Intramedullary Spinal Cord Schistosomiasis

Case Report

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In endemic areas in Africa, Asia, and South America, man frequently is host
to trematodes of the Schistosoma genus. The infection appears in various or-
gans of the body, with different species favoring specific organs for egg-laying sites. Lesions of S. mansoni and S. japonicum are most pronounced in the intestinal tract and liver, whereas those of S. haematobium are most common in the urinary tract. Lesions have also been found in unusual sites such as the skin, the placenta,\(^{28}\) and the conjunctivum.\(^ {10,27}\) A recent review\(^ {21}\) showed that only a few proven cases of schistosomal disease of the central nervous system have been reported, and those in which S. haematobium has been clearly shown to be the causative agent for a spinal cord lesion are indeed rare.

That schistosomiasis (S. haematobium and S. mansoni) is rampant in Nigeria is a well-documented fact\(^ {9,16,17,22,23}\) but to our knowledge only one case of schistosomiasis of the spinal cord has been reported, and that was unverified.\(^ {29}\) We are reporting a case of intramedullary schistosomiasis due to S. haematobium which has been histologically verified in a young Nigerian. The patient made a complete recovery following combined laminectomy and medical treatment.

Case Report

A 13-year-old boy came wobbling into the Neurosurgery Clinic of the University College Hospital in Ibadan one morning with a complaint of progressive low back pain, weakness of both legs, and urinary hesitancy of 6 weeks' duration. He had injured his back slightly in a soccer game 2 weeks before the onset of symptoms. There was no history of abdominal pain, fever, or hematuria.

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Examination. The patient had spastic paraparesis, which was more severe in the right leg. There was no fasciculation, but some occasional jerky involuntary movements of the right leg had been experienced before the onset of the low back pain. There was mild atrophy of the right leg. The gait was ataxic and broad-based. The legs showed equal but decreased deep tendon reflexes; the lower superficial abdominal reflexes were absent but the upper reflexes were present and normal. Bevor's sign was not present. Plantar reflexes were also normal. Patchy and inconstant mild loss of pain and temperature sensations was noted over the L1-L5 dermatomes. Straight-leg raising, Spurling's sign, and Lasegue's sign were normal bilaterally. A 10° list to the left was noted on examination of the spine, and spasm of the dorsolumbar paraspinal muscles was present on both sides.

The blood contained 12.4 gm of hemoglobin and 6,400 white blood cells (WBC) per cu mm, with differential eosinophilia; the hemoglobin genotype was AS. Urinalysis was normal; no schistosome ova or red blood cells were found. The spine radiographs were normal. Lumbar puncture showed an initial pressure of 90 mm of water, with a slow but steady rise to 120 mm on bilateral jugular vein compression. Yet a complete block of the subarachnoid space at D12-L1 was clearly shown on the Myodil myelograms (Fig. 1). The clear, colorless spinal fluid contained 20 WBC (85% lymphocytes and 15% polymorphonuclear cells; there was no eosinophilia). The protein content was 900 mg/100 ml; the sugar, 26 mg/100 ml. Cultures of the cerebrospinal fluid were sterile for pyogenic and tuberculous microorganisms.

Operation. At laminectomy (D-11, D-12, and L-1) the dura sac was opened posteriorly in the midline to expose markedly
swollen segments of the spinal cord with a pale-gray moderately firm but nondiscrete intramedullary lesion. Our first impression was that of a diffuse gliomatous neoplasm. The intramedullary granuloma, as later confirmed by histological examination, was chiefly diffuse, and only a few small discrete nodular masses (pseudo-tubercles?) were seen scattered at the surface. The yellow color characteristic of schistosomiasis was not evident. Three very small pieces of tissue were removed for biopsy from nodular areas at the left posteromedian aspect of the cord at the 12th dorsal vertebra. The dural incision was supplemented by transverse extension and left open. The exposed spinal cord segment was protected with a layer of gelfoam, and the laminectomy wound closed in layers.

