Schistosomiasis of the spinal cord presenting as progressive myelopathy

Case report


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The authors report on a case of schistosomiasis of the spinal cord in an individual returning to Ireland after a 25-year residence in Africa, where the infection affects approximately 200 million people.

Key Words • schistosomiasis • central nervous system • transverse myelitis • granuloma

Schistosomiasis (also known as bilharziasis) is an infection caused by a trematode platyhelminth (flat worms) of the genus Schistosoma. It is endemic in several areas of Africa and South America, affecting approximately 200 million people. It is uncommon for schistosomes to reach the CNS. The presence of ova in the CNS may give rise to seizures if located in the cerebrum or myelopathy if located in the spinal cord. Since schistosome-induced myelopathy was first reported in Africa in the 1930s, approximately 30 tissue-proven and 28 presumed cases have been reported. A similar number of asymptomatic cases have been discovered at autopsy. Most of these cases occurred in endemic areas. We report a case of spinal cord schistosomiasis in Ireland, describing its pathogenesis, clinical manifestations, and management.

Case Report

Presentation. This 62-year-old man returned from Africa where he had been residing for 25 years. He presented with a history of a rapidly progressing lower-limb myelopathy. His symptoms had begun 1 month prior to presentation when he noticed a tingling sensation in his legs, which progressed to numbness. Both lower limbs became ataxic. He lost anal sensation and experienced difficulty initiating and stopping micturition. A course of steroid therapy was instituted based on symptoms that were consistent with some form of transverse myelitis.

Examination. Thoracic MR imaging demonstrated a diffuse patchy signal change in the lower thoracic cord extending over several segments and associated with spinal cord swelling (Fig. 1 left). Tests for human immunodeficiency virus, Mantoux pits, and venereal disease were negative. Examination of CSF samples showed values within normal range, but it was not assessed for schistosomiasis antibodies at that time. Serum schistosomal ELISA was positive at level 5. The coincidence of an unexplained lower cord lesion and evidence of Schistosoma led to a presumed diagnosis of spinal cord schistosomiasis.

Treatment. Treatment with praziquantel was initiated and steroid therapy was continued. Examination of CSF was repeated and schistosomal ELISA was positive. Repeated thoracic MR imaging performed 1 month after the single course of praziquantel revealed reduced signal change and swelling of the spinal cord (Fig. 1 right). An ELISA showed decrease of the serum schistosomal titre to 4 and later to 1.

Posttreatment Course. The patient underwent follow up for an 8-month period, during which there was gradual clinical improvement. Power in both lower limbs increased in strength, ataxia decreased, and lower-extremity sensation also improved bilaterally. The patient ambulated independently. Although marginally better, anal sensation was still diminished and micturition difficulty also persisted.

Discussion

Origin and Pathogenesis of Schistosomiasis

Schistosomes are blood flukes that use humans and other mammals as definitive hosts and aquatic snails as intermediate hosts. Of the schistosomal species, Schisto-
S. mansoni, S. haematobium, and S. japonicum are the most significant to man and the most widely distributed.

The early forms of the trematode life cycle are called cercariae. They are liberated by a freshwater host snail and then infect humans percutaneously causing "swimmer's itch." This often occurs within 24 hours of infection and causes a red pruritic lesion at the site of infection.1,2,11,13

The cercariae migrate to the lungs and liver and then S. mansoni and S. japonicum inhabit portal veins, whereas S. haematobium settles in the urinary bladder veins. In 4 to 8 weeks the cercariae mature to adult worms and begin to lay eggs. At this stage, there may be an acute onset of fever, chills, sweating, cough, diarrhea, hepatosplenomegaly, bronchospasm, and eosinophilia due to an allergic reaction to the egg antigen. Most eggs are passed out in the stool but some become lodged in the intestine and bladder.1,4,9,11

Schistosomes, like many other helminths, can produce eggs for many years in the human body and cause disease long after the host has left the endemic area. Adult worms may live for 20 to 30 years during which cell-mediated immune responses lead to granuloma formation and fibrosis in organs. These granulomatous masses are called bilharzioma.9,11 In the liver the granulomas and fibrosis occur principally in a perisinusoidal distribution.

