

Neuroschistosomiasis: An Unusual Intracranial Space Occupying Lesion

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Background: Neuroschistosomiasis is an uncommonly reported disease. **Case characteristics:** An adolescent Indian boy residing in Kenya presented with headache, visual symptoms and seizures, with MRI showing space-occupying lesions in the occipital lobe and cerebellum. **Observation:** Brain biopsy was diagnostic of neuro-schistosomiasis; complete recovery was seen with praziquantel and corticosteroid therapy. **Message:** This case highlights the importance of considering epidemiology in differential diagnosis and establishing definitive diagnosis even if it is by invasive methods.

Keywords: Headache, Raised intracranial pressure, *Schistosoma hematobium*, Seizures.

Schistosomiasis; although widely prevalent worldwide [1,2], is rarely reported from India due to the absence of the specific intermediate snail hosts [3]. Acute neuroschistosomiasis, usually due to *S. japonicum*, presents as acute meningo-cerephalitis, whereas, chronic neuroschistosomiasis results from ectopic migration of the eggs from the portal mesenteric or pelvic systems to the CNS leading to a granulomatous reaction to the eggs. [4]. These can present as Pseudotumoral encephalic schistosomiasis due to *S. japonicum*, or Spinal cord schistosomiasis in *S. mansoni*/*S. hematobium* [4]. We describe here a case of neuroschistosomiasis in a non-resident Indian adolescent living in Kenya.

CASE REPORT

This 17-year-old boy of Indian origin residing in Nairobi, Kenya had been experiencing headaches, light headedness and episodes of blurred vision for two months before admission. He also reported episodes of flickering balls of light in front of his eyes that came on randomly multiple times a day and left behind a dull occipital headache. There was no fever, weight loss or cough. Magnetic resonance imaging (MRI) done in Kenya showed space occupying lesions in the right occipital parenchyma and left cerebellum. He presented to our hospital with a tonic clonic seizure. The physical examination was normal. Antiepileptic therapy with levetiracetam was initiated.

Radiologic differentials of lymphoma, autoimmune vasculitis and tuberculosis were considered. Initial investigations revealed a normal complete blood count, ESR and CRP, and negative ANA and ANCA. CSF study

including a TB PCR was negative. A contrast enhanced CT scan of the chest and abdomen done to look for extra CNS TB was normal. Repeat MRI showed mild interval progression (**Fig. 1**). Again seen were focal mass- like lesions in the right occipital lobe and left cerebellum with prolonged T1- and T2-signal and perilesional vasogenic edema. The lesions had ill-defined and irregular margins with heterogenous enhancement. Arborised linear enhancement with interspersed enhancing punctate nodules was seen. Punctate nodules formed the larger conglomerate masses. As the diagnosis could not be established through indirect and surrogate tests, an occipital craniotomy was done and brain biopsy taken.

On histopathology, the brain parenchyma showed granulomatous inflammation with numerous well-formed granulomas composed of epithelioid cells, multinucleate giant cells and central necrosis. The periphery showed

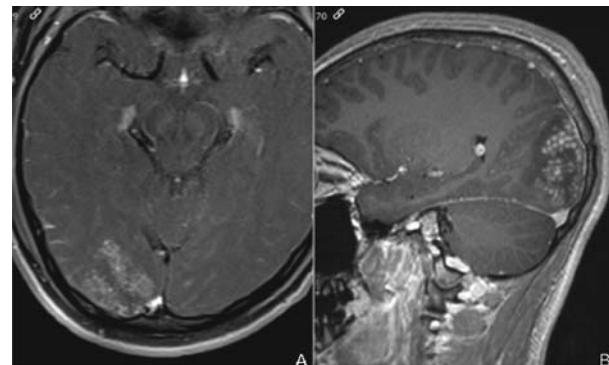


FIG. 1 Axial (a) and sagittal (b) T1 weighted contrast MRI shows conglomerate nodular and linear enhancing nodules with arborization.

lymphoplasmacytic infiltrate along with eosinophils. The center of many granulomas showed one or two non-operculated refractile eggs/ova of trematode with terminal spine (**Web Fig. 1**). Few ova show ruptured and distorted morphology engulfed by giant cells. No vasculitis or viable larvae of parasite were seen. Thus a tentative diagnosis of schistosomiasis was made. The ova morphology with a terminal spine resembled *S. haematobium*. The images were sent to the Department of Parasitic Diseases at Centers for Disease Control, Atlanta, USA where the diagnosis was confirmed.