Histological sections (Fig. 2) of the tissue biopsied from the spinal cord mass were characteristic of schistosomal granuloma. Ova of *S. hematobium* were readily identified in the sections. No schistosome ova were found either in the urine or in stool. The urine specimens were collected around midday on 2 consecutive days; the centrifuged deposit was examined under low-power magnification of the microscope. Stool specimens were examined by the glycerine-ether concentration method. Although the limited parasitological tests were negative, the circumoval precipitin test was strongly positive.

The patient received a course of niridazole (Ambilhar, Ciba), 250 mg orally three times per day for 12 days with supportive daily high fluid intake and promethazine Phenergan) 10 mg orally three times per day.

**Postoperative Course.** The postoperative course was satisfactory. Within 6 weeks of operation and medication, the patient’s gait had returned virtually to normal and only mild L-4 hypalgesia was detectable. The recovery of neurological functions has since become complete, although slight atrophy of the right leg persisted. At 18 months after laminectomy the patient was still well neurologically.

**Discussion**

Previous authors have repeatedly recalled the possible routes in the journey of the schistosome, from its penetration into the skin to its final ectopic settlement within the spinal canal and cord as ova or, occasionally, the adult fluke. From the portal-caval system, the hemorrhoidal-pelvic-vertebral venous plexi are believed to be favored pathways, a “valveless intercommunicating channel,” as aptly termed by Faust. Hutton and Holland discussed the possibility of “diffuse and irregular bombardment (*S. haematobium*) of the cord via the vertebral venousplexus.” They expressed concern that ova are not found frequently enough in the cord to strengthen this possibility. It may well be that other factors, such as preexisting trauma to the cord with some disruption of tissue, are also essential for the ova to take hold by reactive tissue changes once they gain access to the cord.

In schistosomiasis, the criteria for clinical diagnosis have been examined by Gelfand. He noted that Kabure bather’s itch (caused by passage of cercaria from the water) and
Katayama syndrome (including fever, joint swelling, urticaria, enlarged spleen or liver) are not frequently seen in the African patient. In tropical Africa, peripheral blood eosinophilia is too often associated with various parasitic infestations to be of particular diagnostic value. A history of exposure, such as swimming in infested water, serves merely to suggest the possibility of the disease. Among school children in an endemic area, terminal hematuria is the most characteristic symptom of vesical schistosomiasis. Various immunological investigations (skin and serological tests) may assist in establishing the diagnosis. A positive biopsy of rectal or urinary bladder mucosa or the presence of live ova in the urine or feces is considered conclusive evidence of current infection. Improvement under anti-schistosomal drugs also makes the diagnosis reasonably sure.

Presumptive clinical diagnosis of myelopathy due to *S. haematobium* has been made more often than it has been proven. On the other hand, ova may be present in the spinal cord without significant reactive changes.³ Ova of *S. haematobium* have also been recovered from digested spinal cord segments¹¹ and in at least one case without clinical manifestation of spinal cord involvement.

Marcial-Rojas and Fiol¹² illustrated how seldom schistosomes or their ova significantly compromise the human spinal cord in the clinical course of schistosomiasis.

Two such cases were due to *S. japonicum*, eight to *S. haematobium*, and 17 to *S. mansoni*. Two of the eight *S. haematobium* cases had lesions in the brain as well; six of these cases were histologically verified.

