THE ROLE OF CORTICOSTEROIDS IN THE TREATMENT OF CEREBRAL SCHISTOSOMIASIS CAUSED BY SCHISTOSOMA MANSONI: CASE REPORT AND DISCUSSION

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Abstract. A 26-year-old Brazilian man was admitted to The Toronto Hospital with a headache and visual scintillation. His last travel to Brazil was five years previously. A computed tomography (CT) scan of the head showed an occipital mass with surrounding vasogenic edema. Occipital brain biopsy revealed Schistosoma mansoni eggs. The patient was treated with two doses of praziquantel (20 mg/kg) and dexamethasone (10 mg). His symptoms and occipital mass resolved. Cerebral schistosomiasis is, in part, caused by the host’s inflammatory response to Schistosoma. Modes of treatment have included surgical resection, the antiparasitic drugs oxamniquine or praziquantel, and corticosteroids. Corticosteroids may diminish granulomatous inflammation, thereby preventing further tissue destruction, and there is evidence that they also reduce ova deposition. Our review of the literature supports prompt medical therapy in patients with cerebral schistosomiasis. While the minimally or asymptomatic individual may be treated with praziquantel alone, clinicians should consider adjunctive therapy with corticosteroids for patients with prominent neurologic signs or symptoms or mass lesions with evidence of surrounding edema on a CT scan or by magnetic resonance imaging.

Vital signs and results of a general physical examination were normal. In particular, there was no hepatosplenomegaly or lymphadenopathy. A neurologic examination showed that the visual fields and fundi were normal, as were the cranial nerves, power, and sensation, and the results of a mental status examination.

A complete blood count and a differential count were normal; no eosinophilia was noted. Electrolytes, renal and liver function test results, and a chest radiograph were normal.

CASE REPORT

A 26-year-old right-handed Brazilian man was admitted to The Toronto Hospital because of a right-sided headache and a right occipital lobe mass. The patient had been well until three months prior to presentation when he experienced daily right-sided frontal and temporal headaches that were associated intermittently with episodic visual scintillation but no triggering factors. The headaches were not accompanied by nausea, vomiting, or any seizure activity, and acetaminophen was often successful in alleviating them. There was no history of other neurologic symptoms, fever, night sweats, or weight loss.

The patient grew up on a farm in rural Brazil and emigrated to Canada in 1989. During a one-year visit to Brazil in 1992, he swam in fresh water and walked barefoot regularly. He did not ingest uncooked meat or seafood and had no pets. His only significant risk factor for human immunodeficiency virus infection was unprotected heterosexual intercourse.

FIGURE 1. Gadolinium-enhanced magnetic resonance imaging of the brain of the patient showing a right occipital mass with surrounding vasogenic edema.
Axial computed tomography (CT) of the head without intravenous contrast revealed a poorly defined occipital mass with surrounding vasogenic edema. Magnetic resonance imaging of the head with gadolinium contrast revealed a solitary heterogeneously enhancing right intra-axial occipital mass lesion with surrounding vasogenic edema (Figure 1). Ultrasound of the abdomen was normal. The provisional diagnosis was a brain tumor and an open biopsy was performed.

A right occipital lobe brain biopsy revealed numerous microgranulomata composed of epithelioid cells and occasional giant cells, surrounded by lymphocytes, eosinophils, and plasma cells. In the center of the granulomata were oval structures measuring 100×50 microns with an outer wall containing delicate granular material that stained acid fast and silver positive. Some of these structures showed a single lateral spine, diagnostic of *S. mansoni* eggs (Figure 2).

Schistosoma serology was positive by indirect fluorescent antibody at a titer of 1:1,024. Multiple stool concentrates and a stool egg count by the Kato technique revealed no *Schistosoma* ova; the *Schistosoma* hatch test revealed no miracidia after 24 hr.

The patient’s clinical symptoms resolved with two doses of praziquantel (20 mg/kg) and dexamethasone (10 mg). Approximately one month after the treatment, repeat magnetic resonance imaging of the head performed without contrast revealed a small defect in the right occipital bone (from the previous biopsy) and a very tiny amount of residual edema in the white matter of the right occipital lobe (Figure 3). There was complete resolution of the mass. At four months follow-up the patient continues to be asymptomatic.

**DISCUSSION**

The first description of cerebral schistosomiasis based on localization of ova was reported by Yamagiwa in 1889. In the intervening years, there has been extensive documentation of cerebral infection with *S. japonicum*. However, *S. mansoni* very rarely invades the brain as shown by only 12 documented cases reported in the literature. A recent review by Pittella and others outlined the 11 cases published between 1984 and 1995, following which only one additional case report has been noted. Until 1984, there were 14 reports of brain involvement by *S. mansoni* ova. The prevalence of this condition is undoubted much higher, as shown by random cadaver examination from endemic areas that have found schistosome species in up to 28% of selected brains, including 4% infection by *S. mansoni*. Regarding Brazilian patients with fatal hepatosplenic schistosomiasis, 26% had brain involvement on autopsy.

