Clinical Article

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Spinal Cord Schistosomiasis: Diagnosis, Pathological Features and Treatment.

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Abstract

Background Data: Schistosomiasis affects over 200 million people worldwide. Involvement of the CNS is a rare occurrence. Spinal cord involvement is a rare manifestation of schistosomiasis, the conus medullaris being the primary site of spinal involvement.

Purpose: We describe the clinical presentation and the MRI findings of spinal cord schistosomiasis in correlation with surgery and histopatholgical findings.

Study Design: Clinical case study.

Patients and Methods: We report four cases of spinal cord schistosomiasis presented with rapidly progressive paraparesis associated with urinary incontinence. Three patients were males and one was female (mean age 16 years). MRI of the spine demonstrated a diffusely enhancing mass at the conus medullaris with spinal cord edema, in one of the cases, the edema extended to the upper thoracic segment.

Results: In all cases, spinal masses were surgically managed through decompressive laminectomy and either biopsy or debulking. Histopathological examination showed a granulomatous inflammation surrounding bilharzial ova with a final diagnosis of spinal cord schistosomiasis. Patients were treated with Praziquantel and oral steroids. They made a remarkable neurological recovery.

Conclusions: Spinal cord schistosomiasis inspite its rare occurrence, must be considered in the differential diagnosis of conus medullaris lesions. Accurate diagnosis, through recognition of its MRI appearance, allows early treatment. Better prognosis depends on decompressive laminectomy, oral steroids to abort intense immune reaction surrounding the ova and antibilharzial drug. (2014ESJ055)

Keywords: spinal cord schistosomiasis, bilharzial ova, schistosoma mansoni

Introduction

Although infection of the CNS with schistosomiasis is rare, the involvement of the spinal cord accounts for approximately 6% of non-traumatic myelopathy in endemic areas.[12] Reports of spinal cord schistosomiasis date from 1905 when Siamura and Tsunoda[12] described the autopsy of a patient with transverse myelitis. Schistosoma, especially S mansoni, is considered as a primary cause of spinal cord parasitic invasion in Egypt.[27] Spinal schistosomiasis commonly assaults the lower thoracic/upper lumbar regions and is characterized by cauda and lower cord neurologic symptoms as well as characteristic radiological, serologic and pathological findings.[2,3]

The literature review revealed more than 500 cases of spinal cord involvement by schistosomiasis.[2] In this study, we report the MRI findings of four surgically confirmed cases of spinal cord schistosomiasis and correlate them with surgical and pathological findings.

Our objective in presenting these particular cases is to report new cases of spinal schistosomiasis, and to emphasize that a high index of suspicion should be raised in the differential diagnosis of transverse myelitis in endemic areas. This paper describes the clinical manifestations, diagnosis and management of schistosomiasis of the spinal cord in four patients.

Patients and Methods

Three patients were males and one was female (mean age, 16 years). The patients were presented with neurologic manifestations and were referred for spinal MRI. MRI of the spine demonstrated diffusely enhancing masses at the conus medullaris with extensive spinal cord edema, in one case, the edema extended to the upper thoracic segment. Laboratory investigations revealed mild peripheral eosinophilia. In all cases, spinal masses were surgically managed through decompressive laminectomy and either biopsy or debulking. Diagnosis was established by the identification of Schistosoma mansoni ova in histopathological sections in all cases.

The therapeutic regimen in current study included Praziquantel drug in four therapeutic points over one year. It is used 40 mg/kg in 3 divided doses over a day for 3 consecutive days and the drug was ingested by the same previous regimen every 4 months for at least one year. Postoperative MRI lower dorsal and lumbar spine was done in two cases after 6 months showed improvement in cord swelling.

