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Concentric Rings: Updated CT Appearance of Hepatic Schistosomiasis Mansoni

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Schistosomiasis is an infectious disease affecting as many as 300 million people worldwide. *Schistosoma mansoni* infection is associated with gastrointestinal complications. Ultrasound has been used extensively to document characteristic periportal fibrosis of schistosomiasis, but computed tomograph (CT) images of the disease are more scarce. To our knowledge, multidetector CT imaging of hepatic involvement of *S. mansoni* has not been published. We present a case of a Brazilian woman with documented *S. mansoni* infection and secondary portal hypertension. CT images demonstrate concentric ring enhancement representing portal venous structures surrounded by periportal fibrosis and periportal inflammation. We also demonstrate chronic gallbladder wall thickening and nodularity that may represent granulomatous inflammation of the gallbladder, an unusual finding in schistosomiasis. We review the progress made in imaging of the disease over the past twenty years. Although schistosomiasis is not endemic to our region, increasing emigration and rural travel have made *S. mansoni* infection of increasing importance to radiologists worldwide.

Introduction

Schistosomiasis is an infectious disease affecting up to 300 million people worldwide [1]. Of the three main species of Schistosoma, *S. mansoni* has the broadest geographical distribution. It is endemic to portions of South America, sub-Saharan Africa, the Middle East, and the Caribbean. *S. haematobium* is found mostly in North Africa, parts of sub-Saharan Africa, and the Middle East; *S. japonicum* is endemic to Asia. Schistosomiasis presents with different manifestations depending on the offending organism. *S. mansoni* and *japonicum* tend to affect the intestines and the liver, *S. haemotobium* the kidneys and bladder [2].

It is worth noting that most patients infected with schistosomiasis, particularly those from endemic regions, remain asymptomatic and the diagnosis is thus based on imaging and other laboratory studies [3]. Booming industries such as rural tourism have also contributed to the rising incidence of schistosomiasis in non-endemic regions [3,4]. CT findings of *S. mansoni* hepatic involvement have not been extensively published; most series of case reports date back more than ten years. To our knowledge, multi-detector CT (MDCT) images have not been published.

We present a case of hepatosplenic *S. mansoni* infection that demonstrates a concentric ring pattern of periportal fibrosis previously described by ultrasound, but below the resolution of prior CT imaging. We show thickening and nodular changes in the gallbladder wall that may represent *S. mansoni* involvement of the gallbladder, a rare finding. We also present a brief updated review of the past twenty years in imaging of schistosomiasis.

Case Report

A 44-year-old Brazilian woman with documented history of *S. mansoni* infection with esophageal varices was seen in the Gastroenterology Clinic for evaluation of abnormal liver function tests (LFTs) and recent nausea and vomiting. The patient had grown up in Brazil and had been diagnosed with schistosomiasis while living there ten years ago. She believed she had been treated with praziquantel at that time. She had undergone a splenectomy for splenomegaly nine years ago and subsequently had immigrated to this country. She had multiple endoscopies performed over the

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Abbreviations: MDCT, multi-detector CT, LFT, liver function test, AST, aspartate aminotransferase, ALT, alanine aminotransferase, IgG, immunoglobin G, ELISA, enzyme-linked immunosorbent assay, IVC, inferior vena cava

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past seven years, demonstrating stable portal hypertensive gastropathy, gastric varices, and grade 3 esophageal varices. She took propranolol for portal hypertension. She denied any alcohol use.

On physical exam in the clinic, there was no jaundice or hepatomegaly. Laboratory studies revealed stably elevated LFTs with aspartate aminotransferase (AST), alanine aminotransferase (ALT), and ammonia levels at 1.5 times the upper limit of normal. Ceruloplasmin was within normal limits. Hepatitis B and C serologies were negative; Hepatitis A immunoglobin G (IgG) was positive but IgM was negative. Stool cultures were negative for ova and parasites. S. mansoni serology was positive by enzyme-linked immunosorbent assay (ELISA) using microsomal fraction of adult S. mansoni worm as antigen.

