

Imported CNS Schistosomiasis

—A case report—

You-Kyoung Lee, M.D., Tae-Youn Choi, M.D., So-Young Jin, M.D.,
Dong-Wha Lee, M.D.

Department of Clinical and Anatomical Pathology
College of Medicine, Soonchunhyang University, Seoul, Korea

Central nervous system(CNS) involvement may occur in chronic schistosomiasis. It can be produced by any Schistosome species but happens most frequently in chronic Schistosoma japonicum infection. CNS involvement by S. mansoni is relatively rare but it may occur by embolization of eggs or ectopic migration of adult worms. A case of cerebral schistosomiasis caused by S. mansoni in a 40-year-old man, who had worked in Yemen, is reported. Biopsies taken from the cerebellar vermis and the roof of the fourth ventricle, showed granulomatous inflammation due to eggs. S. mansoni was identified by stool examination and ELISA using serum and CSF. This is the first imported case of cerebral schistosomiasis in Korea.

Key Words : *Schistosoma mansoni*, CNS involvement, Biopsy.

INTRODUCTION

Schistosomiasis is prevalent in Africa and the Middle East, but virtually not existent in Korea (Min et al., 1982 ; Lee et al., 1989). The adult of *S. mansoni* lives in the mesenteric venous plexus of the large intestine, and eggs are often spreaded in the CNS by blood-borne migration from the deposition site. Schistosomal encephalopathy and myelopathy by deposition of eggs in the CNS has been rarely reported. Fifty one cases of encephalopathy and 60 cases of myelopathy had been reported in *S. mansoni* infection (Pittella, 1991). Two cases of combined cerebral and spinal schistosomiasis have been reported in Venezuela and Brazil (Dominguez & Bores, 1962 ; Pittella, 1985). In Korea, six imported cases of urinary schistosomiasis by *S. haematobium* (Min et al., 1982 ; Lee et al., 1989) were reported (Table

1) but any cases of *S. mansoni* infection or cerebral schistosomiasis are not reported.

We present a case of imported cerebral schistosomiasis by *S. mansoni* in a returnee from Yemen.

CASE PRESENTATION

Clinical Findings

A 40-year-old man was admitted to Soonchunhyang University Hospital because of headaches and dizziness. He had resided in Yemen from December 1989 to May 1990. During his stay he swam in a local river. On January 1993, he felt discomfort with pain in the lower extremities and tightness in the chest. Three months later, dizziness, headaches, and diplopia developed but these symptoms were aggravated in spite of steroid therapy.

On admission, he complained of headaches, nausea, vomiting, and leg pain. His vital signs and physical examinations were unremarkable. The neurologic examination disclosed normal findings,

Address for correspondence : You-Kyoung Lee, M.D., Department of Clinical Pathology, Soonchunhyang University Hospital, 657-58, Hannam-dong, Yongsan-gu, Seoul, 140-743, Korea.

Table 1. Summary of imported schistosomiasis in Korea.

Author	Organ	Species	Visiting country	Diagnostic method
Min et al. ¹	urinary bladder	<i>S. haematobium</i>	Yemen	egg & biopsy
Min et al. ¹	urinary bladder	<i>S. haematobium</i>	Yemen	egg & biopsy
Min et al. ¹	urinary bladder	<i>S. haematobium</i>	Yemen	egg & biopsy
Min et al. ¹	urinary bladder	<i>S. haematobium</i>	Yemen	egg & biopsy
Min et al. ¹	urinary bladder	<i>S. haematobium</i>	Yemen	egg
Lee et al. ²	urinary bladder	<i>S. haematobium</i>	Yemen	egg & biopsy
Present case	Cerebellum, Spinal cord	<i>S. mansoni</i>	Yemen	egg, biopsy, & serologic test

1. Min DY, Soh CT, Lee MS, Choi HK, Park CI : Urinary schistosomiasis among Korean returnees from the middle east. *Yonsei Reports on Tropical Medicine* 1982 ; 13 ; 36-40.
2. Lee SY, Lee HC, Park YY, Kwon SW, Kim OK : Urinary schistosomiasis : A case report. *Korean Journal of Urology* 1989 ; 30 : 224-7.