Case Report:

A 10-year-old Sudanese female with a history of autonomic dysfunction (sphincteric disturbance) since 3 weeks followed by gradual progressive paraparesis till presented with paraparesis; grade one motor power in both legs with loss of knee and ankle reflex and deficit of all sensory modalities below L1 dermatome. A urinary catheter was necessary to relieve urine retention. No fever or systemic illness was reported. There was no previous history of bilharzial infestation. She had no history of recent vaccinations, trauma, or any medical illness. Laboratory investigations revealed a normal complete blood count (CBC) with eosinophilia. Ultrasound abdomen showed no hepatosplenomagly. MRI lower dorsal and upper lumbar spine showed an intramedullary lesion at the level of the conus medullaris. (Figure 1) MRI spine showed upper thoracic hyperintense intramedullary signal indicating cord edema.

Decompressive laminectomy with biopsy and debulking was done. Histopathology showed parenchymal bilharzial ova deposition associated with a granulomatous inflammation. (Figure 2, 3) Medical treatment with oral steroids and antibilharzial drug [Praziquantel] lead to marked improvement within 8 weeks and can walk without assistance and improvement in the sphincteric functions. Follow up MRI done 6 months post-operative showed improvement in cord swelling. (Figure 4)

Outcome was defined as “full recovery” when the patient presented with complete improvement of neurological status; as “partial recovery without functional limitations” when the patient remained with only minor deficits that did not interfere with daily activities; as “partial recovery with limitations” when the patient was left with permanent disabling weakness (not able to ambulate without help), severe urinary and/or intestinal dysfunction (retention or incontinence of feces or urine), or sensory loss; and as “no recovery” when no change in the clinical picture was observed after treatment.

Results

The clinical features in all the patients were remarkably similar. They all suffered from weakness

in the lower limbs, incontinence of urine and impaired sensation in the lower limbs. The duration of symptoms varied from 2 weeks to 2 months. There was no previous history of bilharzial infestation; none had hepatosplenomegaly or portal hypertension.

MRI images of the patients showed swelling of the spinal cord at D12 to L1 or L2 with enhancing pattern after Gadolinium contrast and one patient's MRI showed hyperintense intramedullary signal indicating cord edema up to the upper thoracic region. MRI showed nodular intramedullary and peripheral enhancement of the distal cord and conus medullaris. Peripheral enhancing lesions correlated to thickened leptomeninges infested by granulomatous thickened inflammatory cells and schistosoma eggs. (Figure 1)

Surgery was done in all cases, laminectomy and biopsy with debulking of the bilharzial mass. Histopathological studies of the specimen revealed schistosomal eggs in the spinal cord. (Figure 2) An interesting finding was negative results of bilharzial egg search in the urine and stool in all cases. The therapeutic regimen in current study included Praziquantel drug in four therapeutic points over one year. It was used as 40 mg/kg in 3 divided doses over a day for 3 consecutive days and the drug was ingested by the same previous regimen every 4 months for at least one year.

Patients responded well to surgical decompression and treatment with Praziquantel and oral steroids. All cases made neurological recovery; one case complete recovery, and three cases partial recovery without functional limitations with corresponding remarkable improvement on follow-up MRI imaging within 6 months of treatment.