Abdominal ultrasound performed on day of clinic visit revealed a small liver with heterogeneously coarse echotexture. In a sagittal plane, there were multiple round echogenic areas present in both the right and left hepatic lobes (Figure 1). A central lucency was surrounded by these echogenic areas, consistent with the previously described "bull's-eye" appearance of periportal fibrosis [5]. A thickened gallbladder wall and recanalization of the umbilical vein were visualized, but there was no dilatation of intra- or extrahepatic biliary systems. No intraabdominal ascites was seen. Color Doppler imaging was negative for portal vein thrombosis or typical appearance of gallbladder varices [6,7].

CT imaging from two years prior had demonstrated periportal enhancement consistent with periportal fibrosis (Figure 2). These images were similar in quality to those of previous case reports demonstrating CT appearance of schistosomiasis [8]. Hypodense lesions were seen traveling with and surrounding the portal venous system; these lesions enhanced during the portal phase, presumably representing inflammation. The gallbladder wall was nodular and thickened to 4 mm. Note was also made at that time of fatty liver and recanalization of the umbilical vein and mesenteric vessels, consistent with portal hypertension.

MDCT scan at the time of current visit showed an irregular liver surface. In the arterial phase, the hepatic arterial branches were seen as enhancing branching structures traveling adjacent to hypodense areas (Figures 3A and B). The hypodense areas represented both periportal fibrosis and the portal venous system prior to contrast enhancement. During the portal phase, the portal venous branches enhanced strongly, and were surrounded by two concentric rings of hypoattenuation and enhancement (Figures 3C and D). The ring of hypoattenuation presumably represented areas of periportal fibrosis with decreased vascularity, whereas the enhancing rim was due to periportal inflammation. These concentric rings correlated with the "bull's eye" appearance of schistosomiasis on ultrasound. Other notable findings included an enlarged gallbladder with a nodular and thickened wall that extended to the porta hepatis. Stranding was seen around the gallbladder; there was no evidence of stones or biliary dilatation (Figure 4). The caudate lobe was noted to be mildly enlarged; the spleen had been removed, and the pancreas was atrophic. There were no varices definitively identified. The inferior vena cava (IVC) was enlarged to a maximal diameter of 3.8 cm. These findings were thought to be consistent with chronic *S. mansoni* infection.

Discussion

The schistosomal life cycle begins with penetration of intact skin by cercarial larvae living in fresh water. After entering the circulatory system, the larvae migrate through the mesenteric, splanchnic, and portal circulations and lodge in the liver. *S. mansoni* then migrates in a retrograde fashion into the venous plexuses of the intestine, where