except a positive Romberg test on the right side and hyperesthesia below the L3 dermatome. Leukocyte count was $1.13 \times 10^9/L$ with peripheral eosinophilia of 18%. Cerebrospinal fluid examination showed $20 \times 10^6/L$ of red blood cells and $2 \times 10^6/L$ of white blood cells. Magnetic resonance imaging-(MRI) examination of the brain and spinal cord revealed irregular shaped lesions of high signal intensity in the cerebellar vermis, the roof of the fourth ventricle(Fig. 1), and the C2 level of the spinal cord (Fig. 2). These radiologic findings were suggestive of CNS tumor or granulomatous inflammation. Biopsies were performed from the cerebellar vermis and roof of the fourth ventricle on the fifth day of his stay in hospital.

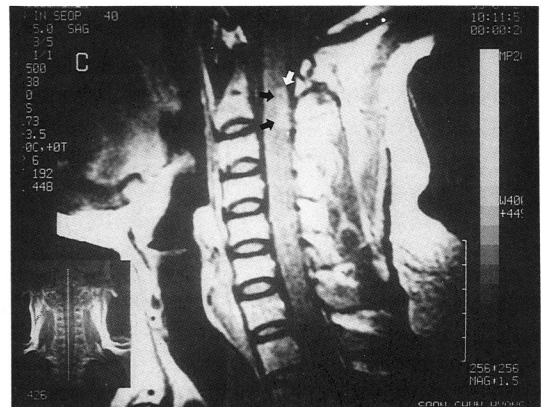


Fig. 2. Contrast enhanced T1 weighted sagittal image shows diffusely swollen cervical cord from C2 to C6 level and patchy nodular enhancement.

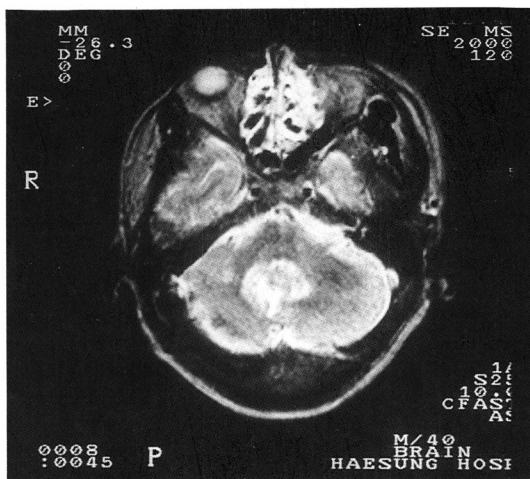


Fig. 1. T2 weighted axial image shows inhomogeneous hyperintense signal mass in cerebellar vermis. The mass is about 4X5cm and has minimal surrounding edema.

Pathologic Findings

Histologic examination revealed multiple relatively well-defined granulomas in gray matter or gray-white matter junction(Fig. 3). Each granuloma consisted of epithelioid cells and foreign body giant cells with infiltration of eosinophils. Some of the foreign body giant cells contained refractile egg shell in their cytoplasm. There was surrounding infiltration of lymphocytes, eosinophils and reactive gliosis(Fig. 4). These findings were consistent with schistosomiasis. We recommended further clinical evaluation to identify the species of schistosomiasis. Fresh CSF, stool, and urine were obtained: The stool revealed a few eggs, each of which was light brown colored, measured $150 \times 60 \mu m$, and had a lateral spine, consis-

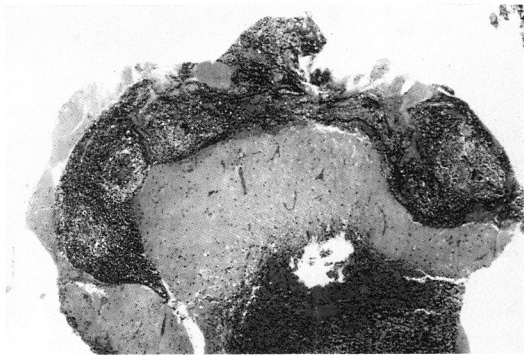


Fig. 3. Low power microscopic findings shows multiple granulomas in cerebellum(H-E stain,X40)

