Radiology's role grows in schistosomiasis mansoni

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Most infected individuals remain asymptomatic carriers, but hepatosplenic form of schistosomiasis can be associated with serious complications

Schistosomiasis, or bilharziasis, results from infection by one of several species of parasitic flukes of the genus Schistosoma. The World Health Organization describes schistosomiasis as a millenary disease, affecting over 200 million individuals living in 76 different countries across Asia, Africa, and the Americas. It is considered the most important helminthic human disease and constitutes a huge public health problem.

The species Schistosoma hematobium was discovered in 1851 by Theodor Bilharz. Just over 50 years later, another species of Schistosoma was discovered and subsequently classified as S mansoni. Schistosomotic patients are affected in many different ways. The majority of infected individuals are asymptomatic carriers of the parasite. At the other end of the spectrum, the hepatosplenic form of schistosomiasis can lead to upper digestive hemorrhage secondary to portal hypertension.1,2

The disease's evolutionary history comprises five major steps. Subjects must first be contaminated with an appreciable quantity of the parasite S mansoni. Live adult worms then affix themselves within the roots of mesenteric veins. Eggs deposited by females and dead worms will migrate toward intrahepatic portal roots in the liver. The resulting inflammatory reactions and scarring constitute Symmers fibrosis. The development of portal hypertension is next. The final stage includes formation of esophageal varices, hypertensive gastropathy and colopathy, and the advent of varices in other organs.

While the acute phase of schistosomiasis is usually asymptomatic, clinical signs of varying intensity may occur. Common manifestations include fever, chills, weakness, weight loss, headache, nausea, vomiting, diarrhea, hepatomegaly, splenomegaly, and marked eosinophilia. The toxemic form of schistosomiasis mansoni is regarded as unusual, especially in areas where this parasitosis is endemic. Reports cite patients who developed multiple small papulo-erythematous lesions scattered across the chest, related to systemic vasculitis. Patients with pulmonary involvement, manifested by micronodules disseminated in both lungs as well as a Loffler-like syndrome, have also been observed.3 We have seen acute schistosomiasis mansoni patients presenting with pulmonary involvement that appeared as ground-glass opacities and septal thickening on high-resolution CT (Figure 1).

The most common presentation of chronic schistosomiasis is the intestinal form, involving the rectal and sigmoid colons. It usually manifests as bowel irregularity of little relevance. Most colitis caused by schistosomiasis mansoni is asymptomatic or oligosymptomatic. Some patients may present with colonic stenosis (Figure 2) and intestinal obstruction. Development of the hepatointestinal form depends on patients' worm burden and nutritional status as well as immunological factors and treatment choice.

Hepatosplenic schistosomiasis, the most severe presentation, is characterized by extensive splenomegaly, periportal fibrosis, portal hypertension, and upper digestive bleeding caused by ruptured esophageal varices. The principal antigenic factor behind hepatic fibrosis is the presence of S mansoni eggs in the portal veins. This causes a granulomatous inflammatory reaction, leading to the formation of fibroid tissue. Subsequent development of an intrahepatic vascular neoformation around the portal ramifications maintains blood flow and preserves hepatic cells. Patients with schistosomiasis consequently do not present with hepatic insufficiency.

Development of portal hypertension, regardless of its etiology, is due to increased vascular resistance or increased portal venous flow, or both. The pathophysiology of portal hypertension also frequently displays a systemic hyperdynamic state due to portal shunting of splanchnic vasoactive mediators.4

