Intestinal and Hepatosplenic Schistosomiasis: Case Series and Review of the Literature

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Abstract

Schistosomiasis is a parasitic infection that is estimated to afflict 200 million people worldwide. Most of these cases are found in Africa, the Middle East, South America and Asia. The primary sites for the infection are the intestines and the liver. This article reviews the literature on the natural history and treatment of Schistosomiasis infections. The authors' experience with 167 consecutive cases is also described.

Key words: Schistosomiasis, intestinal, hepatosplenic.

Schistosomiasis is a parasitic infestation. Intestinal and hepatosplenic schistosomiasis are commonly caused by Schistosoma mansoni and japonicum, and rarely, by Schistosoma intercalatum and haematobium. Schistosomiasis is predominantly an infection of rural and agricultural communities. In the last few years many programs for developing water resources have been established in endemic areas with a pronounced concomitant increase in prevalence and intensity of schistosomiasis.

It has been estimated that over 200 million people in the world suffer from schistosomiasis, and several other millions are exposed to the infection. It affects child development and adult productivity. Infection occurs in areas where water is contaminated by stool or urine containing ova that develop in the intermediate snail host, and from which cercariae emerge. These penetrate human skin and subsequently develop into adult worms located in the venous plexuses.

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DOI: http://dx.doi.org/10.5915/22-1-13542
mainly Japan, China, Thailand and Korea. Intestinal schistosomiasis is a major health problem in endemic areas, and if not diagnosed and treated early, can lead to complications such as chronic intestinal schistosomiasis and hepatosplenic schistosomiasis, both of which carry a high morbidity and mortality.

Clinical manifestation

Schistosoma mansoni and japonicum infection can be divided into acute and chronic forms. The pathological manifestation is mainly due to the presence of ova and depends on intensity of infection, frequency and severity of exposure, and immunological status. The host granulomatous response to the egg is a form of delayed hypersensitivity and plays an essential role in the pathogenesis of schistosomiasis. The egg output accurately reflects the intensity of infection. ²

The acute form can be subdivided into three main forms:

1. Cercarial dermatitis (invasion stage)
This occurs within 24 hours following cercarial penetration of the skin (swimmer's itch).

2. Katayama Syndrome (toxemic phase)
This is the hypersensitivity stage four to six weeks after exposure. It coincides with maturation of schistosome and ova depositing. It is an acute febrile illness with diarrhea and conspicuous eosinophilia, which occurs with all schistosoma species but commonly with japonicum and mansoni, which we reported in five children from Saudi Arabia. ⁴

3. Bilharzial Dysentary (acute intestinal disease)
This results from egg deposition in the bowel wall. It starts about two months following infection with dysenteric-like symptoms of fever, anorexia and abdominal tenderness. Exacerbations occur every few weeks. It may last 6 to 12 months and if not treated, may progress to a chronic form.

The chronic form occurs usually in three forms:

1. Chronic intestinal form
This usually involves the large bowel and to a lesser extent the stomach and small bowel, including the appendix. Other parts of the alimentary tract can be involved, such as the gall bladder and pancreas. Autopsy studies in patients with early Schistosoma mansoni have revealed 19% of the egg load is located in the small intestine and 36% in the colon. If hepatic fibrosis is present, small intestinal involvement reaches 39% and colon, 28%. ⁷

In the large intestine viable ova produce an inflammatory reaction, granulomatous formation, ulceration, bleeding, papillomatose and subsequently, fibrosis, which give rise to the long sequelae of the disease, ⁶ causing abdominal pain, diarrhea, bleeding, anemia and protein losing enteropathy.