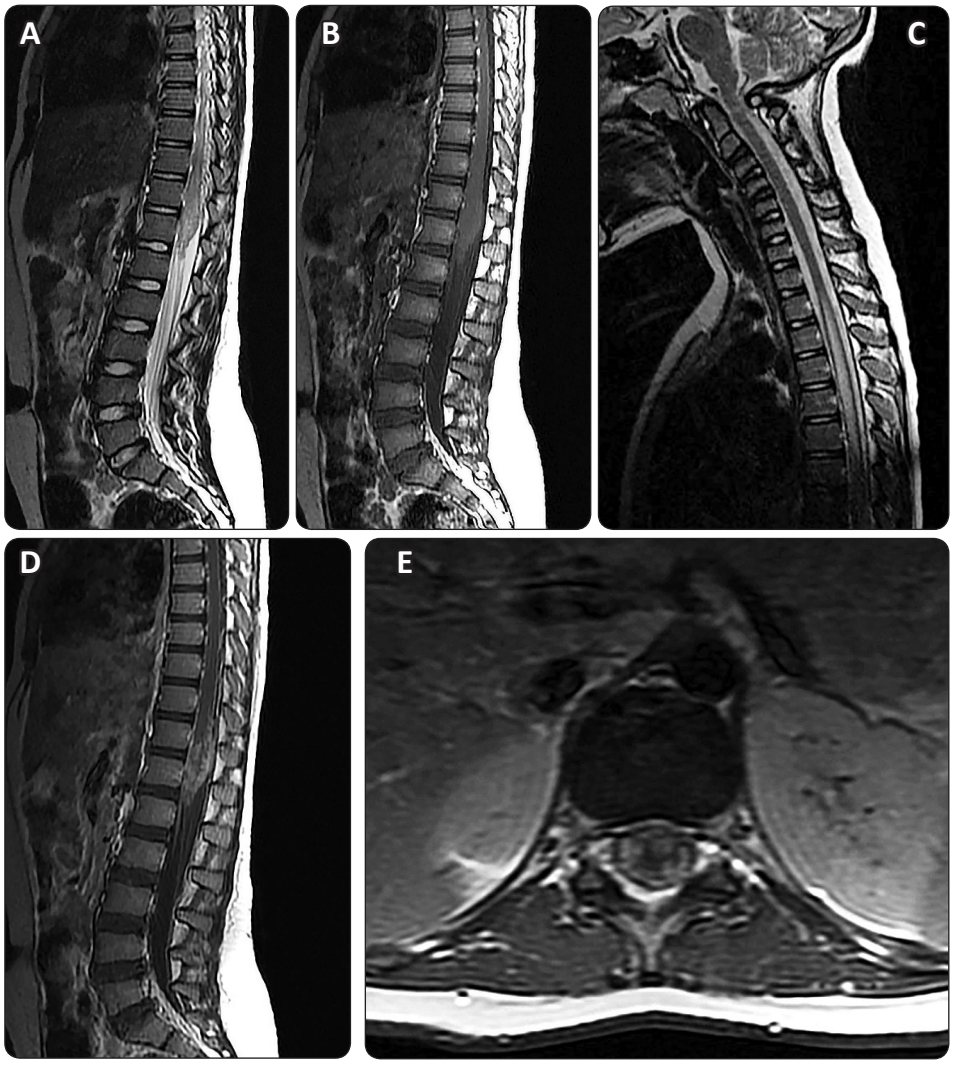

Figure 1. A,B: MRI T2, T1 weighted images showing diffuse hyper intense swelling of the conus medullaris and adjacent spinal ord. C: Sagittal T2-weighted MRI showing associated hyperintense signal involving the spinal cord extend to the whole dorsal cord area indicating associated oedema due to inflammatory reaction. D,E: T1 sagittal and axial postcontrast MRI showing Nodular intramedullary and peripheral enhancement of the distal cord and conus medullaris .Peripheral enhancing lesions correlated to thickened leptomeninges infested by chronic granulomatous inflammatory cells and schistosoma eggs.

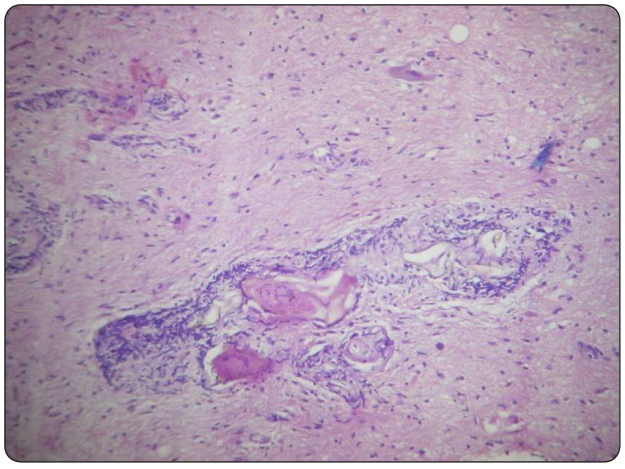

Figure 2. Microscopic examination of Spinal cord schistosomiasis reveals few fragments composed of spinal cord tissue showing focal deposition of bilharzial ova surrounded by lymphocytic infiltrate and variable amount of fibrosis. The deposition is predominantly perivascular in location. The surrounding parenchyma shows gliosis.

