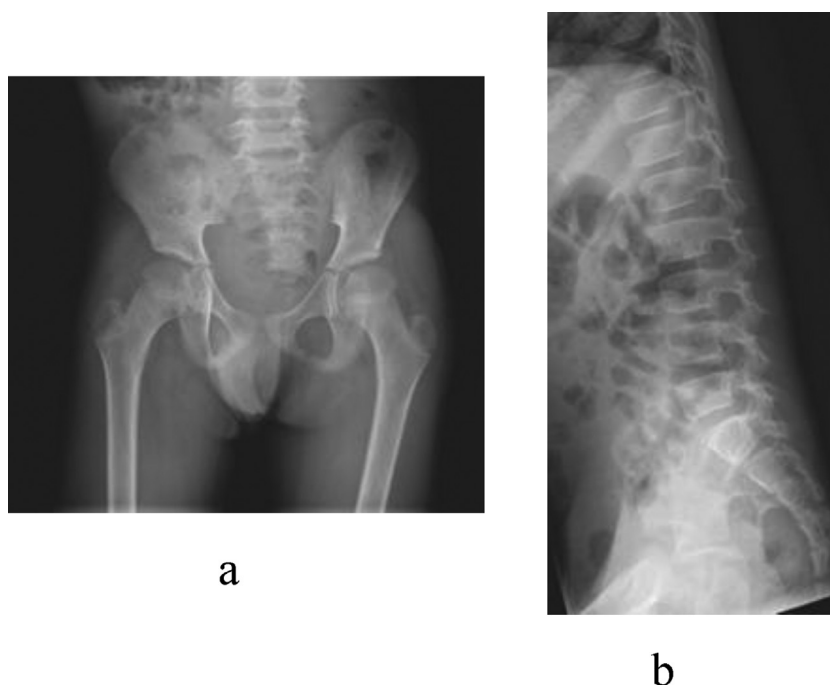


Fig 1 – A B Radiographs demonstrating a normal pelvic ring, hip joints and alignment of the lumbar spine (A). No bladder wall calcification (B)

[WHO] estimated that it is actively transmitted in 78 tropical countries with approximately 800 million people at risk worldwide [2].

Medullary schistosomiasis is an uncommon but significant manifestation of schistosomiasis outside of the urogenital tract. The pathogenesis is related to migration of ova via Batson's venous plexus. Clinically, medullary schistosomiasis has a variable presentation, ranging from severe lower back pain to acute transverse myelitis. If left untreated, the disease may progress with irreversible complications [3]. When patients present with symptoms of severe lower back pain or myelopathy, it is important to consider a differential diagnosis of medullary schistosomiasis, especially in endemic areas or if there is a history of travel to an endemic area [3]. Early diagnosis and treatment is crucial to prevent unnecessary surgery and morbidity.

In this article, we highlight the neurological manifestations of bilharzia [neuroschistosomiasis], specifically in the spine [medullary neuroschistosomiasis], and review the characteristic imaging features and differential diagnoses.

Case report

A 9 year old boy presented with progressive bilateral lower limb weakness but he was still able to walk. There was no history of lumbar pain or headache. He was referred from an area where schistosomiasis is common. On clinical examination there was reduced power [4/5] of the lower limbs. Sensation, bladder and bowel function were normal.

Initial urine microscopy, culture and sensitivity done on admission was negative for *Schistosoma haematobium* ova. A schistosomiasis ELISA done five days post admission was IgG positive. In hospital, serial biochemistry and hematology blood tests were unremarkable except for a Hb 7g/dl [normocytic, normochromic anemia]. Elevated neuron specific enolase of 31.0 μ g/L [0.00-12.5] was documented. No blood eosinophilia was demonstrated. Cerebrospinal fluid analysis and stool microscopy were not performed.

Pelvic and lumbar spine radiographs were normal. No sacral dysgenesis was noted and the pre- and paravertebral spaces were within acceptable limits. No calcifications of the bladder wall were noted (Fig. 1 A, B).

Magnetic resonance imaging [MRI] was done on the fifth day of hospitalization. The pre-contrast T1 weighted sagittal sequence revealed expansion of the distal lumbar spinal cord and conus medullaris from T10-L1. The T2 weighted sagittal sequence revealed patchy areas of hyperintense signal. No chronic hemorrhagic products or cysts were noted. Within the field of view, multifocal areas of bladder wall thickening were seen (Fig. 2 A, B).

Sagittal and axial T1 weighted post gadolinium images demonstrated mixed linear and nodular patchy enhancement of the conus medullaris predominantly anteriorly and along the anterior surface of the meninges (Fig 3. A, B). No mass like enhancement or enhancement of the cauda equina nerve roots was noted. The lower thoracic and proximal lumbar spinal cord was unremarkable. Limited imaging of the brain showed no abnormality. A spinal biopsy done eighteen days after admission revealed granulomas containing non-viable bilharzia ova (Fig. 3).

