

Acute toxemic schistosomiasis complicated by acute flaccid paraplegia due to schistosomal myeloradiculopathy in Sudan

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ABSTRACT

يشكو طبيب سوداني يبلغ من العمر 55 عاماً، من مرض استمر لمدة شهر أعراضه كالتالي: إسهال مع ارتفاع في درجة الحرارة ليلاً، فقد الوزن (10Kg). بعد إجراء منظار للقولون اشتكى المريض من ضعف حاد في الأطراف السفلية واحتباس في البول. تم إدخال المريض للمستشفى وأجرى له فحص بالرنين مغناطيسي للنخاع الشوكي والذي أوضح وجود التهاب حاد في الجزء الأسفل من النخاع الشوكي. من خلال تقرير الحالة أعطينا تفاصيل التحديات التشخيصية والعلاجية التي واجهناها أثناء التعامل مع هذه الحالة، وقمنا بعمل بحث معلومات عن التشخيص النهائي لها وهو: حالة بلهارسيا معوية حادة مع شلل حاد للأطراف السفلية ناتج عن بلهارسيا النخاع الشوكي والأعصاب. لقد قدمنا تقرير عن هذه الحالة بهدف تعريف وتوعية زملائنا الأطباء لتشخيص بلهارسيا النخاع الشوكي والأعصاب لأنه يحتاج لعلاج عاجل ومحدد وهو دواء البرازاكانتيل بالإضافة إلى مثايل بردنيزلون يليه بردنيزلون عن طريق الفم. كما أننا وجدنا إن إجراء فحص رنين مغناطيسي للنخاع الشوكي بعد أسبوعين من بداية بروتوكول العلاج المذكور قد يساعد في تجنب إجراء جراحة للنخاع الشوكي. تبرز الحالة إمكانية الإصابة بالبلهارسيا على ضفاف النيل الأبيض في الأماكن الحضرية وأهمية استخدام الكمبيوتر لتشخيص الحالات الإكلينيكية الصعبة.

A 55-year old Sudanese physician presented with one month history of diarrhea, loss of weight (10kg) and low grade nocturnal fever. Following colonoscopy, he rapidly developed paraparesis and retention of urine. Magnetic resonance imaging (MRI) of the spinal cord showed low cord lesion suggestive of transverse myelitis. We present a detailed account of diagnostic and management challenges and a literature review of the final diagnosis of acute toxemic schistosomiasis, complicated by acute flaccid paraplegia due to schistosomal myeloradiculopathy. We are reporting this case to increase the awareness of physicians of schistosomal myeloradiculopathy, as it needs urgent specific treatment (praziquantel and steroids). An early follow-up with MRI of the spinal cord (2 weeks treatment) may help in preventing unnecessary neurosurgical intervention. Bilharziasis

may be contracted on the banks of the river "White Nile" in urban areas. Finally, clinicians should make use of the Google computer search for diagnosis in difficult cases.

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Schistosomiasis is one of the most common parasitic infections with around 200 million affected subjects with 600 million exposed worldwide.¹ Neuroschistosomiasis due to *Schistosoma mansoni* can present in many ways: a) cerebral or cerebellar lesions (focal central nervous system [CNS] impairment, seizures, or signs of raised intracranial pressure), b) unique spinal cord pathology of *Schistosoma mansoni* CNS involvement namely, myeloradiculopathy of lower spinal cords (conus medullaris), and quada equina presenting clinically as transverse myelitis and radiculopathy. In either case, granulomatous inflammatory reaction occurs as a result of *Schistosoma mansoni* eggs carried via the vascular system to the spinal cord or the brain, or rarely by anomalous migration of adult worms to those organs.²⁻⁴ We are reporting this case to increase the awareness of our colleagues to this rare clinical entity that calls for urgent specific therapy. In addition, we saw 2 unusual features in this case, *Schistosoma mansoni* adult worms in the inflammatory polyp histopathology, and the coexistence of clinical features of acute toxemic schistosomiasis, and schistosomal myeloradiculopathy, which created great diagnostic and management difficulties.

Case Report. A 55-year old Sudanese physician presented with one month history of diarrhea, loss of weight (10 kg), and low grade nocturnal fever without sweating. The stools produced were of no special character. His past medical history and family medical history were not remarkable. On day one, upon arrival at the Emergency Department, the general examination of the patient was not remarkable. His biochemistry, hemogram, chest x-ray, and electrocardiogram were normal. We took blood sample for thyroid function test, and human immunodeficiency virus (HIV) serology. We booked the patient for colonoscopy on an out-patient basis. When the patient came for the colonoscopy examination on day 2, he needed assistance in order to move. At colonoscopy, colonic mucosa was normal up to the splenic flexure, where an inflammatory process of the whole mucosa without skip lesions was seen with erythema, congestion, and some superficial ulceration, extending down to the anus which was spared. A pedunculated inflammatory polyp was removed by snare. On day 3, the patient started to experience difficulty in voiding urine. He returned to the hospital and by inserting Foleys catheter 1200 cc of urine was recovered. Neurological examination was normal with positive signs in the lower limbs as paraparesis, which was asymmetric; left > right with weakness more distally, with grade of power as zero/5 at left ankle and 1-2/5 at right ankle, 3/5 at both knees, and 4-5/5 at both hips. The deep tendon reflexes were absent and plantar reflexes were equivocal. Sensory examination showed a doubtful sensory loss to pain and touch up to the mid left leg. The neurologist admitted the patient with a provisional diagnosis of transverse myelitis and ordered urgent MRI of the spinal cord. Biochemical and cytomorphological examination of the cerebrospinal fluid (CSF) was normal. Over a period of 4 days, the MRI of the spinal cord showed swollen lower cord (conus) with high intensity signal reported as suggestive of transverse myelitis **Figure 1a**. The histopathology of the biopsies carried out on colonoscopy reported *Schistosoma mansoni* eggs, (**Figure 2, arrow A**) with granulomatous reaction and caseating granuloma highly suggestive of tuberculosis (**Figure 2, arrow B**), which was negative for Ziehl-Neelsen stain carried out later on. Adults worms were seen in the histopathology of inflammatory polyp (**Figure 3**). In disbelief, we carried out differential leucocyte count (showed 17% eosinophils) and serology for anti bilharzial antibodies (was positive at a high titre of 1:512). Other blood tests reported normal thyroid function tests, and a negative serology for HIV infection. The neurologist started him on intravenous methylprednisolone (IVMP) one gram daily for 3 days as transverse myelitis. The internist decided to start the patient on a 4-drug anti-TB regimen

with Vitamin B6. At that stage both the patient and the treating team could not understand how he contracted *Schistosoma mansoni* infection. He received the first dose of Praziquantel (50mg/Kg) on the 8th day. In 2-3 weeks, the neurological signs did not improve after 3 days of IVMP, on the contrary there was progressively worsening weakness of the lower limbs with sensory loss in ascending fashion reminiscent of Landry's ascending paralysis of Guillain-Barré syndrome. The neurologist decided to start him on intravenous immunoglobulin (IV IgG) 30 grams per day for 5 days. However, the neurological deterioration continued in a relentless

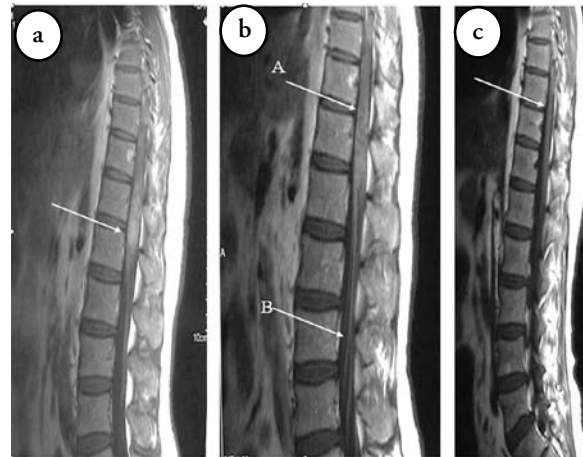


Figure 1 - Magnetic resonance imaging study of the spinal cord a) the arrow points to the high signal abnormality at the time of development of flaccid paraplegia with sensory level at the groin, b) early follow-up study (after 2 weeks of high dose steroids therapy) showing the reduction of the swelling of the speckled appearance of the cord (arrow A), and the thickened cauda equina (arrow B). c) Follow-up study after 2 months of high dose steroid therapy, showing a normal spinal cord.

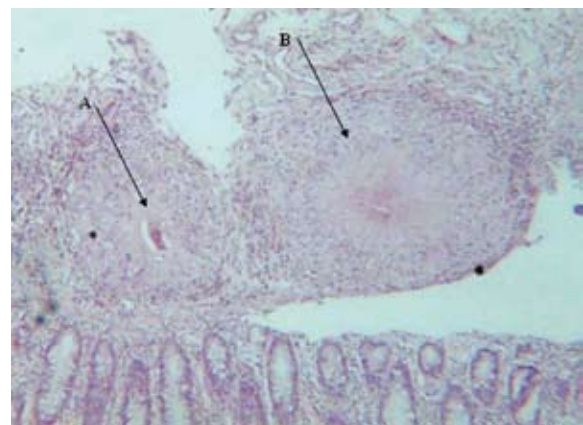


Figure 2 - *Schistosoma mansoni* egg with surrounding granulomatous reaction (arrow A) and a caseating granuloma with 'Giant' cell highly suggestive of tuberculosis (arrow B) - (? probably the section through bilharzial micro-tubercle missed the *Schistosoma mansoni* egg) .

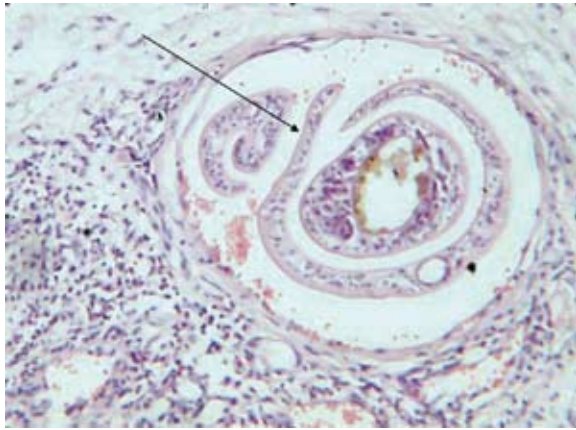


Figure 3 - *Schistosoma mansoni* worm seen in the inflammatory polyp sections.

fashion in spite of IV IgG, by the end of the 2nd week. The patient had flaccid paraplegia with zero power, and crisp sensory level at the groins. At this point of the evolution of this case, he recalled his recent picnic with his family on the White Nile at Khartoum 2 months before the onset of his illness. At this stage, we conducted a Google computer search for diagnosis of this case,⁵ and we came out with diagnosis of myeloradiculopathy of *Schistosoma mansoni* as a very likely diagnosis. We resumed steroids as IVMP one gram per day for 3 days followed by prednisolone 80 mg/day and 2 extra doses of praziquantel 50mg/Kg/day for 2 days. Praziquantel is the drug of choice for all forms of schistosomiasis ovoidal, as it has immunosuppressive, and anti-inflammatory actions.⁶

After the fifth to sixth weeks, we started to doubt the diagnosis of tuberculosis (TB) of the bowel. Since lack of any clinical improvement in spite of treatment the opinion of the neurosurgeons was sought. They raised the chance of a space-occupying intramedullary lesion (ependymoma or astrocytoma) on reviewing the MRI film (Figure 1a). The neurosurgeon asked for consent for urgent surgery to explore for a resectable intramedullary space-occupying lesion or biopsy of the lesion if resection is not useful. Silva et al⁷ stressed the fact that the improvement is delayed and high dose steroids and treatment for 2-6 months may be needed for the treatment of Schistosomal myeloradiculopathy. Under the circumstance, we decided to do a rather early follow-up MRI of spinal cord, after 2 weeks of high steroids therapy. To our relief there was definite improvement with less swelling and speckled appearance of the spinal cord (Figure 1b, arrow A) and thickened quoda equina (Figure 1b, arrow B), which the radiologist reported as an inflammatory process. The neurosurgeons were then convinced that there was no need for surgical

intervention. We decided to discontinue anti-TB medication after completing 2 months. The patient was transferred to the Rehabilitation Hospital.

After the sixth to twelfth weeks, the mainstay of the treatment at this stage was intensive physiotherapy. It was only after 4 weeks of 80 mg of Prednisolone per day that the patient started to experience clinical improvement and the muscles around the pelvic girdle are regaining some tone and are becoming less floppy. The muscle contraction started in the quadriceps. Deep tendon reflexes remained absent. Follow-up MRI of the spinal cord was carried out, after completion of 2 months of high steroids therapy, and showed a normal spinal cord (Figure 1c). The patient left for England, United Kingdom for a second opinion and after 4 months he returned back home. When he came back to Sudan, there was little improvement, with no muscle wasting, and he is in a wheelchair with intermittent catheterization, and he could not go back to work.

Discussion. We aimed to give a detailed and honest account of the diagnostic and management challenges, we encountered in dealing with this extremely interesting case. In retrospect, it is easy to find the many reasons that created those challenges. Being non-immune the clinical picture was atypical in many ways from what we see in victims of Bilharzias in endemic areas.⁸ On top of the presenting illness, he acutely developed clinical features of Schistosomal myeloradiculopathy an extremely rare complication schistosomiasis with only approximately 150 case reports and case series in the literature up to 1999⁹ against around 200 million affected subjects with 600 million exposed worldwide.¹ Moreover, all consultants involved in the management of this patient were not aware of the existence of Schistosomal myeloradiculopathy, a fact highlighted in the literature.⁷ At admission, our patient had the mixture of radiculopathy and myelopathy a combination typically of the more common and unique spinal cord pathology of *Schistosoma mansoni* CNS involvement, namely myeloradiculopathy of lower spinal cords (conus medullaris) and quada equina.⁹ The cerebrospinal fluid analysis carried out at admission was normal, probably due to the fact that it was carried out early in the course of the illness, it was found unnecessary to repeat after we had the histopathology reports of colonic biopsy.

The diagnosis of schistosomal myeloradiculopathy is usually presumptive according to the following criteria CDC (1984):⁷ existence of low spinal lesion, exposure, and evidence of Schistosomal infection, exclusion of other causes of transverse myelitis. A definite diagnosis is by tissue biopsy, which is rare if at all performed now a days. The findings in the initial MRI of spinal cord of

moderate enlargement of lower cord (conus medullaris) with alterations suggestive of myelitic form of disease on T1 with hyperintense signal in T2-weighted sequences and heterogenous diffuse granular enhancement following gadolinium injection, is similar to that reported in the literature.⁷ An interesting finding in our case was the improvement in the MRI changes in an early follow up study, carried out after only 2 weeks of high steroids (80 mg Prednisolone/ day), that prevented a proposed urgent neurosurgical intervention .

Regarding the treatment most authors stress the importance of starting the treatment early after the onset of neurological problem as the likelihood of the response is better and that the clinical improvement may occur late in the course of the disease with maintenance of high steroids therapy and that normalization of the spinal cord MRI is sometimes not indicative of clinical improvement.^{7,9}

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Case Reports

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