Periportal fibrosis, also known as Symmers fibrosis (Figure 3), constitutes a pathognomonic lesion of this chronic form.5 The WHO classification of hepatic fibrosis, established in 1993, is based on
measurements of the periportal tract, 0: >3 mm; I: 3 to 5 mm; II: 5 to 7 mm; and III: > 7 mm.5 Ultrasound makes an important contribution to diagnosis of hepatosplenic mansonic schistosomiasis. Recognizable features include hyperechoic fibrotic bands along the portal vessels, volumetric reduction of the right hepatic lobe, left lobe enlargement, perivesicular fibrotic thickening, splenic nodules, and splenomegaly. The hemodynamic status of the portal venous system, including identification of collateral veins and portosystemic venous shunts, may be evaluated by Doppler analysis (Figure 4). Portal vein thrombosis may be present in a few cases.6 CT depicts periportal fibrosis as rings of low attenuation around portal vein branches throughout the liver. Enhancement is marked following intravenous administration of contrast. Periportal bands are isointense relative to normal liver parenchyma on T1-weighted MRI and hyperintense on T2-weighted MRI. Contrast administration again leads to marked enhancement.7 Mansonic schistosomiasis patients with portal hypertension and hepatofugal venous blood flow will also probably experience hemodynamic changes to the ocular circulation. One study showed that the arrival of fluorescein contrast to the retina was delayed by at least 70 seconds in three (12%) of a group of young patients with the hepatosplenic form of schistosomiasis. No retardation was observed in a control group. Although the difference in mean contrast arrival time between the two groups was not statistically significant, these findings lend support to hypotheses regarding delayed venous blood flow drainage in the eye.8 Portal colopathy is an important finding in hepatosplenic schistosomiasis, and it could be the cause of lower gastrointestinal bleeding in patients with severe portal hypertension. Characteristic signs of portal colopathy in schistosomiasis are similar to those of portal colopathy in cirrhotic patients, though they tend to be more frequent and accentuated. They include increased vascular bed, edema, congestion, scattered “flea-bite” petechial spots, ulceration, and polyps.9 An evaluation of 22 adolescents and young adults with hepatosplenic schistosomiasis associated with bleeding esophageal varices observed that patients had significant deficits in puberal and genital development and in testicular volume (especially the left testicle). The researchers also found a high prevalence of varicocele (61.5%) with no direct association with testicular atrophy.10 Other researchers have demonstrated that patients with hepatosplenic schistosomiasis may present with dilated ovarian veins. Doppler ultrasound can show an increase in the diameter of these vessels and a reduction in flow.