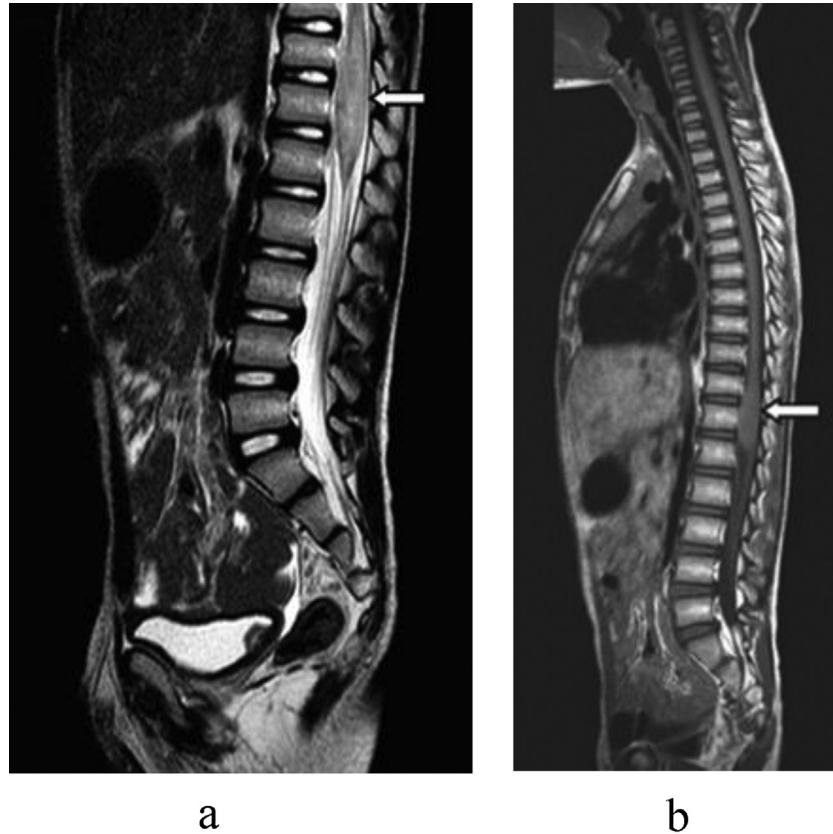


Fig 2 - A B Sagittal T1W (A) image shows moderate expansion of the conus medullaris. The lesion is isointense to the cord. Sagittal T2W (B) image shows patchy hyperintense signal. Multifocal areas of bladder wall thickening are demonstrated

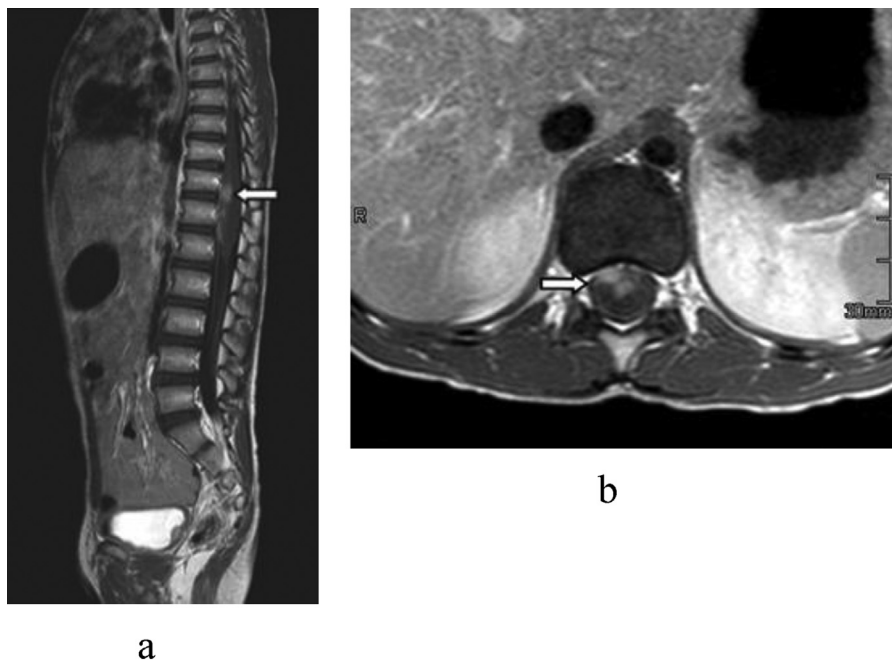


Fig 3 - A B Post contrast sagittal T1W image (A) with a mixed linear and nodular patchy pattern of enhancement predominantly in the anterior aspect of the spinal cord. No discrete enhancing mass. Post contrast axial T1W image (B) demonstrates involvement of the ventral surface of the spinal cord

Table 1 – Comparison of imaging features in the differential diagnoses of schistosomiasis of the spinal cord.

	Location	Leptomeningeal enhancement	Associations
Neoplasm	Any region	-	Increased cord distortion Peritumoral edema Cystic and hemorrhagic components
Multiple sclerosis	Any region	-	Characteristic Dawson's fingers, brainstem and cerebellar lesions
TB	Thoracolumbar	+	Gibbus deformity of the spine, epidural, subligamentous and psoas abscesses
Acute transverse myelitis	Midthoracic	-	
Schistosomiasis	Lower thoracic cord/conus medullaris	-	Less cord distortion Bladder wall thickening maybe present.