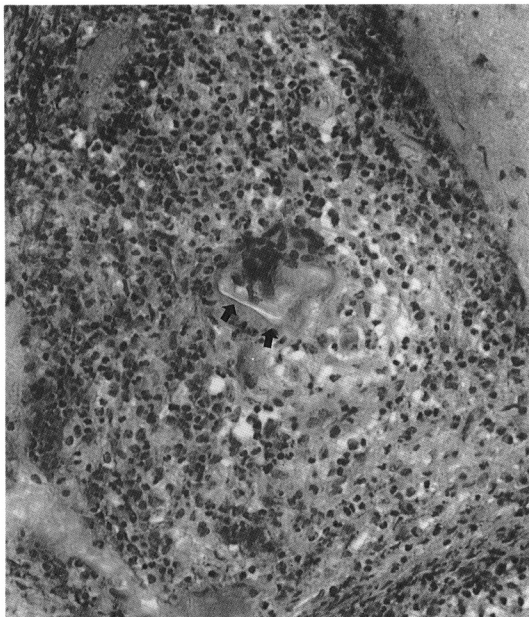


Fig. 4. The granuloma consists of multinucleated giant cells, phagocytizing parasitic egg shells, and inflammatory cells including eosinophils(H-E stain,X200)

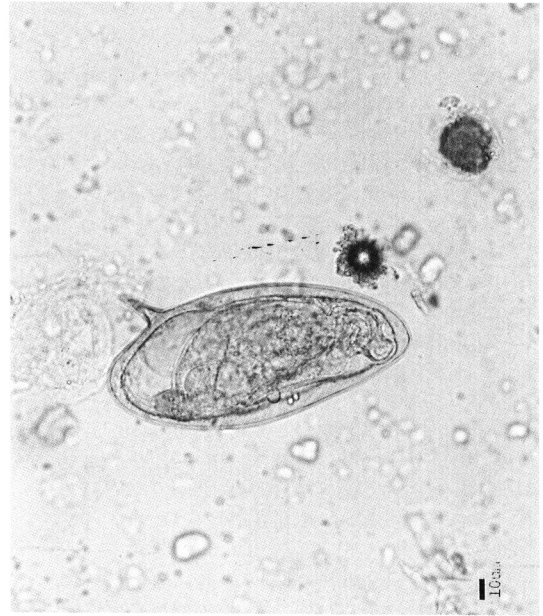


Fig. 5. The egg of *Schistosoma mansoni* measures 150X60 μm, is light brown, and has a lateral spine.(wet smear of stool,X200).

tent with *Schistosoma mansoni*(Fig. 5). Urine and CSF were negative for egg. Enzyme linked immunosorbent assay(ELISA) for parasitic infection including schistosomiasis were performed with CSF and serum, which disclosed significant high titers for *S. mansoni*(Table 2).

Postoperative course

Treatment with praziquantel(40 mg/Kg) was instituted. The patient was discharged after 3 months of postoperative day with improved state. Follow up examination by MRI after 7 months showed no evidence of the disease.

Table 2. The result of micro-ELISA test for various parasite-specific IgG antibodies in the present case

Parasitic antigens	Absorbance in		Cutoff O.D.
	serum	CSF	
<i>Schistosoma mansoni</i>	0.18	0.21	0.20
<i>Sparganum</i>	0.02	0.01	0.22
<i>Paragonimus westermani</i>	0.02	0.01	0.25
<i>Cysticercus</i>	0.02	0.01	0.18
<i>Clonorchis sinensis</i>	0.03	not done	0.25
<i>Fasciola hepatica</i>	0.03	not done	0.25

DISCUSSION

Humans can acquire schistosomal infection through contact with fresh water populated with cercariae-infected snails. A detailed history and duration of a patient's exposure to fresh water in endemic area is an useful step in considering the diagnosis of schistosomiasis.