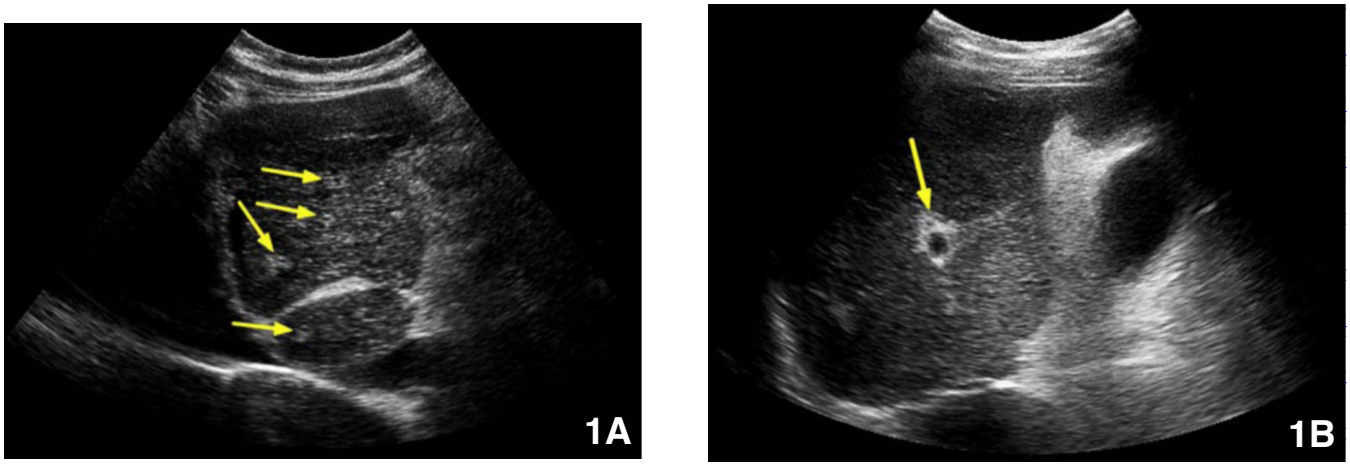


Figure 1. A. Abdominal ultrasound demonstrates a small liver with heterogeneously coarse echotexture. Multiple round echogenic areas with central lucency are seen in this sagittal image of the left hepatic lobe (arrows), consistent with "bull's eye" appearance of fibrosis surrounding portal venous structures. **B.** Close-up view of a round echogenic focus seen in the right hepatic lobe. Central lucency represents portal venous structure.

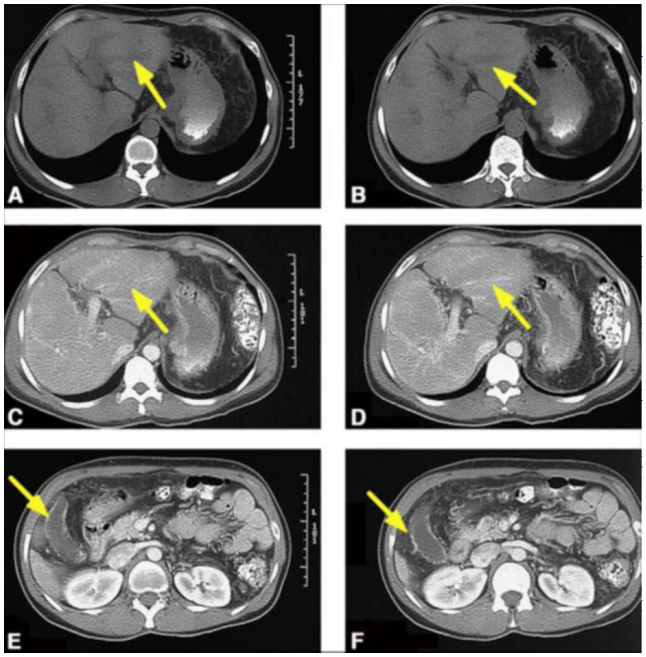


Figure 2. CT from two years prior demonstrating fatty liver and periportal fibrosis. These images are similar in appearance to those of previous case reports demonstrating CT appearance of schistosomiasis. **A, B.** Arterial phase images show hypoattenuated round and linear branching lesions traveling adjacent to enhancing hepatic arterial branches (arrows). **C, D.** These lesions enhance during portal phase (arrows). **E, F.** The gallbladder wall is nodular and thickened and measures 4 mm.