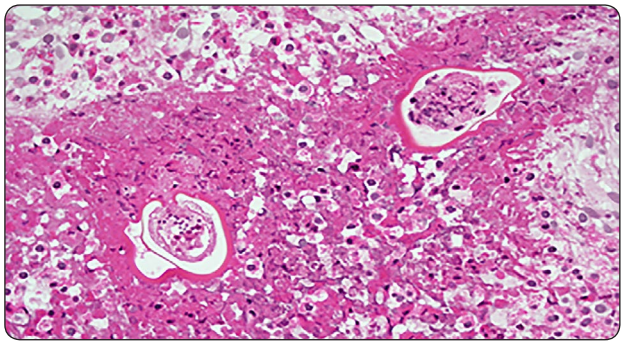

Figure 3. Light microscope of a schistosoma egg with surrounding granuloma.

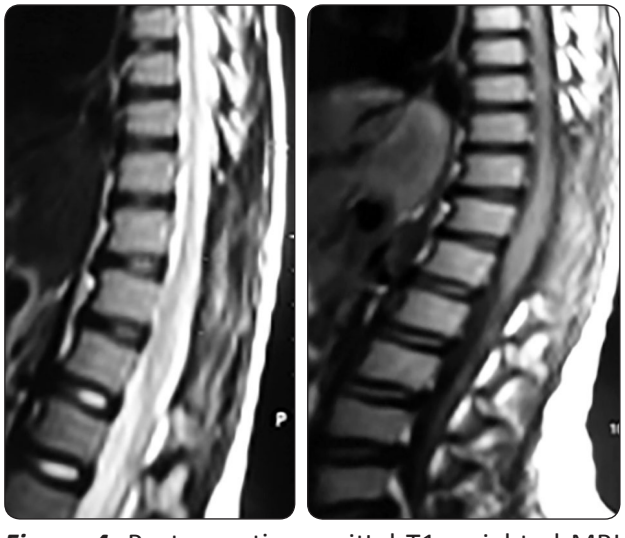

Figure 4. Postoperative sagittal T1-weighted MRI after 6 months with improvement in cord swelling.

Discussion

Schistosoma Mansoni is endemic in the Middle East (Egypt, Iraq), South America (Brazil, Venezuela), the Caribbean (Puerto Rico), sub-Saharan Africa (Mauritania, Senegal) and southern areas of Africa.[17] Schistosomiasis of the spinal cord is commonly caused by Schistosoma Mansoni although Schistosoma Haematobium has been isolated.[2,5,6] Lesions in the spinal cord are most common in the lumbar and lower thoracic parts.[20] A unique clinical picture seen in this disease, in which young patient present with lumbar pain, often of radicular nature , soon followed by rapid gradual progression of weakness of the lower limbs and associated with autonomic dysfunction.[4,5,8,20]

Spinal cord schistosomiasis due to S Mansoni may present as myelopathy, radiculopathy or both and rarely as a syndrome of anterior spinal artery occlusion. Until the clinical presentation with spinal cord symptoms, most patients have neither suggestive history nor a clinical sign of hepatosplenic schistosomiasis. Schistosomal myelopathy affects male patients in more than 80% of cases because men in rural areas are more likely to contact with infected water during childhood and in the course of their work.[22] Although different epidemiological studies have shown variable results, the mean age of presentation is 28 years for myelopathy caused by S Mansoni, while cases caused by S Haematobium present at a lower age (mean=19 years).[14]

Patients with symptoms of a rapidly progressing neurologic deficit due to myelitis or of an intrinsic spinal cord tumor and that have stayed in an endemic area, even without any past history of active intestinal or urinary tract infections,[14] should be evaluated for schistosomiasis by identifying ovae in stool or urine or rectal biopsy.[18] In our study; an interesting finding was negative results of bilharzial egg search in the urine and stool in all cases. Most patients have no clinical evidence of hepatosplenic schistosomiasis and the diagnosis is often made following spinal cord damage. Peregrino et al,[19] in an endemic area, we found only 1 case in 21 patients.

