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*Case Report*

# A Case Study of Intracranial Hypertension due to Neuroschistosomiasis

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## A B S T R A C T

**Introduction:** Neuroschistosomiasis is a rare and severe complication of schistosomiasis. It affects the Central Nervous System. Its prevalence in areas with proximity to freshwater bodies and poor sanitation is attributable to the aquatic life cycle and fecal-oral mode of transmission of the parasite. It presents a multitude of challenges in regards to diagnosis and treatment.

**Methods:** The case study reports of a 34 years old female who was misdiagnosed to have Benign Intracranial Hypertension and presented with severe unrelenting headache and visual disturbance. Upon examination, a fundoscopy revealing severe papilledema. Her other blood tests were unremarkable that led her to the misdiagnosis, until a positive bilharzia IgM antibodies were found. The patient was treated accordingly using Praziquantel with low dose steroids.

**Results:** Initiation of Praziquantel regimen and monitoring for a period of 2 months saw an insidious resolution in symptoms and of elevated intracranial pressure over a period of 9 months.

**Conclusion:** It if often overlooked that raised intracranial pressure maybe due t Neuroschistosomiasis,. Any patient presenting with the same and a history of exposure to schistosoma-endemic areas any time in their past history should be tested and treated with a positive Bilharzia IgM antibody prior to surgical interventions being considered.

**Keywords:** Neuroschistosomiasis; Benign intracranial hypertension; Cerebral schistosomiasis; Papilledema

## Introduction

Schistosomiasis is a widely prevalent and potentially devastating tropical parasitic disease. It affects more than 200 million people with an additional 800 million people at risk of infection worldwide, thus imposing a significant burden on public health. It has the second greatest socio-economic ramification of any parasitic disease, following malaria. According to the Global Burden of Disease Study 2016, the estimated schistosomiasis global burden is 1.9 million disability-adjusted life years (DALYs)[1]. Schistosomiasis is endemic in more than 78 countries, with more than 90% of the infections occurring in sub- Saharan Africa. Approximately six million individuals in Kenya are afflicted with schistosomiasis with fifteen million more at risk of infection[2]. The geographical distribution varies among the species. S. haemotobium is primarily found along the coast, Kano plains in Western Kenya and certain areas around Lake Victoria. S. mansoni is prevalent in Western regions and some parts of Central Kenya[3]. Recent studies report a prevalence of 2.1% for S. mansoni, and 14.8% for S. haematobium among school going children[4].

The World Health Organization (WHO) has advocated for integrated programs involving Mass Drug Administration (MDA)

in endemic areas, with the goal of eradicating schistosomiasis globally using a single dose of Praziquantel 40 mg/kg[5]. Efforts to eradicate schistosomiasis in Kenya have been ongoing since the MDA (Mass Drug Administration) initiative initiated in 2009 with nationwide expansion in 2012[4]. This has come with partial success as schistosomiasis remains endemic in areas with inadequate access to clean water and sanitation facilities.

The parasitic disease is caused by trematode blood flukes of the genus Schistosoma that reside in the vascular system of humans and other vertebrate hosts. The most important schistosomes that parasitize humans are S. haemotobium, S. mansoni and S. japonicum.

Schistosomiasis is characterized by complex pathogenesis. Upon contact with contaminated water sources, infective larvae penetrate human skin, initiating the infection. Once inside the body, they mature into adult worms, residing in mesenteric veins or vesicular venous plexuses. Infections with S. mansoni and S. japonicum are associated GI symptoms and chronic liver diseases. S. haemotobium results in urinary schistosomiasis. Chronic infection may lead to deposition of eggs in host tissue, inciting granulomatous reactions and fibrosis, which form the basis of the disease's diverse clinical presentation encompassing complications affecting various organs, including the rare occurrence in the central nervous system (neuroschistosomiasis). S. haemotobium, S. mansoni or S. japonicum account for most cases of neuroschistosomiasis[6].

Neuroschistosomiasis results from embolization of eggs to the CNS. Once deposited, schistosome eggs release proteolytic enzymes in the nervous tissue inducing a local eosinophilic inflammation. Ectopic eggs may produce granulomatous lesions throughout the body. Chronic, severe infections, due to accumulation of a vast number of eggs in tissues, leads to fibrosis, calcification and occasionally dysplasia and malignant change. Cerebral schistosomiasis is caused by S. japonium, resulting in acute encephalitis. The higher incidence of CNS involvement of S.japonicum may be owed to their small size eggs, which are released in higher numbers from the worm, and can be carried easily to the brain. Neuroschistosomiasis may present in two clinical syndromes, Spinal cord neuroschistosomiasis and localized Cerebral or Cerebellar neuroschistosomiasis[7].

The patients with cerebral neuroschistosomiasis may present with signs and symptoms of elevated intracranial pressure and focal neurological deficit, whereas those with spinal cord neuroschistosomiasis experience progressive myelopathy, inclusive of cases of Cauda-equina root involvement[8]. Cerebral neuroschistosomiasis is clinically classified into acute schistosomal encephalopathy and pseudotumoral encephalic schistosomiasis (PES). The latter is a chronic form of cerebral neuroschistosomiasis and is rarely encountered in clinical practice[9].