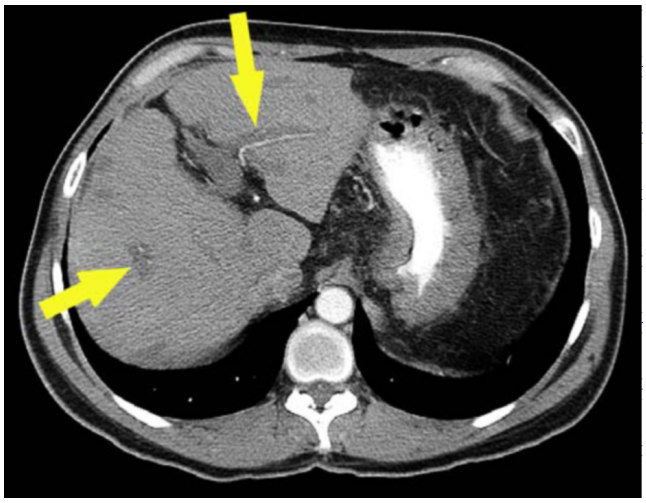
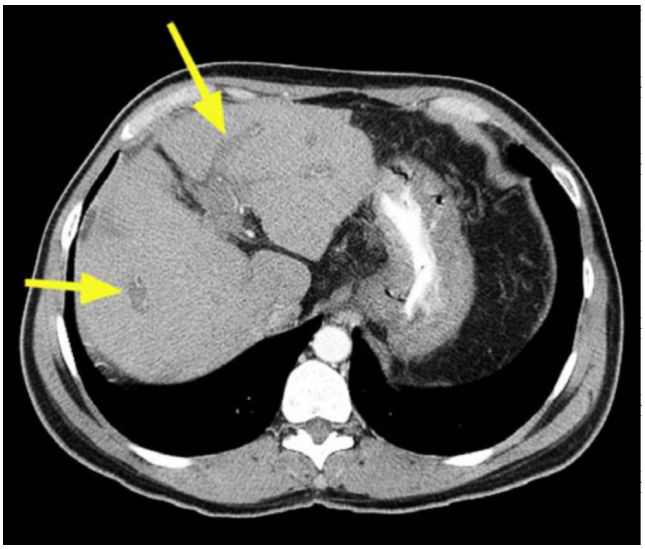


Figure 3B. MDCT for evaluation of abnormal LFTs, illustrating irregular liver surface and periportal fibrosis as seen in schistosomiasis. Arterial phase demonstrates hepatic arterial branches as enhancing structures with surrounding hypodensities (arrows). Hypodense areas represent both periportal fibrosis and the portal venous system prior to contrast enhancement.



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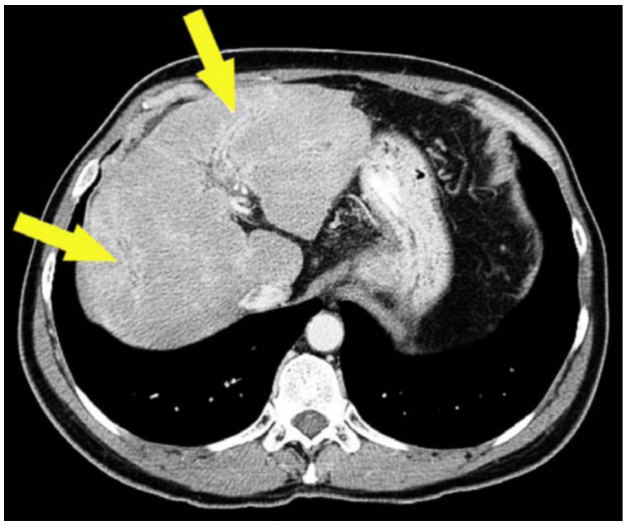

Figure 3C. MDCT for evaluation of abnormal LFTs, illustrating irregular liver surface and periportal fibrosis as seen in schistosomiasis. Portal phase demonstrates enhancement of portal venous structures surrounded by inner ring of hypoattenuation and outer ring of enhancement (arrows). Hypoattenuated ring presumably represents areas of periportal fibrosis with decreased vascularity, whereas enhancing rim may be due to periportal inflammation. These concentric rings correspond to the "bull's eye" appearance of periportal fibrosis previously described with ultrasound.