Histology of the spinal biopsy revealed sections of normal tissue with a focus of poorly formed granuloma. The lesion was characterised by aggregates of epithelioid cells. In addition non-viable bilharzia ova were also seen.

Ethical statement

Ethical clearance obtained by Faculty of Health Sciences Research Ethics Committee, University of Pretoria, Ethics reference no: 150/2021

Discussion

Incidence

Schistosomiasis of the central nervous system is uncommon. Spinal cord involvement in particular is a rare but well-documented presentation [4] affecting approximately 230 million people in 74 countries across Africa, Asia and America [5] of which approximately 123 million are children [1]. Schistosomiasis is second to malaria in terms of worldwide socio-economic impact [8].

Schistosomiasis is estimated to affect 20 million people, who have severe disease progression including involvement of the central nervous system [CNS] [5]. Early detection and treatment is crucial to preventing disability [5].

Pathogenesis

Schistosomiasis may enter the CNS via two major pathways. Adult worms may migrate and deposit eggs in the CNS or parasitic eggs may travel via either the arterial or venous system, especially the valveless peri-vertebral venous plexus of Batson [6,7]. This plexus connects the deep iliac veins and inferior vena cava with the spinal cord veins [8]. Schistosomiasis in the spine has four standard clinical forms, namely, granulomatous, myelitis, radicular or vascular [6]. The schistosomi-

asis eggs release proteolytic enzymes that induce inflammation, leading to the eventual formation of granulomas [9].

Clinical

People of any age may be affected but most people are affected in their second decade of life. Men are also more affected than women. Spinal cord schistosomiasis may present acutely or sub-acutely with lower back pain, radiculopathy and associated lower limb weakness, as with our patient, and bladder dysfunction. The medullary form which involves the cord has a rapid course with severe weakness and symmetrical fallout. This is in contrast to the conus medullaris form, which has a slower course with less severe symptoms and an asymmetrical distribution [5]. Other cerebral symptoms may be present including headache, encephalopathy and seizures [10].

Blood eosinophilia is suggestive but nonspecific of schistosomiasis [8]. Analysis of the CSF may show CSF pleocytosis, elevated protein content or oligoclonal IgG bands [8]. Stool smear examination is positive with quantitative Kato-Katz [1]. Ova in stool or urine samples provides supportive but not direct evidence of medullary neuroschistosomiasis [8]. The definitive diagnosis is tissue biopsy via surgery which demonstrates schistosome ova with surrounding necrosis and inflammation [5].

Imaging

Imaging plays a major role in detecting spinal cord schistosomiasis and helps to exclude other possible diagnoses. MRI is the modality of choice which demonstrates an inflammatory myelopathy with moderate expansion specifically of the distal cord and conus medullaris [5,11]. The myelopathy may affect variable lengths of the spinal cord and can be up to a few centimeters [8]. Signal intensity abnormalities may include isointense to cord on T1, patchy hyperintensity on T2 over several segments and no blooming on susceptibility weighted imaging [4,12].

Contrast enhancement is almost always present but variable. Saleem, Belal [4] describe three different forms of enhancement. This includes intramedullary nodular enhance-

ment, peripherally enhancing lesions, as seen in our patient, on the anterior surface of the cord and radicular enhancement of the cauda equina nerve roots [4]. The enhancement pattern is usually heterogenous [8]. A small reactive syrinx without enhancement has been documented [11]. Atrophy of the cord may be present in chronic disease [8,12]. Extrapinal findings include bladder thickening [12].

Differential diagnosis

The differential diagnosis of spinal cord schistosomiasis includes neoplastic and non-neoplastic cord lesions. Neoplastic lesions include astrocytoma, ependymoma, hemangioblastoma and metastases [11] and the latter includes transverse myelitis, infarction and demyelinating diseases i.e., multiple sclerosis [4] as well as granulomatous disease such as sarcoid and tuberculosis. A comparison of the distinguishing imaging of the differential diagnosis is provided in Table 1. The most important distinguishing imaging feature is the location [4].

Treatment

Safe and effective treatment has been achieved with Praziquantel [1,8]. Corticosteroids are used in the acute setting to inhibit the inflammatory response and to avoid further neurological complications [8]. There are no current guidelines stating the appropriate duration of corticosteroid use, which can vary from 7 days to 12 months [2]. Late diagnosis and treatment can result in severe neurological sequelae. Surgical treatment is only indicated in acute paraplegia, release of nerve roots, cord compression or if there is obstruction to CSF flow [2].

Conclusion

Medullary neuroschistosomiasis is a rare but easily treatable complication of schistosomiasis which has an excellent prognosis. It should be considered in the differential diagnosis of lesions affecting the lower thoracic cord -conus medullaris particularly in children from endemic areas who present with back pain. A high index of suspicion is required during imaging. MR imaging can demonstrate the extent of involvement, typical signal changes in the cord and enhancement patterns suggesting the diagnosis. Early detection and correct diagnosis allows for early treatment and avoids devastating neurological sequelae and disability.

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