In our unit we have studied the symptoms of 167 patients with colonic schistosomiasis. The commonest symptom was abdominal pain, in 38.92%. (Table 1)

Few patients may present with abdominal mass due to pericolic or mesenteric granuloma, and these may present with intestinal obstruction. In our hospital during the last three years, three patients had surgery for intestinal obstruction due to complicated schistosomiasis. Five patients presented with appendicitis and two with cholecystitis. All had surgery and the histology showed that inflammation of the appendix and the gall bladder was associated with schistosoma eggs. (Figure 1)

The stomach and small intestine rarely are involved. If the small intestine is involved, the disease can lead to stunting of the villi and development of malabsorption-like syndrome. Schistosoma mansoni involving the small intestine has been diagnosed by endoscopic appearance and biopsies from the duodenum. ⁷

Schistosoma mansoni ova were recovered from endoscopic biopsies in a patient with severe schistosoma duodenitis who presented with haematemesis. Schistosoma ova were also recovered from duodenum, jejunum and ileum from surgical specimens in a patient who had intestinal obstruction due to schistosomiasis. ⁶

Stomach ulcers have been reported with Schistosoma japonicum, and multiple eggs were found in the biopsies of the stomach in these patients. ⁹

2. Hepatointestinal form
Thirty percent of the infected population with early schistosomiasis have significant hepatomegaly, especially children. This is usually a result of heavy infection and is unusual in mild cases. Hepatomegaly disappears with treatment and resolution of the infection.

3. Hepatosplenic form
This, the main complication, develops late in the disease process, probably occurring from 5 to 10 years after the initial infection. It is estimated that 10% of infected patients develop this form. Em-

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>No. of Patients</th>
<th>Percentage (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Non-specific abdominal pain</td>
<td>65</td>
<td>38.9</td>
</tr>
<tr>
<td>Diarrhea</td>
<td>50</td>
<td>29.9</td>
</tr>
<tr>
<td>Rectal bleeding</td>
<td>33</td>
<td>19.8</td>
</tr>
<tr>
<td>Alternate diarrhea and constipation</td>
<td>16</td>
<td>9.6</td>
</tr>
<tr>
<td>Constipation</td>
<td>3</td>
<td>1.8</td>
</tr>
</tbody>
</table>

Table 1. Symptoms in 167 Patients

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bolization of the ova to the liver leads to granulomatous formation and later, fibrous formation, resulting in periportal fibrosis (Symmer's fibrosis). This usually leads to the development of portal hypertension. The resultant esophageal or gastric varices bleed in about one-third of patients and is the main cause of death from this form.\(^{10}\)

Hepatocellular function is usually well preserved until late in the disease so that portal hypertension becomes evident long before the deterioration of liver function.\(^{11}\) Hypersplenism usually develops due to the enlarged spleen. Rarely, liver cirrhosis occurs in schistosomiasis following anoxia to the hepatocyte, usually as a result of massive gastrointestinal bleeding. (Chronic active hepatitis has been observed in liver biopsies of patients with hepatic schistosomiasis but the exact etiology is not yet established.) Portal hypertensive gastropathy, when developed, may also lead to bleeding from gastric congestion and erosions.

Schistosomiasis and malignancy

There is an established association between Schistosoma haematobium and carcinoma of the urinary bladder.\(^{12}\) Schistosoma japonicum is reported to play an etiological role in the development of malignancy. Amano\(^9\) from Japan has reported an association between Schistosoma japonicum and different types of adenocarcinoma of the stomach. Chai et al\(^{13}\) from China studied 454 colorectal carcinoma specimens, of which 289 were associated with Schistosoma japonicum infestation, with a history of diffuse schistosomal involvement of the large intestine and ten years or more of colitic symptoms. However, there were no definite reports on an association between colorectal cancer and Schistosoma mansoni.\(^6\) Dimmetee et al\(^{14}\) studied 98 Egyptian patients with carcinoma of the large bowel. Of these 17 had Schistosoma mansoni infestation in which detailed histologic studies revealed no outstanding features to distinguish parasitic from non-parasitic groups. We have studied 167 patients with schistosomal colonic disease, and six patients with schistosomal polyps in whom there was no evidence of development of metaplastic or carcinomatous changes.\(^{15}\)
There is an association between chronic hepatitis B liver disease and hepatoma, but until now there has been no association between chronic schistosomal liver disease and hepatoma.

**Diagnosis**

The diagnosis of schistosomiasis is easy to establish in endemic areas, usually based on finding the ova in stool by simple smear examination, concentration methods or quantitative techniques such as the Kato/Katz method. However, in the chronic form the passage of ova in stool is not constant. In such situations a diagnosis can be made by:

1. **Serological test**
   
   Haemoagglutination techniques might not always be positive and usually are not specific. The new developments in immunodiagnosis, radioimmunoassay and ELISA are promising.