On enquiry, the patient who was an avid swimmer gave a history of frequent swimming in natural fresh water lakes in Kenya. There was no history of urinary or intestinal symptoms, and his urine and stool analysis were negative for parasitic eggs. His serum was positive for schistosomiasis by hemagglutination (2560, reference range <160), ELISA (6.8, reference range <0.8 negative, 0.8-1.2 equivocal and >1.2 positive) as well as Western Blot (positive for the p30-32 / p20-24 bands)

The patient was started on praziquantel at a dose of 40 mg/kg/day for 3 days along with dexamethasone 4 mg thrice daily which was subsequently tapered over 1 month. His clinical symptoms resolved completely and a repeat MRI after 2 weeks showed remarkable regression of the CNS lesions. The boy was asymptomatic at the six-month follow-up, and a repeat MRI was normal.

DISCUSSION

Neuroschistosomiasis is commoner than perceived, diagnosed or reported [4-6]. An autopsy study from Africa reported that half the patients with urinary schistosomiasis had brain lesions [7]. Another pathological study in Africa found scattered ova of *S. haematobium* or *S. mansoni* in the brain at autopsy in around 25% of 150 unselected cadavers [5]. Only a handful of cases of indigenous urinary schistosomiasis have been reported from India, but none of neuroschistosomiasis [3].

In the index case, commoner differentials like tuberculosis, lymphoma, and autoimmune etiology were considered, but biopsy revealed the rare etiology. Even in most cases reported from endemic areas, the diagnosis is usually made inadvertently following brain biopsy and lesion excision. This is because the clinical symptoms of the disease are non-specific and mimicked by other infectious and non-infectious illnesses, including tumors. Some investigators have described arborised linear enhancement circled by multiple enhancing punctuate nodules as more characteristic for schistosomiasis [8]. CSF findings are also non-specific. Recent studies have reported on the utility of specific schistosoma serology and

PCR in CSF for diagnosis [9]. In some cases, the speciation was done by PCR analysis of brain tissue [5].

Treatment of chronic neuroschistosomiasis is not well standardized but good results with combination therapy with praziquantel and steroids [4] are reported. Current consensus is to reserve surgery only for non-responding lesions, severely elevated intracranial pressure and intractable epilepsy [10]. This case highlights the need to consider epidemiology in clinical differential diagnosis of any illness as well as the importance of establishing a definitive diagnosis even if it is by highly invasive methods.

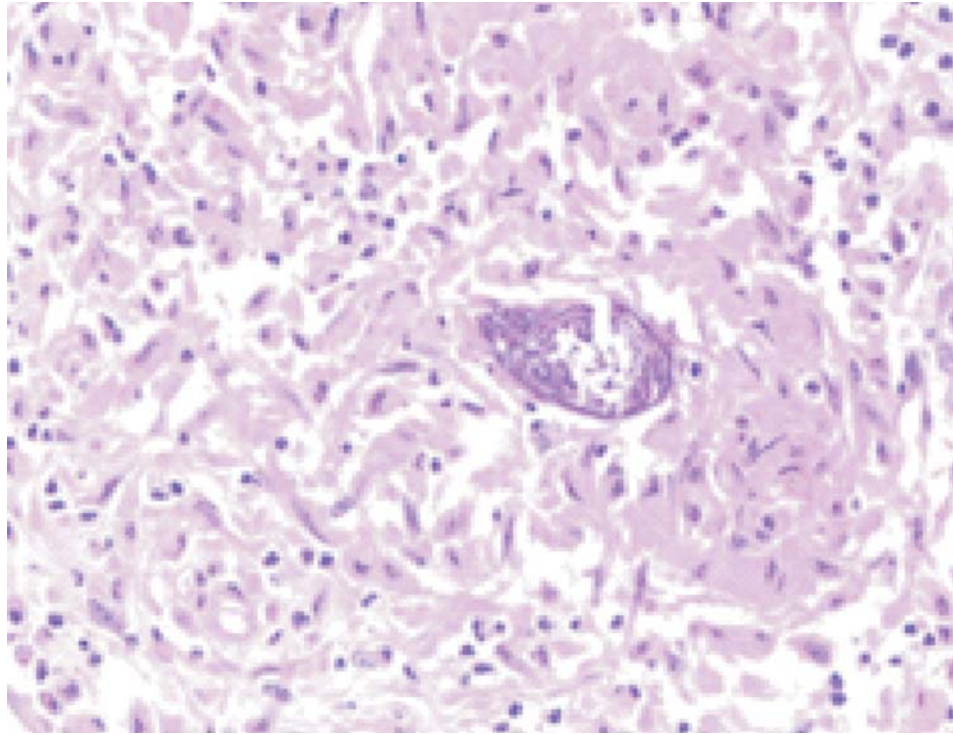
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WEB FIG. 1 Histology section of brain tissue show characteristic *Schistosoma haematobium* ova with terminal spine (Haematoxylin and Eosin, x 40 magnification).