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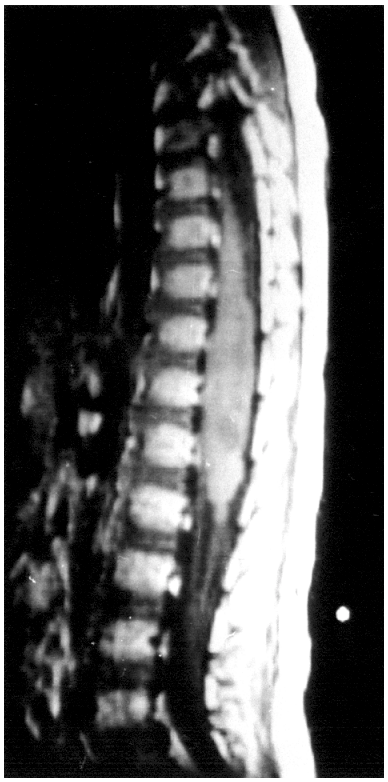

Fig. 1 Unenhanced mid- sagittal spin echo T1-weighted MR image of the thoracolumbar spine.

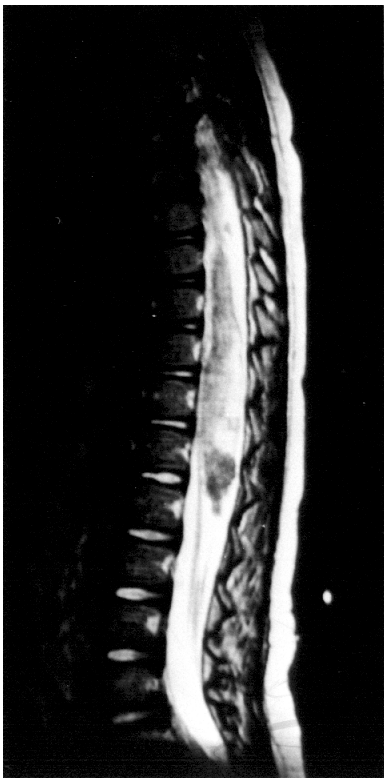

Fig. 2 Unenhanced mid-sagittal fast spin echo T2-weighted MR image of the thoracolumbar spine.

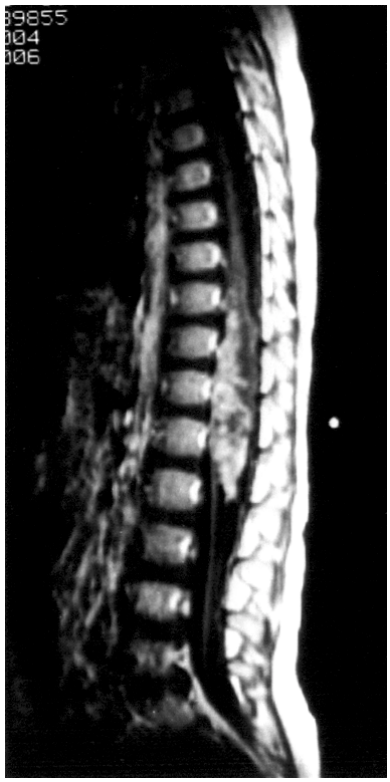

Fig. 3 Enhanced mid- sagittal spin echo T1-weighted MR image of the thoracolumbar spine.

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CASE PRESENTATION

A 2-year-old Brazilian boy presented with leg weakness and constipation. He then developed progressive paraparesis and bladder dysfunction. On neurological examination, he was found to have a flaccid paraparesis. The lower deep tendon reflexes were absent. Haematological laboratory tests showed eosinophilia. Cerebrospinal fluid (CSF) analysis revealed 50 cells/mm^3 with 4 eosinophils/mm^3, and a protein level of 106 mg/dl. Magnetic resonance (MR) imaging of the spine was performed. What do the MR scans show (Figs. 1-3)? What is the diagnosis?

IMAGE INTERPRETATION

MR imaging showed a heterogeneous fusiform intramedullary mass that extended from T9 to L1 levels. The mass caused marked expansion of the cord, and had a cystic component that was hypointense on T1-weighted images (Fig. 1) and hyperintense on T2-weighted images (Fig. 2). The solid component of the mass was T1- and T2-isointense relative to the normal spinal cord. Following intravenous (IV) gadolinium-DTPA administration, there was intense enhancement of the solid component of the mass (Fig. 3).

DIAGNOSIS

Spinal cord schistosomiasis.

CLINICAL COURSE

The stool tests of this patient revealed the presence of *Schistosoma mansoni* eggs.

The patient underwent surgery during which a T8 to L3 laminectomy was performed. The spinal cord mass was partially resected. Histolopathological examination showed a granulomatous inflammatory process and the presence of *Schistosoma mansoni* eggs. The final diagnosis was spinal cord schistosomiasis. After the surgery the patient was treated with praziquantel (three doses of 20mg/kg/day for 1 week) and oxamniquine (20mg/kg, twice daily for 1 day). The patient was discharged with partial improvement of his lower extremity strength.

DISCUSSION

Schistosomiasis is a parasitic infestation that affects an estimated 200 to 300 million people worldwide[1]. Three major species of schistosoma are known to affect man, namely: *Schistosoma mansoni*, *Schistosoma haematobium and Schistosoma japonicum*. *Schistosoma mansoni* is endemic in South America, the Caribbean region, the Middle East, and Africa. *S. haemotobium* is found in the Middle East and North Africa, and *S. japonicum* is endemic in Japan, China, and the Philippines[2,3]. In South America, Brazil is an endemic region for *Schistosoma mansoni*[4].

