Schistosomotic choroiditis. II. Report of first case

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SUMMARY The first case of granulomatous choroiditis produced by Schistosoma mansoni with histological confirmation is reported. The patient had the hepatosplenic and cardiopulmonary forms of the disease and presented with cerebral schistosomiasis. The funduscopic aspects of the lesion and the possible pathways taken by the parasite to reach the choroid are discussed.

Uncommon sites for infection by Schistosoma mansoni seem to be associated with certain anatomico-clinical forms of the disease. One of these unusual sites, the brain, was recorded in 26% of patients with the hepatosplenic form of schistosomiasis. One case has been reported of infection of the eye by an immature male S. mansoni parasite, in the anterior chamber. The funduscopic observation made by Oréfice et al. of choroidal nodules in five cases of the hepatosplenic form of schistosomiasis prompted one of the authors (Pittella) to carry out a systematic search for ova and granulomas in semiserial histological sections of the eyes of two deceased patients who had the hepatosplenic form of S. mansoni infection. In one case S. mansoni ova and granulomas were found in the choroid. We report this case, as no histologically confirmed granulomatous choroidal schistosomotic infection has been reported so far.

Case report

A female aged 17 years, born in Medina (Minas Gerais State) and living in Belo Horizonte, had been asymptomatic until 1½ years ago, when she presented with dyspnoea on effort, which became worse over the next year or so. Recently she had suffered severe dyspnoea even at rest, with cough and vomiting. No palpitations or haemorrhagic sputum were noted, nor thoracic or abdominal pain.

On examination there was moderate tachypnoea, cyanosis, and reduced capillary perfusion, but no oedema. The blood pressure was 140/80 mmHg, the heart rate 108 per minute, with normal rhythm. The heart had a pulmonary presystolic click, with normal first and second heart sounds. There was hepatosplenomegaly. Funduscopy was not done. The patient died soon after hospital admission. The clinical diagnosis was severe pulmonary hypertension of unknown origin (cardiopulmonary schistosomiasis?).

Pathological investigations showed a schistosomotic type of Symmers hepatic fibrosis, a sclero-congestive spleen, a cardiopulmonary form of schistosomiasis, and moderate ascites. An intratral communication was found, with patent foramen ovale, 0.4 cm diameter. Neuroschistosomiasis was present, with perivascular S. mansoni ova and monocellular inflammatory infiltrate, a variable glial reaction, and occasional necrotic-exudative granulomas in the leptomeninges, cerebral cortex, and subcortical right frontal and temporal white matter, the left putamen, and close to the left dentate nucleus. Calcified S. mansoni ova agglomerates were present in the left lateral globus pallidus, adjacent to a small lenticulostriate artery. There were slight cerebral oedema, a small cavum septi pellucidi, and a pineal gland cyst. Anatomical variation of the circle of Willis was noted.

EYE EXAMINATION

No changes were noted macroscopically on the internal surface of the eye. Semiserial haematoxylin-eosin stained histological sections (7 μm) showed three schistosomotic granulomas characterised by embryonic and non-embryonic S. mansoni ova in the choroid, projecting slightly into the retina, surrounded by epithelioid cells in palisade formation and more external lymphocytes, plasmocytes, and eosinophilic granulocytes. The largest granuloma also showed periocular necrosis as a homogeneous acidophilic
Fig. 1 Schistosomotic granuloma in the necrotic-exudative phase in the choroid. (=Haematoxylin and eosin, ×100).

Area (granuloma in the necrotic-exudative phase), which was absent in the other two granulomas—in the productive phase (Figs. 1, 2, 3).

Discussion

The finding of schistosomotic granulomas in the choroid of a patient with hepatosplenic schistosomiasis mansoni confirms the suggestion made by Orefice of choroiditis caused by S. mansoni. These granulomas may correspond to those nodules seen at funduscopy by Orefice et al. One of the most important morphological characteristics of the nodules is their variation in size, which may be correlated with different phases in their development, as was noted in our case. As in the present case the patients studied by Orefice et al. presented with hepatosplenic schistosomiasis and, with one exception, the cardiopulmonary form. Another point to be mentioned is the finding of S. mansoni ova in the brain. Cerebral parasitism in schistosomiasis mansoni is found in 26% of patients with the hepatosplenic form; most of these cases also presented with the cardiopulmonary form of the disease.

The possible pathways by which ova might arrive at the choroid are worth considering. The discussion in Pittella and Lana-Peixoto on cerebral schistosomiasis is pertinent. There are two possibilities: embolism of ova through the arterial or retrograde venous systems, and local laying of ova following anomalous parasite migration. Arterial embolism requires the presence of pre-existing pulmonary arteriovenous shunts or shunts related to the parasitism. However, vascular changes in pulmonary schistosomiasis which were interpreted as arteriovenous fistulae are at present considered to result from the organisation of thrombi in the pulmonary arterial circulation. Two alternative routes for arterial ova embolism to the choroid are the porto-pulmonary azygous anastomosis, favoured by the portal hypertension in our reported case, or the passage of ova to the systemic circulation through direct communication between the right and left heart, which was found in our case.

On the other hand ova might arrive at the choroid by retrograde venous routes through anastomosis between the ophthalmic vein, the cavernous sinus, cerebral veins, the spinal cord, and the portal system by means of the vertebral venous plexus of Batson.
The increase in flow due to portal hypertension would favour the spreading of ova by this route.12

Ova lying in the choroid following anomalous parasite migration in the venous circulation can be postulated after the finding of a pair of ova of S. haematobium in the orbital vein of a 12-year-old child13 and ova agglomerates at a similar site in cases of conjunctival and lacrimal gland bilharzia.14-15 In our case an agglomerate of ova was found in the globus pallidus, a finding already noted in other parts of the brain and spinal cord in cases of schistosomiasis mansoni,16-18 haematobium,19-20 and japonica.21-23 Similarly, adult parasites have been found in the central nervous system in a few cases.7, 20-24 Finally, another possibility by which ocular S. mansoni infection may occur is the entry of cercariae through the conjunctiva and a posterior local parasite maturing.3,4, 25 However, in experimental models Queiroz,26 Abboud et al.,27 and Lester and Freeman28 showed that ocular entry of the parasite is not responsible for unusual eye lesions, resulting only in hepatointestinal schistosomiasis.

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References
21 Greenfield MG, Fritchard B. Cerebral infection with Schistosoma japonica. Brain 1937; 60: 361-72.