In the case of Hutton and Holland,¹⁸ the authors raised some doubt in determining the specific schistosome, but their judgment favored *S. haematobium*. A search of the literature available to us has added only two more verified cases to this short list. The present proven case represents the ninth in the *S. haematobium* group known to us (see Table 1). We have excluded the incompletely documented case which Marcial-Rojas and Fiol¹² attributed to Ferguson.¹² A few other cases reported give only indirect evidence, such as positive rectal or bladder mucosal biopsy, or the presence of ova in

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Fig. 2. *Left*: Numerous ova of *Schistosoma haematobium* in cross section in intramedullary granuloma of spinal cord. H. & E. ×250. *Right*: Terminal spine demonstrated in ovum sectioned longitudinally. H. & E., ×750.
<table>
<thead>
<tr>
<th>Author, Year, Country</th>
<th>Age, Sex (yrs)</th>
<th>Clinical Presentation</th>
<th>Clinical Pattern</th>
<th>Type of Lesion</th>
<th>Peripheral Blood Eosinophilia</th>
<th>Cerebrospinal Fluid</th>
<th>Histological Evidence</th>
<th>Treatment</th>
<th>Result</th>
</tr>
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<tbody>
<tr>
<td>Day and Kenawy 1936 Egypt</td>
<td>10 M</td>
<td>sudden flaccid paraplegia with sensory loss; urinary incontinence</td>
<td>acute lumbar myelitis</td>
<td>? diffuse</td>
<td>no normal</td>
<td>30</td>
<td>ova in cord (autopsy)</td>
<td>—</td>
<td>died (sepsitcemia)</td>
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<tr>
<td>Bayoumi 1939 Egypt</td>
<td>20 M</td>
<td>root pains and waist girdle pain; paresis of legs; incontinence</td>
<td>low lumbar transverse myelitis</td>
<td>(½ ins. above conus) chronic fibrotic mass</td>
<td>? no excess</td>
<td>30 not done</td>
<td>ova in cord (autopsy)</td>
<td>—</td>
<td>died</td>
</tr>
<tr>
<td>Gelfand 1950 Rhodesia</td>
<td>50 F</td>
<td>spastic paraplegia</td>
<td>acute transverse myelitis</td>
<td>?</td>
<td>6</td>
<td>45</td>
<td>no block</td>
<td>ova in digested cord (autopsy)</td>
<td>—</td>
</tr>
<tr>
<td>Pepler &amp; Lombaard 1958 South Africa</td>
<td>8 M</td>
<td>low back &amp; thigh pains; paraparesis</td>
<td>lumbar &quot;spinal cord tumor&quot;</td>
<td>granuloma mass</td>
<td>— 1 lymph</td>
<td>3750</td>
<td>L1 total block</td>
<td>surgery, Triostam</td>
<td>improved</td>
</tr>
<tr>
<td>Hutton &amp; Holland 1960 (Uganda) S. Rhodesia, Tanganika</td>
<td>73 M</td>
<td>fall with back &amp; hip injury; shooting leg pains; flaccid paraparesis; sensory loss urinary incontinence</td>
<td>thoraco-lumbar myelitis</td>
<td>diffuse granulomatosis</td>
<td>yes 55 lymph</td>
<td>90</td>
<td>ova in cord (autopsy)</td>
<td>—</td>
<td>died</td>
</tr>
<tr>
<td>Author, Year, Country</td>
<td>Age, Sex (yrs)</td>
<td>Clinical Presentation</td>
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<td>Barnett 1965 South Africa</td>
<td>35 M</td>
<td>low back pain; Katayama syndrome; paraplegia with sensory loss; chronic urinary infection</td>
<td>lumbar (L1) cord tumor</td>
<td>large granuloma mass</td>
<td>yes</td>
<td>20 (lymph 18)</td>
<td>60</td>
<td>D12-L2 indention</td>
<td>ova in cord (biopsy)</td>
</tr>
<tr>
<td>Bird 1965 South Africa</td>
<td>34 M</td>
<td>fall, injury with low back pain, bilateral sciatica; paraparesis; urinary retention</td>
<td>radiculitis (cauda equina)</td>
<td>tumor granuloma</td>
<td>(yes)*</td>
<td>(15)* lymph</td>
<td>(81)* normal</td>
<td>ova in cauda equina root (biopsy)</td>
<td>laminectomy and sodium antimonyl gluconate + leucanthone hydrochloride + corticosteroids</td>
</tr>
<tr>
<td>Odeku, et al. 1968 Nigeria</td>
<td>13 M</td>
<td>fall, injury low back pain; spastic paraparesis</td>
<td>subacute dorsolumbar myelitis</td>
<td>diffuse granulomatosis</td>
<td>yes</td>
<td>20 (lymph 17)</td>
<td>900</td>
<td>D11-12 total block</td>
<td>ova in cord (biopsy)</td>
</tr>
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* Obtained after laminectomy.
the urine or feces in patients with impaired spinal cord function. Although immunological tests may indicate probable infection or previous exposure, the species cannot be identified since the tests are not type specific.20,24 Bird,5 in an analysis of nine cases from South Africa, gave the details of only one case (see Table 1), positive for S. haematobium on biopsy from the cauda equina. One other biopsy failed to yield any ova or the adult worm. In still another case where the sections of the spinal cord at autopsy revealed schistosome ova, their identification as S. haematobium was not achieved because spines were not seen in the plane of section. In the case reported by Raper,26 it was not possible to identify specifically the calcified extradural adult schistosome found at laminectomy.