All cases in our study were of the common granulomatous type. Granuloma is the better-defined entity of the disease because it is diagnosed following operation and biopsy. Transverse myelitis is seldom proved , and its diagnosis depends on

circumstantial evidence[4,20] It is believed that the eggs reach the cord either as a result of embolization through the vertebral venous plexus of Batson or they may be deposited locally by ectopic worms in the vicinity of the cord.[16,20] Alternatively, arterial embolization of the ova through the great radicular artery could explain why conus medullaris is the primary site of spinal lesions.[5,6,11,25] We have shown in serial sections that the eggs appeared to reach the cord via the Virchow–Robin space.

Most frequently they are caused by an intramedullary granuloma of the conus mdullaris, in some cases with extension to the cauda equine. Symptoms may begin suddenly or progress rapidly in spite of ova present within the spinal cord for months or years. There is no obvious reason for the beginning of symptoms, that is usually ascribed to a immunological reaction to the ovae of a delayed hypersensitivity type with rapidly evolving tissue-destruction or mass effect.[12]

In our study, the spinal abnormalities were located in the conus medullaris and the lower thoracic cord .This very consistent localization in the lower cord and conus region is explained by the free anastomosis between pelvic veins and the valveless vertebral veins, as well as between the hemorrhoidal and systemic veins.[5,10] Our data show the good sensitivity of MRI examination of the spinal cord in SMR, previously suggested by retrospective studies. MRI of the spinal cord was important in the diagnosis of SMR, allowing identification of images compatible with infectious myelitis (described previously as spinal cord enlargement, high signal intensity in T2-weighted images, and diffuse nodular enhancement after contrast administration).[7,24]

The nodular intramedullary enhancement was linked to the pathological findings of multiple granuloma, each surrounding ova or more ova. The peripherally enhancing lesions seen in all patients represented infested meninges. It was noted in previous work.[15,20,26]

The diagnosis was made based on clinical presentation, consistent neuroimaging studies, identification of S Mansoni on a rectal biopsy or spinal cord biopsy showing granuloma. Diagnosis was established by identification of ova in histopathologic studies Schistosoma Mansoni in all cases. Intramedullary nodular enhancement was correlated to multiple schistosomiasis microtubercles. Peripheral enhancing lesions correlated to thickened leptomeninges infested by chronic granulomatous inflammatory cells and Schistosoma eggs. Linear radicular enhancement correlated with thickened resected nervous roots infested by granulomatous cells and Schistosoma eggs.[20]

The damage to the cord is attributable to the presence of the eggs and transverse myelitis as evidenced by the demyelination in the cord which was associated with a chronic inflammatory cellular infiltrate around the eggs. The Schistosomal etiology was only made after biopsy of the spinal cord lesions. This is true of most cases reported in the literature.[21,22]

The differential diagnosis of spinal cord Schistosmiasis includes neoplastic and other non-neoplastic cord lesions like transverse myelitis, multiple sclerosis, and infarction. Spinal cord Schistsomiasis shows less anatomic distortion than intramedullary tumors.[9,20,23] Multinodular intramedullary contrast enhancement of the distal cord enabled correct presumptive preoperative imaging diagnosis of spinal cord Schistosomiasis.[20]