Before discussing therapy of cerebral schistosomiasis, it is important to understand the pathogenesis of this disorder. Cerebral symptoms result from the granulomatous inflammatory response of the host to egg deposition in the brain. Although the precise source of the eggs has not been fully defined, most investigators believe that eggs released into
towards the portocaval system reach the brain via pulmonary or portopulmonary venous shunts. Although adult worms may anomalously enter the vertebral venous plexus and ascend to the brain, such findings are extremely rare. In spinal schistosomiasis, eggs appear to reach the spinal veins via the valveless venous plexus of Batson that joins the deep iliac veins and inferior vena cava with the veins of the spinal cord.

The optimal method of treatment of cerebral S. mansoni remains to be determined. Both surgical resection and medical therapy have been used. In the 12 documented cases, nine underwent surgical resection (five total resections, three partial resections, and one unspecified resection). Five of these received adjunctive oxamniquine and two received praziquantel. One received oxamniquine and corticosteroids. Two received praziquantel and corticosteroids. One without any therapy died. Of these patients, four of five treated with total resection and adjunctive medical therapy had a complete remission, whereas the three patients treated with a partial resection and adjunctive medical therapy had slight residual symptoms. The two treated with praziquantel and corticosteroids without any surgical intervention had complete resolution of their symptoms, as did our case.

The use of either oxamniquine or praziquantel seems logical. Both drugs cause death of the adult worm, resulting in cessation of oviposition and thus a reduction in the inflammatory response. Their use in non-CNS S. mansoni infection is well documented.

On the other hand, the benefit of concomitant corticosteroid use is not well established. Corticosteroids are expected to diminish granulomatous inflammation and edema, thereby preventing further tissue damage. In addition, there is some evidence that corticosteroids reduce ova deposition by adult worms and concomitant administration of an antiparasitic agent can further enhance this effect. In establishing the effectiveness of single-day praziquantel treatment for cerebral S. japonicum, Watt and others administered corticosteroids only to patients with symptoms of intracranial hypertension. They found good clinical outcomes in the group that received corticosteroids and in the group that did not. They did not witness any severe treatment-related inflammatory reactions, as can occur in cerebral cysticercosis, in those treated with praziquantel alone.

In spinal cord schistosomiasis, use of combination therapy is more frequent. Haribhai and others described 14 patients; eight showed rapid clinical and radiologic improvement after treatment with praziquantel and corticosteroids. One patient improved soon after being treated only with corticosteroids. Although the importance of early antischistosomal and corticosteroid therapy has been emphasized by some investigators, previous experience has shown that early treatment with praziquantel alone can also be effective.

Pollner and others reported a case of cerebral schistosomiasis with significant mass-related edema caused by S. haematobium. The patient was treated with praziquantel and dexamethasone. Discontinuance of dexamethasone on two occasions led to immediate recrudescence of headache, suggesting that the drug played an important role in reducing inflammatory edema.

Kirchhoff and Nash reported a case of acute cerebral S. japonicum in a healthy marine officer who presented with neurologic symptoms and had multiple focal cerebral lesions on a CT scan. Treatment with dexamethasone alone resulted in complete resolution of his symptoms and cerebral CT lesions. The parasitic infection was identified only after the patient completed a 10-week course of corticosteroids. He subsequently received praziquantel.

In view of the rarity of the disorder, controlled trials to assess the efficacy of corticosteroids in cerebral schistosomiasis have not been conducted. Based on our literature review, CNS lesions may be treated with mass resection alone, with anti-schistosomal therapy alone, or with both modalities. Corticosteroids have been used in conjunction with each of these treatments and rarely in isolation.

At present, a consensus has not been reached on the optimal mode of therapy for all presentations of cerebral schistosomiasis. However, given the pathogenesis of the disorder and our current literature review, we would not encourage surgical excision unless the diagnosis is uncertain, as undoubtedly it was in the majority of cases for which surgical therapy was undertaken. In a patient with neurologic symptoms and cerebral lesions believed to be caused by a schistosomal infection, prompt medical therapy with both praziquantel and corticosteroids has been shown to be effective. Praziquantel should be administered according to the infecting schistosome species and a steroid dose equivalent to 1 mg/kg/day of prednisone can be initiated and continued for several days as clinically warranted. The minimally symptomatic or asymptomatic patient with cerebral mass lesions may be treated with praziquantel alone. It is important to point out that therapy-induced dead worm reactions of the....

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**Figure 3.** Post-treatment magnetic resonance imaging of the brain of the patient showing resolution of the previously seen mass lesion.
type seen in cerebral cysticercosis do not occur with worm or egg death in schistosomiasis; therefore, steroids are not mandatory in minimally or asymptomatic individuals with CNS schistosomiasis. A randomized, blinded, and placebo-controlled trial comparing the use of praziquantel alone with that of praziquantel and corticosteroids might be possible in endemic areas. This would help to further diminish the uncertainty that exists regarding the role of corticosteroids in the treatment of cerebral schistosomiasis.

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