2. **Radiology**

   An ultrasound of the liver has shown to be a good diagnostic tool in demonstrating periportal fibrosis. However, CT scan and MRI can also show evidence of fibrosis. Barium swallow may show esophageal varices, and portal venography and hepatic angiography may show evidence of portal hypertension. Conventional radiographic methods may be helpful in diagnosis of advanced schistosomiasis. Nuclear scanning may show hepatosplenic enlargement. Barium enema may reveal the presence of schistosomal polyps.

3. **Liver biopsy**

   This shows the characteristic ova and granuloma (Figure 2) but sometimes may be difficult to perform due to coagulopathy, which may be further aggravated by thrombocytopenia due to hypersplenism.

4. **Endoscopic appearance**

   Fiberoptic sigmoidoscopy or colonoscopy are very helpful in establishing a diagnosis, not only because their may reveal characteristic lesions, as in our patients (Table 2), but also because a biopsy from these lesions can be obtained.

5. **Colonic or rectal biopsy**

   This is a very important diagnostic test especially in chronic forms of intestinal schistosomiasis, as the passage of ova in stool is infrequent and scan-
Table 2. Endoscopic Findings in 167 Patients with Colonic Schistosomiasis

<table>
<thead>
<tr>
<th>Findings</th>
<th>No. of Patients</th>
<th>Percentage (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rectum:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Patchy mucosal congestion +</td>
<td>20</td>
<td>16.8</td>
</tr>
<tr>
<td>Petechiae</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Patchy erosions + tiny ulcerations</td>
<td>8</td>
<td>16.8</td>
</tr>
<tr>
<td>Colon:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Patchy mucosal congestion +</td>
<td>9</td>
<td>7.2</td>
</tr>
<tr>
<td>Petechiae</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Patchy erosions + tiny ulcerations</td>
<td>3</td>
<td>7.2</td>
</tr>
<tr>
<td>Rectum:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Patchy mucosal congestion +</td>
<td>26</td>
<td>21.6</td>
</tr>
<tr>
<td>Petechiae</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Patchy erosions + tiny ulcerations</td>
<td>10</td>
<td>21.6</td>
</tr>
<tr>
<td>Polyps:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Colon or Rectum</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Single</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>Two or three</td>
<td>2</td>
<td>3.6</td>
</tr>
<tr>
<td>Telangectasic-Like Lesions in</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Rectum or Colon</td>
<td>4</td>
<td>2.4</td>
</tr>
<tr>
<td>Normal Looking Mucosa</td>
<td>81</td>
<td>48.5</td>
</tr>
</tbody>
</table>

Management

Drugs

Different antimonial compounds, hycanthone and niridazole, were used. New antischistosomal drugs (Oxaminquine and Praziquantel) are safe and effective in treating acute schistosomiasis and in halting progress of the disease in chronic forms. Praziquantel is highly effective against all forms of human schistosomiasis. Based on the dose, the parasitological cure rate ranged between 75% and 78%. Oxaminquine is highly effective against Schistosoma mansoni with egg output reduced by 80% to 90%. In one series the cure rate in children was low (55%) compared with adults (87%). Oxtipraz was also found to be effective in Schistosoma mansoni infection.

Sclerotherapy

This is an effective method for arresting acute variceal bleeding and preventing recurrence of bleeding. Sclerotherapy has a special place in treat-

Figure 3: Endoscopic biopsy from colon showing schistosoma ova.
Forty-three patients had no recurrence of bleeding after sclerotherapy, making the mortality rate 8%. Six patients, three of whom died postoperatively, had complications including perforation, sepsis, and liver failure. The mortality rate following sclerotherapy was 8%.

Surgery
Several surgical procedures have been advocated for the treatment of variceal bleeding. Some reports have shown good surgical outcome in patients with hepatosplenic schistosomiasis. One from Egypt and a second from Saudi Arabia employed Hassab's operation, and a third from Brazil employed selective distal spleno-renal shunt. Surgical procedures may carry a high risk of morbidity and mortality when compared with sclerotherapy.

Endoscopic polypectomy:
In cases of schistosomal polyps this procedure will prevent rectal bleeding. Antischistosomal drugs should also be given to these patients.

References:


