Spinal Cord Schistosomiasis: A Case Report and Review of the Literature

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Summary

Spinal cord involvement by Schistosomiasis is considered to be rare. We report a case of acute transverse myelitis due to Schistosoma mansoni in which treatment with praziquantal and steroids was started three days after the onset of a paraparesis, with complete neurological recovery.

Key words: Spinal cord disease; Schistosomiasis

Introduction

Schistosomiasis is considered to be a rare cause of an acute transverse myelitis. However, the true incidence of the disease is unknown, as it is rarely diagnosed and even more rarely treated effectively during life (Bird, 1964; El-Banhawy, 1971; Barnett, 1965), the diagnosis usually being made at autopsy. Symptomatic and asymptomatic nervous system involvement, particularly of the spinal cord, is probably more common than suspected, considering the striking infestation rates in endemic areas, and the estimate that about 200 million people throughout the world are infected with schistosomiasis (World Health Organization, 1965).

Case report

A 19-year-old male was admitted for investigation of recurrent acute urinary tract infections. A previous history of terminal haematuria, as well as a history of swimming in rivers while on holiday in an endemic schistosomiasis region 1 year and again just prior to admission, was obtained. Symptoms and signs of a urinary tract infection were present on admission, and a Klebsiella species was cultured from the urine and successfully treated. No parasite ova were found on urine microscopy. The serum leukocyte count was 14 200/mm³ with 60%

neutrophils and 3% eosinophils. Following treatment an excretory urogram demonstrated features consistent with chronic pyelonephritis and chronic cystitis. A subsequent bladder biopsy revealed viable ova of *Schistosoma mansoni*.

While still under investigation, the patient developed an acute spastic paraparesis (grade 3/5 power in an upper motor neurone distribution), and a sensory level from T10 to all modalities, with retained bladder and bowel function. A myelogram was performed and it was normal, but the cerebrospinal fluid had a raised protein (1.2 mmol/litre), normal glucose and chloride values, and 68 lymphocytes.

Viral Studies, stains and cultures for tuberculosis and fungi were negative. The Fluorescent Treponemal Antibody Test, Reactive Precipitin Reagen, and antinuclear factor were negative in the blood and cerebrospinal fluid.

The bilharzia enzyme linked immunosorbent assay (Elisa), Radio-Immunoassay and fluorescent antibody tests in the cerebrospinal fluid and serum were all positive. In view of these results treatment was started on the third day following the onset of paraparesis with praziquantal (50 mg/kg/day) for three days and prednisone 50 mg/day for the first week and then slowly withdrawn over four weeks (i.e., 40 mg/day on week two, 30 mg/day on week three, 20 mg/day on week four and 10 mg/day on week 5). On the seventh day of treatment the motor and sensory deficit began to improve. He was discharged from hospital after 6 weeks without any residual neurological deficit, and normal cerebrospinal fluid (Table I).

Table I Cerebrospinal fluid and blood parameters

Cerebrospinal fluid (csf) +	Days after the onset of the paraparesis			
	1	7	30	40
Protein	1.2	0.6	0.5	0.5
Neutrophils	1	0	0	0
Lymphocytes	68	26	8	4
Bilharzia Elisa + + CSF	0.625	0.411	0.385	0.365
blood	0.441	0.405	0.359	0.316
Bilharzia RIA + + + CSF	3	2	2	2
blood	3	2	2	2
Bilharzia fluorescent Csf	+	—	—	—
Test (IgM) blood	+	—	—	—
+ Cerebrospinal fluid chloride & glucose were always normal				
+ + Less than 0.21 negative				
0.22–0.40 low positive				
0.41–0.74 mid positive				
0.75 and greater high positive				
+ + + 0 negative				
1–4 positive				

Discussion

Since the first report of spinal cord involvement by Schistosomiasis (Shimamura and Tsunoda, 1906), there have been numerous reviews of the literature. Herskowitz (1972) found 104 cases of central nervous system schistosomiasis, 25 of which were limited to the spinal cord. Norfray (1978) found 43 cases of spinal schistosomiasis in the world literature.

To date we have found 64 cases reported in the literature (Table II) (Norfray,

Table II The total number of reported cases of spinal schistosomiasis, the types of organisms involved in the various modes of presentations, and the sex ratio

Type	Number of cases	Modes of presentation					Sex ratio	
		Granuloma	Myelitis	Radiculitis	Anterior spinal artery occlusion	Unknown	Male	Female
S. Mansoni	41	16	19	0	1	5	34	7
S. Haematobium	13	5	6	2	0	0	10	3
Mixed mansoni & haematobium	1	0	1	0	0	0	1	0
S. Japonicum	1	0	1	0	0	0	1	0
Unspecified	8	6	0	0	0	2	7	1

Schlachter, Heiser, Weinberg, Jerva, Wizgird, 1978; Rosenbaum, Ishii, Tano-witz, Wittner, 1972; Neves, Marinho, De Araujo, Raso, 1973; El-Banhawy, Elwan, Tahar, 1972; Queiroz, Nucci, Facure, Facure, 1979; Dar, Zimmerman, 1977; Pool, 1978–1979; Lechtenberg, Vaida, 1977; Luyendijk, Lindeman, 1975; Wood, Srolovitz, Schetman, 1976; Saxe, Gordon, 1975; El-Banhawy, 1971; Scaff, Riva, 1971; Nozias, Pichard, Develoux, Gentilini, 1983; Chait, Gannon, 1969; Warren, 1973; Eftimiou, Denning, 1984; Adeloye, Folami. Aghadiung, 1982; Levy, 1970; Ghaly, El-Banhawy, 1973; Siddorn, 1978; Ibrahim, 1941; Sparrow, 1966; Arean, 1966; Scully, Mark, McNealy, 1985).