The schistosomas (cercaria) are initially released from host snails. These cercaria are then liberated in the fresh water by the snails and enter humans directly through the skin. A typical "swimmer's itch" may develop at the site of penetration within 24 hours of the infestation. After penetration of the skin, the larvae migrate through blood vessels and lymphatics to the lung and liver, where they mature. Their final human habitat is the venous system of the rectum, intestines (*S. mansoni*), and urinary bladder (*S. haematobium*). They mature to adult worms in 4-8 weeks and then begin to lay eggs. Typically, their eggs lodge in the intestinal wall, urinary bladder wall or liver, afterwhich they are expelled in the faeces and urine. Dissemination of ova to ectopic locations may occur via the venous system to ultimately infest the liver, ureter, bladder, lungs and central nervous system[2,5].

Spinal involvement is a rare complication of *Schistosoma mansoni* or *S. haematobium* infestation[3,4,6-9]. The prevalence of spinal cord schistosomiasis in patients with systemic disease ranges from 0.3% to 13% in different published studies[4,6]. It is believed that the ova migrate to the spinal cord by way of the valveless perivertebral venous plexus of Batson[1,3,6,9-11]. The spinal form of the disease may be either asymptomatic or can cause lumbosacral pain, paraplegia, bowel and bladder sphincter dysfunction, and sensory disturbances[10-11]. The majority of patients diagnosed with spinal schistosomiasis present concomitantly with the intestinal form of the disease[11].

The pathological findings of spinal schistosomiasis include a granulomatous intramedullary mass of the caudal spinal cord, radicular involvement with granulomatous changes surrounding the conus medullaris and nerve roots of the cauda equina, granulomatous necrosis and haemorrhage, and asymptomatic deposition of ova in the spinal cord[12]. The granulomatous reaction of the host to the presence of the ova is the major factor in the pathogenesis of schistosomiasis[8]. Four pathological patterns have been identified in association with spinal schistosomiasis, namely: extra-axial mass formation with medullary compression, granulomatous caudal equina root involvement, acute transverse myelitis, and inflammatory spinal artery occlusion (i.e. arteritis) with spinal cord infarction[8].

The most common gross finding of spinal schistosomiasis is an intramedullary mass[7]. The diffuse or localized granulomatous lesions primarily affect the conus medullaris or cauda equina[3,5]. Patients may present with the spinal form of schistosomiasis weeks to years after the initial infection. The clinical syndrome is usually that of an acute or subacute myelopathy, with or without polyradiculitis. At initial presentation of spinal schistosomiasis, specific ova may or may not be found in the patient's urine or stool samples. A nonspecific eosinophilia can be detected in the peripheral blood and CSF. The ELISA or RIA methods are reliable tests of the serum and CSF for the detection of antibodies against *Schistosoma* organisms[3,6,9,10].

Conventional water-soluble contrast myelography, computed tomography (CT) and CT-myelography may demonstrate enlargement of the conus medullaris or a complete CSF block in the lower thoracic or lumbar level. In some cases of transverse myelitis,

conventional myelography, CT and CT-myelography are normal. Affected nerve roots of the cauda equine may show irregular thickening and matting[1,3,4,6,8,9]. The MR findings in advanced spinal cord schistosomiasis include an intramedullary mass that is typically hyperintense on T2-weighted images, and produces heterogeneous or homogeneous enhancement following IV gadolinium- DTPA administration[1,3,4,5,7,9,10,13]. The cauda equina may also show diffuse enhancement[8].

The differential diagnosis thus includes primary and secondary tumours, and spinal cord myelitis of a different nature, such as bacterial and acute disseminated encephalomyelitis. Although treatment with antiparasitic drugs can be effective and curative, and surgery can be avoided with a correct pre-operative diagnosis, a specific diagnosis of spinal cord schistosomiasis can be difficult based on imaging features alone[6-8]. Praziquantel is the treatment of choice for eradication of the adult worms of *S. mansoni* and to prevent the release of further ova[3,5,6]. Oxamniquine can be used simultaneously for adjunctive treatment[5]. The supplemental use of corticosteroids is however more controversial[9].

In conclusion, the index of suspicion for spinal cord schistosomiasis should be high in patients who have had a recent history of travel to endemic areas. This history should be considered together with the known clinicoradiological constellation of findings, coupled with the laboratory data. The MR diagnosis of systemic schistosomiasis with spinal involvement should enable prompt and appropriate treatment[3,6-8].

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ABSTRACT

A 2-year-old Brazilian boy presented with bilateral leg weakness and constipation, followed by development of progressive paraparesis and bladder dysfunction. Neurological examination revealed flaccid paraparesis. Blood tests and CSF analysis showed eosinophilia. The MR examination revealed a spinal cord mass extending from T9 to L1 levels, with a heterogeneously-enhancing solid component and a cystic component. Stool tests for *Schistosoma mansoni* eggs were positive. The patient underwent surgery, the intramedullary mass was partially resected, and the diagnosis of spinal cord infection by *Schistosoma mansoni* was confirmed. After surgery, the patient was treated with praziquantel and oxamniquine. He was discharged with partial improvement of the lower extremity weakness and bowel/bladder function. The clinical and imaging features of spinal cord schistosomiasis are reviewed.

Keywords: Spinal cord, parasitic infections, schistosomiasis, *Schistosoma mansoni*, Magnetic resonance imaging

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