Manson’s schistosomiasis presenting as a brain tumor

Case report

IAN R. A. MACKENZIE, M.D., F.R.C.P.(C), AND ABHIJIT GUHA, M.SC., M.D., F.R.C.S.(C)
Department of Pathology and Laboratory Medicine, University of British Columbia, Vancouver, British Columbia, Canada; and Division of Neurosurgery, University of Toronto, Toronto, Ontario, Canada

Neurological complications arising from schistosomiasis are uncommon, and reports of Manson’s schistosomiasis presenting as an intracerebral mass lesion are particularly rare. The authors describe the case of a 26-year-old man with a 3-month history of headaches and visual abnormalities. He had immigrated to Canada from Brazil 4 years previously. The patient’s general physical and neurological examinations were unremarkable. Magnetic resonance imaging revealed an enhancing lesion with surrounding edema and mild mass effect in the right occipital lobe. A stereotactic brain biopsy demonstrated intraparenchymal granulomas surrounding Schistosoma mansoni eggs. The patient’s symptoms resolved following treatment with praziquantel and steroid medications; follow-up MR imaging yielded normal findings. This case demonstrates that neuroschistosomiasis should be considered when an individual presenting with an intracerebral mass has lived in a region in which this disease is endemic.

KEY WORDS • schistosomiasis • brain lesion • brain tumor

Schistosomiasis is a metazoal infection caused by trematodes of the genus Schistosoma. More than 200 million people are infected worldwide. Three species cause significant disease in humans: Schistosoma mansoni is endemic to countries in northern South America, central and southern Africa, and Saudi Arabia; S. haematobium is distributed more widely throughout Africa and the Middle East; and S. japonicum is restricted to portions of Japan, China, and southeast Asia. Humans are the definitive host, although certain aquatic snails act as the intermediate host. The adult parasite is a flat, elongated fluke that lives in the mesenteric (S. mansoni and S. japonicum) or pelvic (S. haematobium) veins. Females of this genus lay hundreds to thousands of eggs per day, which are excreted in human urine or feces. Intestinal or urinary symptoms are most common; they develop when eggs remain in the wall of the bowel or bladder and incite a localized inflammatory response. Other organs such as the lungs, liver, and central nervous system (CNS) may be involved by hematogenous dissemination of the eggs or by migration of adult worms to ectopic locations.

Neurological complications arising from schistosomiasis are uncommon. Cerebral lesions most frequently occur in cases of Japanese schistosomiasis, whereas Manson’s and urinary or bladder (haematobium) schistosomiasis may involve the spinal cord. We describe an unusual case in which Manson’s schistosomiasis presented as an intracerebral mass lesion.

Case Report

History. This 26-year-old man grew up in rural Brazil and had worked on a banana plantation before moving to Canada in 1989. He returned to Brazil in 1992 to await formal immigration to Canada the following year. The patient was married with one child and was employed part-time as a construction laborer. He had appeared well until 3 months before hospital admission when he developed right-sided, frontotemporal headaches. The headaches were throbbing in nature, radiated posteriorly, and were often associated with visual scintillation. They could occur at any time during the day but did not awaken him from sleep. There was no association with position or movement and no nausea or vomiting. Between episodes, the patient felt well. By the time he sought medical attention, the headaches were occurring daily and were no longer relieved by administration of acetaminophen.

The patient’s medical history included a fractured arm, which occurred when he was 7 years old, and minor trau-
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Examination. The patient appeared to be a healthy young man in no distress. His blood pressure, heart rate and temperature were normal. A general physical examination proved to be unremarkable, with the patient exhibiting no rash, organomegaly, or lymphadenopathy. The neurological examination was also normal, with no visual defect identified by confrontation and no papilledema. Routine laboratory tests included normal electrolytes, renal and liver function tests, erythrocyte sedimentation rate, and white blood cell count without eosinophilia. A magnetic resonance (MR) image of the patient’s head revealed a heterogeneously enhancing mass in the right occipital lobe with some surrounding edema (Fig. 1). The clinical impression was that of an intrinsic brain tumor and a stereotactic biopsy was performed.