A high index of suspicious is needed to diagnose spinal Schistosomiasis for the following reasons: a- Patients often do not have systemic involvement. b- Routine tests for detection of ova and parasites in urine and stool are often negative. c- Onset of myelopathy may be delayed by months or even years in some patients. d- Whereas histopathologic detection of Schistosoma ova and inflammatory granulomas in the spinal cord provides a definitive diagnosis, such invasive surgery is not always feasible option.[1]

Some clinicians, however, argue against the need of biopsy to establish. It has been suggested that, if the diagnosis of spinal cord Schistosomiasis can be made on the basis of clinical presentation and imaging evidences, medical therapy alone with imaging follow up would be appropriate and less invasive treatment[26] Other clinicians believe as do we , that most of the patients with spinal cord Schistosomiasis will benefit from surgery by partial resection and laminectomy, with the additional use of anti-Schistosomal drugs and oral steroids.[8,13,22] Carod et al,[1] observed full neurological recovery in 30% of patients affected by spinal Schistosomiasis. The favorable outcome in this study was attributed to early surgical intervention and medical treatment before irreversible damage occurred.

The therapeutic regimen in current study included

Praziquantel drug in four therapeutic points over one year. It is used 40 mg/kg in 3 divided doses over a day for 3 consecutive days and the drug was ingested by the same previous regimen every 4 months for at least one year. Praziquantel is a broad-spectrum Schistosomacidal drug with parasitological cure in 70-90% of patients, and is considered the drug of choice for all forms of Schistosomal myelopathy. Improvement in motor, sensory, and sphincter function usually occur within 6 weeks following Praziquantel treatment.[2] Although controversial, corticosteroids could have a role in patients with severe pathology to mitigate inflammatory responses participating in granuloma formation. Schistosomiasis of the spinal cord is an uncommon but potentially curable form of Schistosomiasis, if diagnosed and managed early.

Conclusion

We emphasize that a high index of suspicion should be raised in the differential diagnosis of transverse myelitis in endemic areas. Spinal cord Schistosomiasis is rare but an important and potentially treatable cause of myelopathy. It should be considered in any patient who lives in endemic areas especially if peripheral eosinophilia is present and ova are identified in stool, urine, or rectal biopsy. The role of surgery is decompression, particularly in large lesions and to provide material for pathological diagnosis. This should then be followed by anti-Schistosomal drugs and oral steroids.

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الملخص العربي

بلهارسيا الحبل الشوكي: طرق التشخيص و العوامل الباثولوجية والعلاج

يصيب مرض البلهارسيا حوالي ٢٠٠ مليون شخص حول العالم وإن تأثر الجهاز العصبي هو أمر نادر الحدوث وتحدث إصابة البلهارسيا بالحبل الشوكي في الجزء المخروطي.

أهداف البحث: تهدف الدراسة إلى توضيح الأعراض الإكلينيكية ونتائج التصوير بالرنين المغناطيسي وتحليل الأنسجة واهميه التدخل الجراحي.

شملت الدراسة علي أربعه حالا ت مصابة ببلهارسيا الحبل الشوكي وقد حدث تدهور سريع للحالة العصبية مما أدى إلى ضعف شديد في الأطراف السفلية مع عدم القدرة على التحكم في التبول. وكان يوجد ثلاثة ذكور وأنثى واحدة. وأظهرت أشعة الرنين المغناطيسي وجود كتلة داخل النخاع الشوكي مع حدوث إرتشاح في أنسجة الجهاز العصبي.

النتائج: أجريت الجراحة لكل الحالات مع تحليل عينة الأنسجة من الحبل الشوكي وأظهرت وجود بويضات البلهارسيا وبعد العملية تم إعطاء المرضي عقار البرازكوانتيل مع الكورتيزون مما أدى التحسن في الحالة العصبية.

الخلاصة: يتطلب تشخيص مرض بلهارسيا الحبل الشوكي دراسة دقيقة للرنين المغناطيسي وتتضمن الخطة العلاجية تدخل جراحي مع إعطاء المرضي عقار البرازكوانتيل مع الكورتيزون.