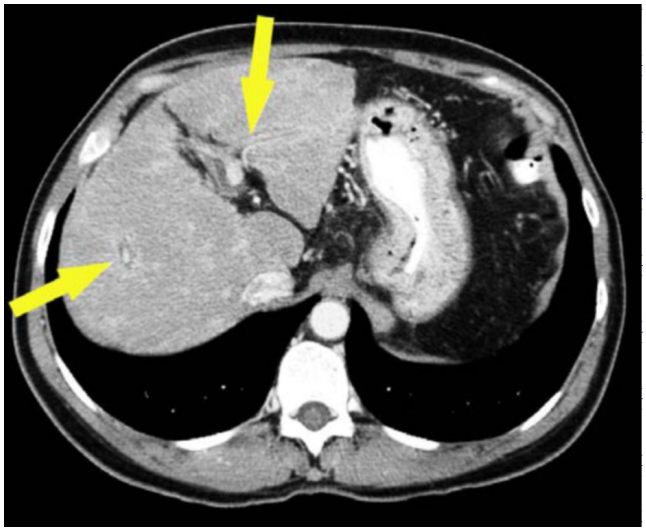

Figure 3D. MDCT for evaluation of abnormal LFTs, illustrating irregular liver surface and periportal fibrosis as seen in schistosomiasis. Portal phase demonstrates enhancement of portal venous structures surrounded by inner ring of hypoattenuation and outer ring of enhancement (arrows). Hypoattenuated ring presumably represents areas of periportal fibrosis with decreased vascularity, whereas enhancing rim may be due to periportal inflammation. These concentric rings correspond to the "bull's eye" appearance of periportal fibrosis previously described with ultrasound.

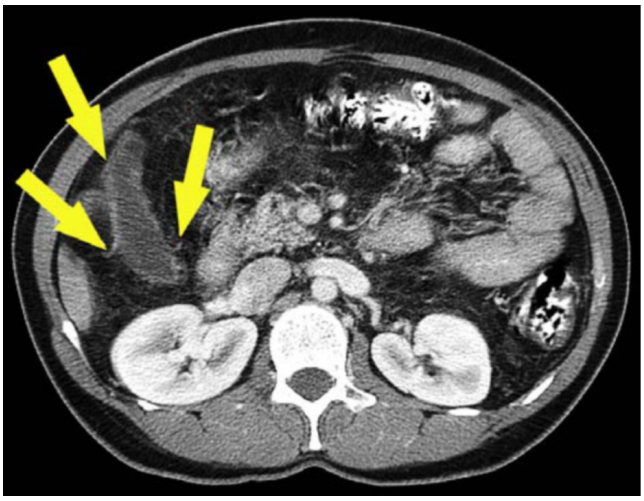

Figure 4. Additional CT findings of schistosomiasis. The gallbladder demonstrates chronic wall thickening and nodularity (arrow), present since two years prior to this study. There is stranding surrounding the gallbladder (arrowheads). There are no stones or sludge, pericholecystic fluid, or dilatation of the biliary structures.

adult helminths may live for years. The female worm here produces ova that are excreted in the feces into fresh water, in search of a snail host. The ova then develop into cercarial larvae to begin the process anew [2].

In the host, ova spread hematogenously to the intestines and liver, where a granulomatous reaction ensues, secondary to release of enzymes and toxins. This eventually leads to fibrosis and scarring of the presinusoidal radicals of the liver. Schistosomiasis is a common cause of noncirrhotic portal fibrosis, known as Symmers' or "pipestem fibrosis" [5,9]. The portal venous hypertension seen with schistosomiasis is secondary to the resulting obstruction of portal venous flow [2].

Acute schistosomal infection may manifest as either "swimmer's itch," a localized dermatitis, or Katayama fever, the flu-like syndrome that occurs one to two months after exposure. The first is a result of invasion of the skin; the latter a result of migrating larvae through the circulatory system. Although most infected adults remain clinically asymptomatic [1], chronic schistosomiasis can result in jaundice, abnormal LFTs, abdominal pain, and secondary portal hypertension with all of its attendant comorbidities [5,8]. Extra-intestinal schistosomal infection can affect the spinal cord, brain, and lungs [10,11].