**ATYPICAL MANIFESTATIONS**

Intestinal polyps are an atypical manifestation of schistosomiasis. A retrospective review of 500 autopsy cases found 21 subjects with polyps (38.8% schistosomotic).11 A separate publication has detailed the case of a 35-year-old woman with hepatosplenic schistosomiasis, portal hypertension, and esophageal varices who presented with intestinal bleeding of recent onset. Colonoscopy revealed four hyperplastic polyps with mild to moderate inflammatory infiltrations around egg shells of S mansoni.12 Intestinal obstruction and even perforation are infrequently associated with schistosomiasis mansoni. Obstruction may be related to stenosing granulomatous processes and extensive pericolonic infiltration.13-16 Interstitial or granulomatous pulmonary schistosomal disease corresponds to granulomatous vasculitis excited by widespread deposition of ova within the pulmonary vasculature. Radiography shows interstitial infiltrates (typically nodular or micronodular) and sometimes frank fibrosis or typical lung fibrosis.17 Radiographic patterns mimicking acute tuberculosis and lung neoplasms have also been reported. CT findings have been correlated with histopathological appearance in S mansoni patients with chronic, stable lung involvement.17,18 Pulmonary hypertension develops in 7% to 23% of patients with chronic pulmonary schistosomal disease. This is due to granulomatous endarteritis that obliterates pulmonary arterioles and capillaries and leads to secondary fibrotic changes in the lung parenchyma. These progressive changes may occur over several decades. Young adults with hepatosplenic schistosomal disease may be lacking in bone mineral content. Medical treatment of the parasite, followed by surgical treatment of portal hypertension, is associated with an improvement in bone mineral content. Brandt at al demonstrated a significant linear association between the histomorphometric degree of Symmers fibrosis and bone mineral deficit. No patients with marked fibrosis presented with normal bone mineral content, while no patients with slight portal fibrosis presented with osteoporosis.19 Adult S mansoni worms usually lodge in venules of the colorectal mucosa and submucosa. This localization means that ova are found frequently in the colon and liver. Development of portal hypertension and establishment of a portacaval shunt can lead to ova being found in the lungs. The discovery of schistosome eggs or worms in other organs is rare and can be associated with serious
clinical complications. Schistosomal myeloradiculopathy is the most severe ectopic form of schistosomal infection. It is more frequently associated with the intestinal and hepatointestinal chronic forms of schistosomiasis. Clinical diagnosis may be strongly suggested when a patient from an endemic area presents with rapidly progressing signs and symptoms of myelitis involving the lower spinal cord segments, usually in association with involvement of the cauda equina roots. Neither myelography, CT myelography, nor spinal MRI shows specific findings. The most frequent abnormalities are spinal cord enlargement (Figure 5) and enlargement of the conus medullaris, which may or may not be associated with thickening of the cauda equina roots. MRI sometimes shows areas of hypo/hyperintense signal, with contrast enhancement but no changes in the diameter of the spinal cord.20 Cerebral neuroschistosomiasis often presents as a slowly expanding intracranial lesion. Clinical manifestations depend mainly on the lesion site and on the increase in intracranial pressure caused by the mass effect of granulomas. Headache, seizures, papilledema, visual abnormalities, speech disturbances, sensory impairment, hemiparesis, nystagmus, and ataxia are common manifestations. CT usually shows a hyperdense enhancing area surrounded by a hypodense halo (edema) and an associated mass effect. Multiple focal lesions may be seen as well. MRI demonstrates one or more areas of hypointense and hyperintense signal with contrast enhancement.20 The overall incidence of glomerular disease with schistosomiasis has been shown to be about 5% to 6%. The hepatosplenic form of S mansoni infection may be accompanied by glomerulopathy in 12% to 15% of cases.21 Infection is characterized clinically by variable proteinuria ranging from asymptomatic to nephrotic syndrome. Immunofluorescence and electron microscopy reveal the presence of immune complexes containing immunoglobulins IgM, IgG, IgA, and IgE. They will also show schistosomal antigens in the mesangium and along the endothelial side of the capillary wall. Renal histological changes will sometimes precede clinical manifestations. Cardiac involvement in schistosomiasis is typically secondary to pulmonary involvement. A possible association between endomyocardial fibrosis and schistosomiasis has been investigated.22 The breast is a very rare site of schistosomiasis manifestation. Lima et al reported the first case of a breast lesion caused by S mansoni in a 23-year-old woman. The clinical appearance and ultrasound findings were suggestive of fibroadenoma.23 Cutaneous schistosomiasis is a rare complication. A literature review conducted in 1998 turned up 25 cases of ectopic cutaneous schistosomiasis mansoni. The lesions were usually asymptomatic and most commonly found in the anterior thorax and abdomen.24 It is estimated that at least 5% of girls and women with intestinal schistosomiasis also have genital lesions at an age when sexual activity peaks. The ovaries are most commonly affected, followed by the cervix, fallopian tubes, and uterus. Vulval lesions account for 6% of all case reports. Schistosomiasis of the cervix, with or without concomitant human papilloma virus, must be considered as a risk factor for the development of cervical cancer.25 In the male genital tract, epididymitis can be related to schistosomiasis. S mansoni infection can also cause orchitis and has long been associated with malignant neoplasia. A total of five cases of prostatic adenocarcinoma and concomitant gland schistosomiasis have been described in the literature.26 References 1. Coura JR, Amaral RS. Epidemiological and control aspects of schistosomiasis in Brazilian endemic areas. Mem Inst Oswaldo Cruz 2004;99(5 Suppl 1):13-19. 2. Lambertucci JR, Cota GF, Pinto-Silva RA, et al. Hepatosplenic schistosomiasis in field-based studies: a combined clinical and sonographic definition. Mem Inst Oswaldo Cruz 2001;96(Suppl):147-150. 3. Lambertucci JR, Rayes AAM, Barata CH, et al. Acute schistosomiasis: report on five singular cases. Mem Inst Oswaldo Cruz 1997;92(5):631-635. 4. Cleva R, Saad WA, Herman P, et al. Portal hyperflow in patients with hepatosplenic mansonic schistosomiasis. Rev Hosp Clin Fac Med Sao Paulo 2004;59(1):10-14. 5. Magalhaes A, Miranda DG, Miranda RG, et al. Cytokine profile associated with human chronic schistosomiasis mansoni. Mem Inst Oswaldo Cruz 2004;99(5 Suppl 1):21-26. 6. Machado MM, Rosa ACF, Oliveira IRS, Cerri GG. [Ultrasound findings in hepatosplenic schistosomiasis]. Radiol Bras 2002;35(1):41-45. In Portuguese. 7. Cerri GG, Alves VA, Magalhaes A. Hepatosplenic schistosomiasis mansoni: ultrasound manifestations. Radiology 1984,153(3):777-780. 8. Souza ACD, Brandt CT, Ventura L, Orifice F. Retinal fluorescein contrast arrival time of young patients with hepatointestinal form of the schistosomiasis mansoni. Mem Inst Oswaldo Cruz 2004;99(5 Suppl 1):13-19.

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