Histological Findings. The biopsy produced two cores of tissue, each measuring 10 × 1 mm. Granulomatous inflammation was present within the cerebral cortex and overlying leptomeninges (Fig. 2). The granulomas consisted of a central collection of epithelioid cells and multinuclear giant cells surrounded by a dense cuff of lymphocytes, plasma cells, and eosinophils. At the center of each granuloma was a foreign body, the appearance of which was consistent with that of a parasite egg. Most of the eggs had degenerated and fragments of the shell were being phagocytosed by multinuclear giant cells. The more intact embryonate eggs were oval and measured approximately 150 × 50 μm. A few eggs possessed a prominent lateral spine. When the Ziehl–Neelsen technique was used for staining, the shells of the eggs were found to be acid-fast. A diagnosis of cerebral S. mansoni (Manson’s schistosomiasis) infection was made.

Postoperative Examination and Treatment. A repeated physical examination, chest x-ray film study, and abdominal ultrasonography failed to disclose evidence of systemic disease. No ova or parasites were identified in multiple samples of feces or urine; however, serological testing for schistosomiasis, using an indirect fluorescent antibody technique, proved positive with a titer of 1:1024. The patient was treated with two doses of praziquantel (20 mg/kg) and dexamethasone (10 mg). When he returned for follow-up examination 4 months later, he was free of symptoms. Repeated MR imaging performed at 10 months yielded normal results.

Discussion

Neurological complications arising from schistosomiasis are uncommon, but may occur when eggs reach the CNS and elicit an inflammatory response. Symptomatic brain involvement is most common in cases of Japanese schistosomiasis. Rarely, encephalopathy or encephalitis develop during the acute stage of the infection, referred to as “Katayama fever.” In chronic disease, raised intracranial pressure and focal signs and seizures can result from granulomatous inflammation, which may be disseminated or focal (tumoral form). Neuroschistosomiasises caused by S. haematobium and S. mansoni infections usually involve...
the spinal cord with miliary granulomas or a focal granulomatous mass that produces lumbosacral transverse myelitis.

It is perhaps surprising that cerebral or cerebellar symptoms do not develop more frequently in patients with Manson’s schistosomiasis. Several autopsy studies in patients from regions in which this disease is endemic have provided histological evidence of brain involvement in a significant proportion of infected individuals, particularly those with the hepatosplenic form of the disease associated with pulmonary hypertension.\(^*\) In these cases, embolization of eggs is probably facilitated by the arteriovenous shunts that result from hepatic and pulmonary hypertension. The lack of neurological symptoms in such cases may be explained by the relatively small numbers of eggs that are disseminated over a wide area and are often associated with only mild inflammation. Symptoms are more likely to occur when a large number of eggs is concentrated in one area and the associated granulomatous inflammation produces a tumorlike mass. It is believed that this results from the anomalous migration of adult worms to the CNS, resulting in situ egg deposition.\(^*\) Fortunately, this event seems to be a rare occurrence.

In a recent review, Pittella, et al., described four cases of the tumoral form of Manson’s schistosomiasis and identified seven other previously reported cases with histological confirmation. Similar to our case, most of their patients were young adult males (eight males and three females; age range 11–38 years, mean 25 years) who had previously been healthy. These patients typically presented within 3 months of the onset of neurological symptoms, the most common being headache, localized signs, and seizures. Radiological examinations identified a solitary mass lesion in the cerebellum in five patients, the cerebrum in four patients, and the temporal subdural space in one patient. One patient had two discrete lesions, one in the frontal lobe and one in the thalamus. It is interesting to note that in several patients, the initial neuroimaging was interpreted as being normal.\(^*\) In all cases, the resected material showed schistosomal granulomas. Patients who were treated with complete surgical resection and antihelmintic medication (praziquantel or oxamniquine) tended to have a good outcome, whereas those who underwent partial resection or biopsy more often had residual or recurrent symptoms despite antihelmintic therapy. One patient who was only treated with niridazole (no surgery) died; the diagnosis was confirmed at autopsy. Our patient is currently asymptomatic, despite minimal surgical intervention. However, we must caution that his follow-up study has lasted only 1 year.

**Conclusions**

Although an extremely rare complication, Manson’s schistosomiasis should be considered in the differential diagnosis when an individual from an endemic region presents with a mass lesion in the brain. In most reported cases including this one, the clinical history and preoperative laboratory investigations failed to suggest the diagnosis. It was only recognized after biopsy.\(^*\)

**References**


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Address reprint requests to: Ian R. A. Mackenzie, M.D., F.R.C.P.(C), Department of Pathology and Laboratory Medicine, Vancouver General Hospital, 855 West 12th Avenue, Vancouver, British Columbia V5Z 1M9, Canada. email: imackenz@vanhosp.bc.ca.