The small round eggs of S. japonicum travel all the way to the brain. The eggs of S. mansoni and S. haematobium are large and bear protruding spines. This is thought to be why S. mansoni and S. haematobium lodge in the spinal cord and not the brain. Spinal cord schistosomiasis is a relatively rare manifestation of chronic schistosomiasis. Schistosome-induced myelopathy results from the inflammatory reaction accompanying the deposition of ova in the venules located in and around the spinal cord. The ova, which normally are deposited in the inferior mesenteric vein of the portal system, reach the spinal veins via the valve-free venous system connecting the intraabdominal and spinal veins, known as Batson plexus. Because this system links only the drainage of the lower spinal cord, it would certainly explain the predilection of myelitis for the lower spinal cord and conus medullaris levels. Anomalous migration of adult worms may also cause elimination of eggs directly within these spinal vessels, accounting for the occasional finding of adult worms and eggs in a row inside vertebral vessels. The extent of the lesions depends on the degree of infestation and the host immunological response. The interval between the supposed infestation and the onset of the spinal cord presentation varies from several days up to 6 years.5,6,8,12

Clinical Picture

In most cases of schistosomal CNS involvement, patients are asymptomatic. The most frequent manifestation of spinal cord schistosomiasis is acute or subacute transverse myelitis. Urinary retention, back pain, and lower-extremity motor and sensory disturbance are common symptoms. Granuloma is less common and is usually at the conus medullaris level. Schistosoma mansoni and S. haematobium have also been reported to cause anterior spinal artery occlusion and multiple nerve root syndrome with multiple granulomas of spinal roots.3,2,4

Diagnosis of Infection

Schistosoma haematobium ova are found in the urine in...
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25% of cases. In 60% of the patients with S. mansoni, eggs are identified in the feces; however, if the stool culture is negative an examination of a rectal biopsy sample is indicated (M. Wilson, personal communication). Eosinophilia may be seen in the peripheral blood. Examination of CSF may show mild pleocytosis and an increased protein level. In our case all these tests were negative.

Diagnosis can also be established if antibodies against schistosomes are detected; these were positive in our patient.

Treatment of Schistosomiasis

The treatment of spinal cord schistosomiasis relies on schistosomicides. Praziquantel or oxamnique are the antischistosomal agents recommended to treat schistosome-induced transverse myelitis. These agents destroy the adult worms and thereby prevent further oviposition. In up to 80% of cases a cure is established following one course of treatment. Corticosteroid agents counter the granulomatous inflammation. Treatment in our case was based on the association of antischistosomal drugs and corticosteroid medication. Nobre, et al., reported that 22% of their patients exhibited a full response to treatment, 57% a partial response without functional limitations, and 17% partial improvement with limitations or no response to treatment. They also reported that because three of the four patients who ceased steroid treatment before 45 days experienced recurrence of myelopathy, they recommended that steroid treatment should be continued for months after clinical improvement.

If there is no clinical improvement after institution of antischistosomal medications or if neurological deterioration occurs, surgical decompression and biopsy sampling may be indicated. This was not required in our case because there was no spinal cord compression and the patient improved after medical treatment was initiated.

Conclusions

Spinal cord schistosomiasis should be included in the differential diagnosis of unexplained myelopathy in patients who traveled to geographical areas in which it is endemic.

When a biopsy specimen is not obtained, a presumptive diagnosis of spinal cord schistosomiasis is based on the following considerations: 1) the finding of low thoracic/upper lumbar neurological symptoms; 2) demonstration of exposure to schistosomes by using parasitological or serological techniques; 3) the exclusion of other known causes of transverse myelitis; and 4) response to antibilharzial medications. The presence of eosinophilic pleocytosis of serum and CSF is suggestive of schistosomiasis of the CNS but is often not observed, as in our case.

Given the potential long-term implications of failure to recognize and adequately treat this rare neurological pathological entity, we would advocate the presumptive treatment of patients with diagnoses of transverse myelitis and histories of water exposure in endemic areas while awaiting definitive serological results. Early diagnosis is essential because of the excellent prognosis on initiation of a specific therapy.

References


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