SCHISTOSOMIASIS MANSONI PRESENTING AS A CEREBELLAR TUMOR

Case report

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ABSTRACT - The Manson's schistosomiasis tumoral form rarely affects the brain. There are only 12 cases prior related with a mean age of 25 years and a male predominance. We describe a 16-year-old Brazilian Northeastern boy with a cerebellar mass lesion. The radiological aspect was considered compatible with glioma and a gross total resection was performed. Microscopic examination disclosed intraparenchymal granulomas surrounding Schistosoma mansoni eggs. The case is compared with the literature findings and some peculiar aspects of this trematode infection are reviewed.

KEY WORDS: schistosomiasis, tumor, cerebellum.

Esquistossomose mansônica apresentando-se como tumor cerebelar: relato de caso


PALAVRAS-CHAVE: esquistossomose, tumor, cerebelo.

Schistosomiasis is one of the most common parasite infections. Approximately 200 million people are carriers and another 600 million are exposed to the risk of infection in Latin America, Africa and Asia. This metazoal infection is caused by trematodes of the genus Schistosoma and three species cause significant disease in humans: Schistosoma mansoni (endemic to countries in South America, central and southern Africa and Saudi Arabi); S. haematobium (distributed throughout Africa and Middle East); and S. japonicum (restricted to Japan, China and southeast Asia). Humans are the definitive host, although aquatic snails act as the intermediate host. Adult parasites live in the mesenteric (S. mansoni and S. japonicum) or pelvic (S. haematobium) veins and females of this genus lay hundreds of eggs per day, which are excreted in human urine or feces. The most common urinary or intestinal symptoms are caused by a localized inflammatory response surrounding these eggs; 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 1-5. CNS clinical involvement is uncommon, cerebral lesions most frequently occur in cases of Japanese schistosomiasis 6, whereas the other two types (Manson's and haematobium) may involve the spinal cord 5. The cerebral or cerebellar tumoral form of schistosomiasis mansoni is extremely rare, there are only 12 cases prior related 1,5,7-10.

We describe a curious case in which Manson's schistosomiasis simulated a posterior fossa tumour.

CASE

We received the informed consent from the patient’s family permitting this publication.

History – A Northeastern Brazilian 16-year-old boy developed bilateral fronto-temporal headache 2 months before the admission. The headaches radiate posteriorly...
and were not associated with visual scintillation, position, movement, nausea or vomiting. They were occurring daily and were not relieved by administration of common analgesics. The patient’s medical history disclosed no relevant findings.

Examination – The patient appeared to be a healthy young boy. General physical examination was normal. Neurological examination showed slight gait ataxia and incoordination of the movements of the right limbs. Computer tomography (CT) scan showed a heterogeneous, hyperdense area, of irregular borders, enhanced by intravenous contrast, in the right cerebellar hemisphere (Fig 1).

Surgery – The lesion was considered compatible with glioma and suboccipital craniotomy with gross total resection was performed. The “tumour” infiltrated the cerebellar cortex and white matter. It was bloodless and the consistency was slightly more firm than normal tissue.

Histological findings – Microscopic analysis showed sections of leptomeninges and of the cerebellum cortex and white matter containing numerous S. mansoni eggs involved in chronic granulomatous inflammatory reaction (Fig 2).

Postoperative evolution – The patient was submitted to chemotherapy with oxamniquine. His symptoms gradually improved.

DISCUSSION

Neurological complications arising from schistosomiasis are uncommon and was initially described in 1899 by Yamagiva. This author performed an autopsy in a carrier of Katayama disease who suffered focal seizures and identified a brain granuloma.

Symptomatic brain involvement is most common in cases of Japanese schistosomiasis. Eventually, 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 or seizures can result from granulomatous inflammation. Neuroschistosomiasis caused by S. haematobium and S. mansoni infections usually involve the spinal cord with multiple granulomas or a focal granulomatous mass that cause lumbarosacral transverse myelitis.

Fig 1. CT scan disclosing a cerebellar heterogeneous and irregular lesion.

Fig 2. Specimen obtained showing granulomatous inflammation. Photomicrograph H&E.
It is curious that clinical brain involvement do not develop more frequently in patients with Manson’s schistosomiasis. Some autopsy studies provided histological evidence of cerebral and cerebellar ectopic eggs location in infected individuals, particularly those with the hepatosplenic form of the disease associated with pulmonary hypertension. The lack of neurological symptoms in such cases may be explained by the small amount 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. The tumoral form of cerebellar schistosomiasis mansoni, as it is observed in our case and the twelve other in the literature, showed not to be associated with other severe forms of visceral involvement by S. mansoni. Probably, we may expect that those patients were carriers of mild intestinal or hepatointestinal forms.

It must be discussed how the S. mansoni ova reach the brain. There are two possibilities: the eggs may be carried into the brain through the arterial system or through the retrograde venous path, or in other way, it is possible an anomalous migration of adult worms and local deposition of ova. The first mechanism occurs in carriers of the hepatosplenic form of schistosomiasis, in which eggs distributed in the brain and surrounded by inflammatory reaction were found in 26% of autopsy cases. This finding leads to the conclusion that cerebral Manson’s schistosomiasis is quite common, although it is not clinically relevant in the majority of cases. The hypothesis of arterial embolization supposes the presence of either pre-existing or disease-related pulmonary arteriovenous shunts resulting of organizing thrombi in the pulmonary circulation causing pulmonary arterial hypertension. Another possible path for the arterial embolization would be related to the portal-pulmonary anastomoses through the azygos veins facilitate by the presence of portal hypertension. The local deposition of eggs after migration of adult worms is suggested by two facts: worms are occasionally found inside the leptomeningeal veins and eggs grouped in the same area are found.

The main region involved in the tumoral form is the vertebro-basilar circulation territory. This fact could be explained by the anomalous migration hypothesis: a local deposition of eggs after the adult worm reach the CNS through venous anastomoses between brain, spinal cord and the portal system by the Batson’s venous vertebral plexus, probably the portal hypertension facilitates this migration.

The majority of Manson’s schistosomiasis tumoral cases, as in our report, were young adult males (9 males, 3 females, mean age 25 years) who had previously been healthy. These patients typically presented within three months of the onset of neurological symptoms, the most common being headache, focal signs and seizures. The ideal management is a controversial issue because this is an extremely rare and almost unexpected diagnostic. Patients who were treated with complete surgical resection and anthelmintic medication tended to have a good outcome, whereas those who underwent partial resection or biopsy more often had residual or recurrent symptoms despite anthelmintic therapy. It is not predictable if there is a possibility of clinical remission without gross resection.

In conclusion, Manson’s schistosomiasis must be considered in the differential diagnosis when a young male patient from an endemic region presents with a brain mass lesion in the posterior circulation territory. There is evidence, from the previous reported cases, of a beneficial effect of a aggressive surgical resection followed by specific chemotherap with praziquantel or oxamniquine.

REFERENCES