Cerebral schistosomiasis can be produced by portal system encephalopathy in *S. japonicum* infection (Brown & Neva, 1983), but those of *S. mansoni* infection may rarely occur. Involvement of the CNS can be explained in any stage of infection; in the early stage of infection (non-toxic form), during evolution of the disease to its chronic forms, or concomitantly with the intestinal and hepatic forms (Pittella, 1991). Two possible mechanisms of CNS involvement can be considered. The first is embolization of eggs from the portal mesenteric system to the CNS via the arterial or retrograde venous flow (Spencer, 1977; Calabresi & Abelmann, 1957; Stein & Stein, 1961). And the second possibility is in-situ egg deposition following the anomalous migration of adult worms (Pittella & Lana-Peixoto, 1981). The random distribution of eggs in the CNS indicates their embolization hypothesis. Thus CNS involvement in the present case seems to be considered by the first mechanism. Because of the small and dispersed inflammatory reaction, and sparsity of ova, neurologic symptoms cannot be elicited in most *S. mansoni* infection (Pompeu & Sampaio de Lacerda, 1979). Sometimes, the mass effect produced by numerous eggs grouped in a single area and surrounded by granulomas, severe neurological symptoms observed in most patients. But this patient did not complain of any specific cerebellar symptoms, except for signs of increased intracranial pressure.

Blood examination often reveals eosinophilia, however it is not the rule (Rosenbaum et al., 1972; Queiroz et al., 1979; Selwa et al., 1991). The present case showed 18% of peripheral eosinophilia. Specific diagnosis depends on the characteristic eggs in stools in early stage of infection. When chronic or lightly infected, proctoscopic aspiration and biopsy revealed eggs in some cases (Herskowitz, 1972). The CSF is usually clear but in some patients with myelopathy, xanthochromia, mild pleocytosis, or eosinophilia may be present (Herskowitz, 1972; Ross et al., 1952; Odeku et al., 1968; Ghaly & El-Banhawy, 1973; Bambirra et al., 1984). Protein levels in the CSF may be slightly raise-

d (Odeku et al., 1968; Herskowitz, 1972), but glucose levels are normal. In the present case, eggs of *Schistosoma mansoni* were observed in stool, but not in CSF. The specificity of the serologic test for active infection is too low for the recovery of patients in endemic areas (King, 1991). But in non-endemic areas such as Korea, serologic tests are useful diagnostic tools for schistosomiasis. Many serological tests including circumoval precipitin (COP), indirect haemagglutinin (IHA), immunofluorescent (IF), complement fixation (CF) and double gel diffusion (DD) tests, have been used to assess the reactivity to Schistosome antibodies of serum and CSF in patients with suspected CNS schistosomiasis. All of these tests are sensitive while the specificity is variable. The ELISA is superior to IF, IHA and CF tests in respects of sensitivity and specificity (Suchet et al., 1987). Antibody responses are often elevated in the CSF than in the serum in the cerebral infection (El-Banhawy et al., 1972). ELISA for *S. mansoni* revealed positive both in the serum and CSF of the present case, and an absorbance in the CSF is higher than that in the serum. It is a supportive diagnostic tool to determine whether involvement of CNS of schistosomiasis by serology. The biopsy is essential for confirmation of CNS schistosomiasis.

Myelography in schistosomal myelopathy is an useful technique in documenting the extent of inflammation. A characteristic abnormality is seen in the granulomatous form of the illness usually at T12 to L1 levels, as a "swollen" or "trifid" image (El-Banhawy et al., 1972). However normal myelogram cannot exclude the possibility of CNS involvement (Scrimgeour & Gajdusek, 1985). CT and MRI are also of value in observing CNS schistosomal granulomas (King, 1991). The present case revealed a high signal intensity lesion seen in the C2 level of the spine and an irregular shaped high signal intensity lesion in cerebellar vermis and the fourth ventricular roof. These abnormal densities localize in the lesions and enable targetting of granulomatous lesions by the neurosurgeon.

Eggs of Schistosome induce granulomatous inflammation in infected tissue. The degree of inflammatory response to eggs in the CNS varies with the status of the immune system of the patient. Delayed cell-mediated immunity is an important factor in schistosomiasis (Warren, 1980). The continuous enzymatic and antigenic secretions of these eggs may induce a huge granulomatous cell-

mediated response of lymphocytes, eosinophils and macrophages, that is, approximately 100 times as much as the volume of the egg (Mahmoud, 1977). The present case revealed multifocal granulomatous inflammation due to infested eggs in cerebellar vermis and the roof of the fourth ventricle, which provided a diagnostic clue to cerebral schistosomiasis.

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