Human infestation with S. haematobium is more heavily endemic than with S. mansoni in western areas of Nigeria,9,10 and the more common urinary problems arising from it are well documented.1,16,17 Yet our case represents the first well-established instance of schistosomiasis in the spinal cord in a Nigerian.

As in nearly all other cases, our Nigerian patient was a male. He showed eosinophilia in the peripheral blood, pleocytosis without eosinophilia in the CSF, and a marked increase in the protein content of the fluid (900 mg/100 ml). Some authors17,20 found no eosinophilia in the blood and no pleocytosis in the CSF.

Aside from his purely neurological difficulties, our patient showed no systemic or organ evidence of the disease. As previously observed by Faust,12 it is possible that clinical indications in other organs had appeared much earlier and had been resolved. In a number of patients, trauma (such as a fall) has been noted as a precipitating factor. Other differential considerations in a tropical environment include tuberculous epidural granuloma, pyogenic epidural or fungal abscess (due to Histoplasma duboisii), and tropical ataxic paralysis believed to be the cumulative effect of increased intake of cyanogenetic compounds in the diet. Our case was essentially one of diffuse myelopathy with elements of the schistosomal radiculitis emphasized by Bird.5 No large discrete granulomatous lesion was present at the exposed segments of the spinal cord. Complete neurological recovery was obtained by operative decompression, immediately followed by a course of niridazole. Laminectomy was mandatory because of the complete myelographic block in the subarachnoid space and the high protein content of the CSF.

In diffuse granulomatosis, a small but adequately selected biopsy should be sufficient for examination. If the cord is diffusely swollen the dura should be left open; anti-schistosomal drug is then employed to combat the infestation. Large granulomatous masses require great restraint in their surgical handling. Barnett2 cautioned against a radical approach when the disease is suspected, since further damage to the cord during the operation may prove to be devastating and permanent. Simple surgical decompression is timely when done for some immediate relief, particularly where neurological deficits are progressing rapidly and myelographic study indicates the presence of an intraspinal canal mass. Laminectomy also affords an opportunity for a direct positive proof by biopsy.

One should not forget that the basic and most effective way to combat the disease is with anti-schistosomal drugs. Marcial-Rojas and Fiol21 believe, as do many others, that where indirect evidence leads to suspicion of schistosomiasis as the probable underlying process of spinal cord impairment, an appropriate anti-schistosomal regime should be instituted. It is essential that treatment begin early; by the time of complete loss of function, recovery or improvement is unlikely no matter what the treatment.22

Summary

We have presented a rare instance of schistosomiasis (S. haematobium) of the spinal cord in a Nigerian African boy who completely recovered from severe neurological deficits under the combined treatment of decompression by laminectomy followed by anti-schistosomal medication. We have also reviewed other comparable reports of this condition.

References

2. BARNETT, A. M. Bilharzial granuloma of the