Neuroschistosomiasis, although rare, carries significant morbidity and mortality. It affects between 2% and 4% of the estimated 200 million people with systemic schistosomal infections[10]. 90% of cases of neuroschistosomiasis are found in sub-Saharan Africa, with cases of travel induced neuroschistosomiasis outside of endemic areas[11]. Due to the smaller number of cases reported and the increased rate of misdiagnosis, only about 500 cases have been reported globally since 1930[12].

This case study involves a patient who was misdiagnosed to have Benign Intracranial Hypertension instead of Neuroschistosomiasis. Benign Intracranial Hypertension is an idiopathic disorder characterized by increased Cerebro-spinal Fluid (CSF) pressure within the intracranial cavity. It produces signs and symptoms such as visual disturbances, headache, nausea and neurological deficits. Granuloma formation in neuroschistosomiasis due to deposition of eggs may obstruct flow of CSF within the ventricular system or subarachnoid space also causing similar intracranial hypertension.

There is a gross lack of understanding of the mechanisms of pathogenesis, diagnosis and treatment evidenced by the common misdiagnosis of this condition in common clinical practice.

## Case Report

A HIV-seronegative 34-year-old female of african ethnicity presented with a severe 8/10 headache and bilateral visual disturbance, notable blurring, without concomitant nausea or vomiting. Consciousness and cognition were preserved. Upon fundoscopic examination, pronounced bilateral papilledema was confirmed prompting an urgent MRI scan. The findings revealed features of Intracranial Hypertension possibly Idiopathic, without any MR evidence of dural sinus thrombosis or space occupying lesions. Further, bilateral optic nerve vertical tortuosity with peri-optic nerve sheath effusions were observed along with prominence of draining cortical veins, however, without identifiable intraluminal filling defects. The results of the Glaucoma Hemifield Test (GHT) indicated abnormal values for both eyes with deviations from normal limits.

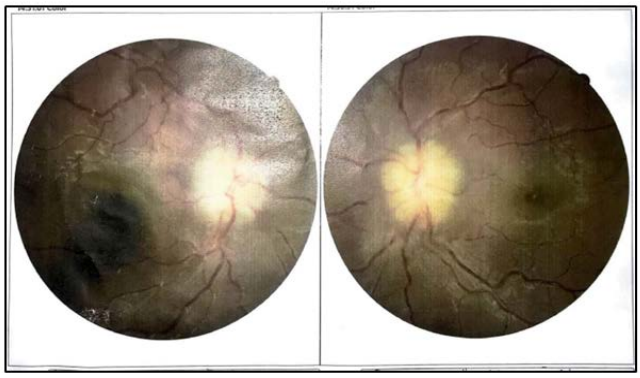

**Figure 1**: Fundoscopy on initial examination prior to treatment.

A complete blood count revealed a mildly iron deficient picture with normal leukocyte and differential levels, inclusive of eosinophil levels within normal limits. Malaria antigen was tested negative, urine and stool lab examinations were unremarkable except for a positive Helicobacter Pylori Antigen and ESR was elevated (54 mm/hr)

Based on the available results, a Ventriculoperitoneal (VP) shunt was recommended as a measure to alleviate intracranial hypertension. Upon physician assessment and review for possible prior to surgery, a serum Interferon Gamma Release Assay (IGRA) was performed to confirm the suspicion of ocular tuberculosis, yielding an equivocally positive result. Most importantly, serum ELISA confirmed the presence of Bilharzia IgM antibodies.

After confirmation of the presence of Bilharzia IgM antibodies, prompt intervention with praziquantel 2400mg twice a day for 3 days was prescribed and repeated after a month, with 12mg of daflazocort once a day for 2 weeks upon initiation of treatment, that was tapered to 6mg once a day thereafter. Diamox (acetazolamide) 500mg twice a day was continued for 6 months

to maintain intracranial pressure and progress was monitored. Follow up conducted after two months revealed relative improvement and in at 4 months, the absence of Bilharzia IgM was achieved. A complete resolution of symptoms of intracranial hypertension without diamox was found at 9 months from initial dose of praziquantel given. A control MRI displayed normal findings indicating an overall positive response to treatment. In spite of the resolution of infection, anemia persisted on follow up.