Spinal involvement in schistosomiasis is usually due to schistosoma haematobium or mansoni, schistosoma japonicum more commonly affects the brain. Reaction to the eggs rather than the adult worms are the cause of the major neurological syndromes, with the diversity of the host's immune response accounting for the various types of presentations (Wilcocks, Manson-Bahr, 1972; Gelfand, 1973; El-Banhawy, 1972).

In cases presenting as an Acute Transverse Myelitis, multiple small granulomata are found, each surrounding one or more ova lying together in the spinal cord (Levy, 1970). Sometimes instead of many small granulomata, a large solitary granuloma forms. It presents as an intraspinal mass lesion and appears grey and granular, closely resembling a glioma macroscopically (Cohen, Capildeo, Rose, Pallis, 1977). Unless schistosomiasis is suspected the diagnosis is only made histologically following surgical biopsy.

A third form of presentation is the radicular form, with granuloma forming around spinal roots or the cauda equina. This form is usually associated with the intra-spinal granuloma, and there is some doubt of its independent existence (El-Banhawy, 1971; Ghaly and El-Banhawy, 1973).

Finally, vascular syndromes may occur. One well documented case (Siddorn, 1978), and possibly a second (Bird, 1965), of anterior spinal artery occlusion have been reported. Lymphocytic infiltration with intimal thickening and fibrinoid degeneration of spinal vessels have been reported (Marciel-Rojas and Fiol, 1963; Maciel *et al.*, 1954). Another possibility is that ova are disseminated, as arterial emboli from the pulmonary circulation, explaining the cases with skin (Wood *et al.*, 1976; Saxe & Gordon, 1975) and conjunctival (Ibrahim, 1949; Sparrow, 1966) involvement. Some authors have surmised that there is actual oviposition in situ by aberrant worms which have made their way into the vessels of the spinal cord (Jane *et al.*, 1970; Raper, 1948). With the exception of one case no adult worms have been found inside the spinal cord. The most

common areas of the spinal cord affected are the lower thoracic or lumbar spine. Only one case involving the cervical cord has been reported (Molyneux *et al.*, 1978). This predilection is probably explained by the free anastomosis between the pelvic veins and the valveless vertebral plexus (Batsons Plexus) (Batson, 1940) as well as between haemorrhoidal and systemic veins.

Diagnosis

Because of the variability of presentation, a high index of suspicion should be maintained in patients presenting with spinal signs, and who live, or have visited endemic areas. Numerous consecutive urine and faecal examinations for ova should be made, as eggs are discharged intermittently, and to a less extent in chronic cases (Norfray, *et al.*, 1978; Areal, 1966). Eosinophilia, particularly in chronic cases, may be absent. The cerebrospinal fluid is occasionally normal, but more commonly reveals a lymphocytosis and elevated protein. A raised eosinophil count has rarely been noted. Rectal, bladder or liver biopsies may be required, but highly specific non-invasive immunological tests are now available with high sensitivity and specificity (Nillyer *et al.*, 1979; Ruiz-Tiben *et al.*, 1979). The immunological tests used are the double diffusion test (DDT), circumvocal precipitin test (COP), enzyme linked immunosorbent assay (Elisa), radioimmuno assay (RIA). The various skin tests are outdated because of their relative insensitivity, as well as the numerous cross reactions that occur with other trematode infections. So by using both the Elisa (92% specificity and 75% sensitivity) and RIA (79% specificity and 95% sensitivity) in our patient, a high degree of both specificity and sensitivity was achieved.

Treatment

Praziquantal has revolutionised the treatment of schistosomiasis. It is highly effective against all three types of schistosomiasis (Kardaman *et al.*, 1983; Pearson, 1983). Praziquantal appears to increase permeability of the plasma membrane of susceptible worms to calcium ions, resulting in a loss of intracellular calcium and contraction and paralysis of the worms (Pearson, 1983). Side effects are usually slight and include nausea, vomiting and abdominal pain. Cure rates range from 71–98% for *Schistosoma haematobium* and 66–94% for *Schistosoma mansoni*. Steroids have been used with conflicting results in cases with spinal involvement, as in most reported, therapy has been delayed for at least one week after the development of an advanced deficit (Lechtenberg and Vaida, 1977). Experimental studies by Domingo *et al.* (1967) indicate that the size of granulomata are considerably diminished by steroids. Bird is also a proponent of steroid therapy while Colley has questioned the use of corticosteroids in acute transverse myelitis. The use of steroids to counteract the allergic inflammatory reaction plus treatment with praziquantal directed at the adult worms would seem to be a reasonable therapeutic course.

Delay in treatment as in the case of a young man who went on a 3 month trip to Kenya (Scully *et al.*, 1985), usually results in a complete, irreversible paraplegia. Harries *et al.* (1986), in their controlled trial concluded that all expatriates

returning to Britain from endemic areas in Africa who may have been exposed to infection be screened for schistosomiasis. Positive serology requires further investigation and treatment if necessary, so that tragedies may be avoided.

Conclusion

Complete neurological recovery in our patient may be explained by the early commencement of treatment with steroids and praziquantal, prior to the onset of a complete paraplegia.

Because of the extent of schistosomal endemic areas, often with high infestation rates, the increased mobility of the world's population, and the frequent asymptomatic nature of the disease, schistosomiasis should always be considered in patients, presenting with spinal lesions who may have been exposed to the infestation. Vigilance and appropriate investigation will reveal the true extent of central nervous system schistosomiasis, and clarify the possible importance of early and specific treatment nowadays.

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