Diagnosis of schistosoma infection is made in one of several ways, including detection of ova in the stool, serology, or histology. In this case, serology was used to confirm the presence of previously established schistosomiasis. This assay tests for the presence of antibody to microsomal fraction of adult *S. mansoni* worms, and has a specificity of 99% with a sensitivity of 96% [12]. The assay is not able, however, to distinguish between active or past infection, or to quantify the severity of the disease.

Radiologic diagnosis of schistosomiasis has been mostly focused on ultrasound imaging. Several case reports from 1984 demonstrated periportal fibrosis, which appeared as echogenic tubular shadows with central lucencies that radiated from the porta hepatis [5,13,14]. These were described as "bull's eye" lesions due to the appearance of the concentric ring of fibrosis surrounding portal venous vasculature. Our case showed similar findings. Grading of the intensity of periportal fibrosis on ultrasonography was later shown to correlate with disease burden [15,16]. In addition to the stigmata of portal venous hypertension, other ultrasonographic signs of schistosomiasis included hypertrophy of the left hepatic lobe, atrophy of the right hepatic lobe, gallbladder wall thickening, granulomas, and splenic nodules [5,17]. Acute schistosomiasis has been associated with hypodense hepatic nodules on both ultrasound and CT [18].

Reports of CT correlation of ultrasound findings in chronic *S. mansoni* infection have not been as extensive. In 1985, a published series of five such patients showed that periportal fibrosis on CT appeared as low-density periportal zones with strong contrast enhancement. Similar to the ultrasonographic findings, these enhancing periportal re-

gions could be seen as both rounded foci and linear branching patterns, depending on the cross-sectional orientation [8]. Our report demonstrates high-resolution MDCT imaging of concentric layers of periportal enhancement, previously seen on ultrasound, but not yet described with CT. Whereas periportal enhancement is nonspecific and can also be associated with Kaposi sarcoma or chemotherapy [19], this pattern of concentric ringed enhancement may prove to be a more specific indicator of schistosomiasis.

This case demonstrates a nodular and thickened gallbladder wall with evidence of stranding. Granulomatous reaction of the gallbladder in schistosomiasis is an unusual finding. As a gallbladder biopsy was not performed in this case, we do not have pathologic proof of correlation with schistosomiasis, although its extension to the porta hepatis supports this claim [13]. The differential diagnosis for a thickened gallbladder wall is broad, but would include gallbladder varices secondary to portal hypertension and chronic acalculous cholecystitis. However, given absence of portal vein thrombosis and identification of other varices on CT imaging, isolated gallbladder varices were thought to be unlikely [7]. Additionally, Doppler ultrasound appearance of the thickened gallbladder wall was not consistent with published reports of gallbladder varices [6]. The patient did not have signs or symptoms suggestive of acalculous cholecystitis.

Our case also demonstrates an irregular liver contour and an enlarged caudate lobe. Previous reports have noted ascites, portal collaterals, hypertrophy of the lateral segment, and irregular contour of the hepatic surface as associated indicators of disease [20,21]. Although most cases describing dystrophic calcification of schistosomal ova in a "turtleback" or "tortoiseshell" appearance are associated with S. japonica infection [21], there have been rare reports of calcification of the liver, spleen, small bowel [22], and large bowel [23] with *S. mansoni*. Magnetic Resonance Imaging of *S. mansoni* has shown similar findings with strong periportal enhancement after gadolinium administration [24].

In our era of increasing global travel and immigration, exotic diseases previously thought to be under the purview of infectious disease physicians are now becoming increasingly commonplace. Although the incidence of schistosomiasis in endemic areas has been decreasing, largely due to public health measures, increasing emigration and interest in rural tourism has led to an increase in the incidence of the disease in non-endemic areas. Reaquaintance with the radiographic findings of schistosomiasis, particularly with newer MDCT imaging, is therefore helpful in establishing the diagnosis in many asymptomatic patients.

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