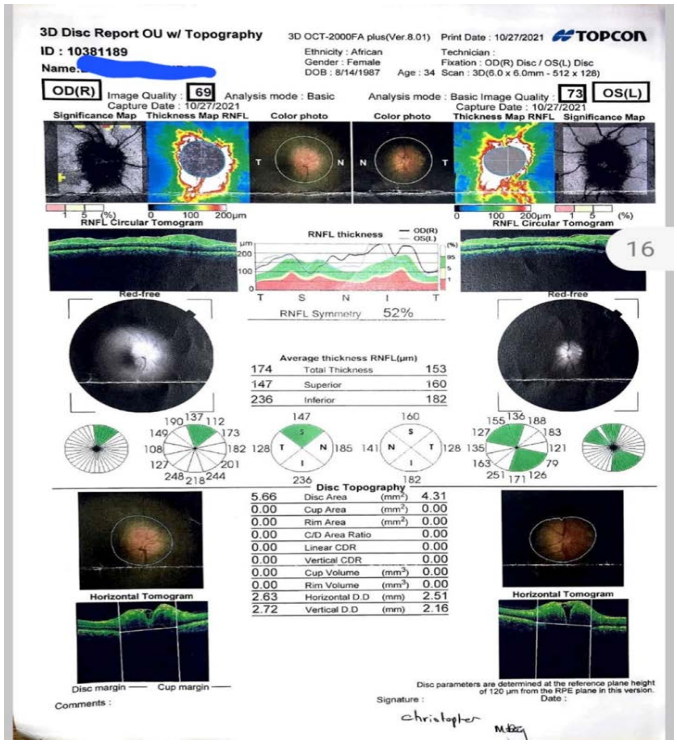


**Figure 2**: OCT scans prior to treatment.

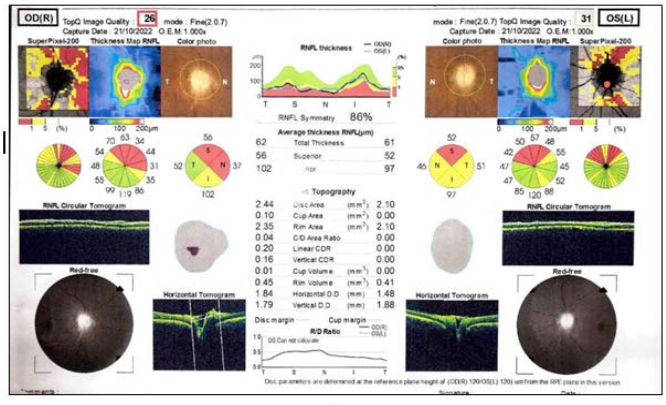


**Figure 3**: Fundocscopic examination and OCT scan 1 year post-intervention.

A review of this relatively unique case done 18 months post treatment confirmed a non recurrence of symptoms and a successful complete resolution of the disease.

## Discussion

Cerebral neuroschistosomiasis commonly present with symptoms of elevated ICP (headache, Cushing response, dizziness, vomiting, papilledema, visual disturbances, speech disturbances, hemiparesis, nystagmus and ataxia)[13], neurological manifestations (disturbed consciousness levels, dilated poorly reactive pupils, increased muscle tone, exaggerated deep tendon reflexes and hyperventilation with deep inspiration and expiration) and complications of increased ICP (Ischaemia, tonic convulsions and herniation syndromes). Focal neurological deficits may be present. These clinical manifestations closely mimic those of brain tumors.

Diagnosis of cerebral schistosomiasis is a challenge as neurological symptoms and imaging findings can be overlap with other conditions such as brain tumors. Granuloma formation around eggs is a typical finding in histopathology[14]. This diagnostic approach remains the gold standard to confirm the diagnosis[15].

Neuroimaging is an essential modality in diagnosis of neuroschistosomiasis. Contrast-enhanced magnetic resonance imaging may exhibit a linear enhancement pattern surrounded by multiple enhancing punctate nodules, also known as ‘arborized’ appearance. These imaging findings are suggested to be specific for cerebral neuroschistosomiasis. A review of 33 patients with cerebral schistosomiasis, MRI scans presented a typical pattern of single or multiple lesions compromising multiple intensely enhancing nodules, sometimes with areas of linear enhancement[16].

Praziquantel is the most effective drug used in the treatment of neurochistosomiasis[17]. Following diagnosis, Steroids, 8mg daily, may be considered before praziquantel to decrease inflammation that may result from the cytotoxic effect of praziquantel. Administration of 40-60 mg/kg of Praziquantel every 8 hours for three days duration should be given, it acts by causing tetanic contraction and paralyzing the parasite[18]. A second dose is required 4-6 weeks following initial treatment to eliminate any remaining parasites as praziquantel is ovastatic[19] and has little effect on eggs or immature worms, which also makes it ineffective in the early stages of infection[20].

The case being studied is of a 34 year old female highlights the diagnostic challenges associated with neuroschistosomiasis attributable to atypical symptoms of raised intracranial pressures alone. Past medical history and relevant exposure history is a significant guide to the possibility of such presentation being a form of the an often overlooked cerebral neuroschistosomiasis. This would necessitate the confirmation of this suspicion through serology. The presence of Bilharzia IgM antibodies would allow non surgical treatment for the condition.

It is yet to be established whether the progression to neuro schistosomiasis, in the absence of immunosuppression, has a genetic predisposition in certain populations as is the case here.

## Conclusion

Many patients presenting with idiopathic benign intracranial hypertension of late onset without and apparent cause may actually have an infective etiology. Screening for infectious such as schistosomiasis should be a strong consideration for physicians handling such cases prior to recommending surgical interventions.

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