

CME Article

Clinics in diagnostic imaging (I36)

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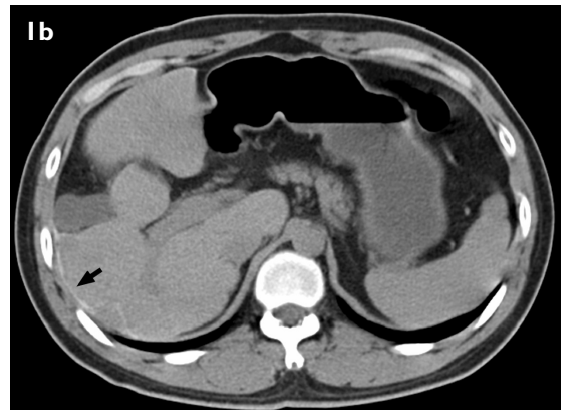
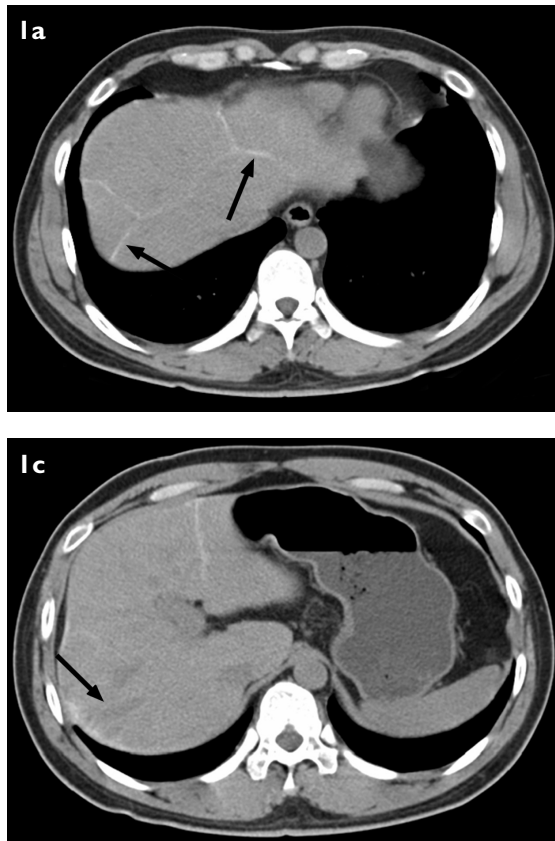


Fig. 1 Unenhanced CT images of the liver show (a) septal (arrows); (b) capsular calcifications (arrowhead); and (c) linear hypodensities at the periphery of the liver (arrow).

CASE PRESENTATION

A 37-year-old man from China was initially seen at the urology clinic for symptoms of urinary colic. Plain computed tomography (CT) imaging of the kidneys, ureters and bladder was requested for workup of renal

calculi. Besides demonstrating bilateral non-obstructing small renal calculi, the CT study revealed incidental findings in the liver (Figs. 1a-c). Triphasic hepatic CT imaging was then performed for further evaluation. What is the diagnosis?

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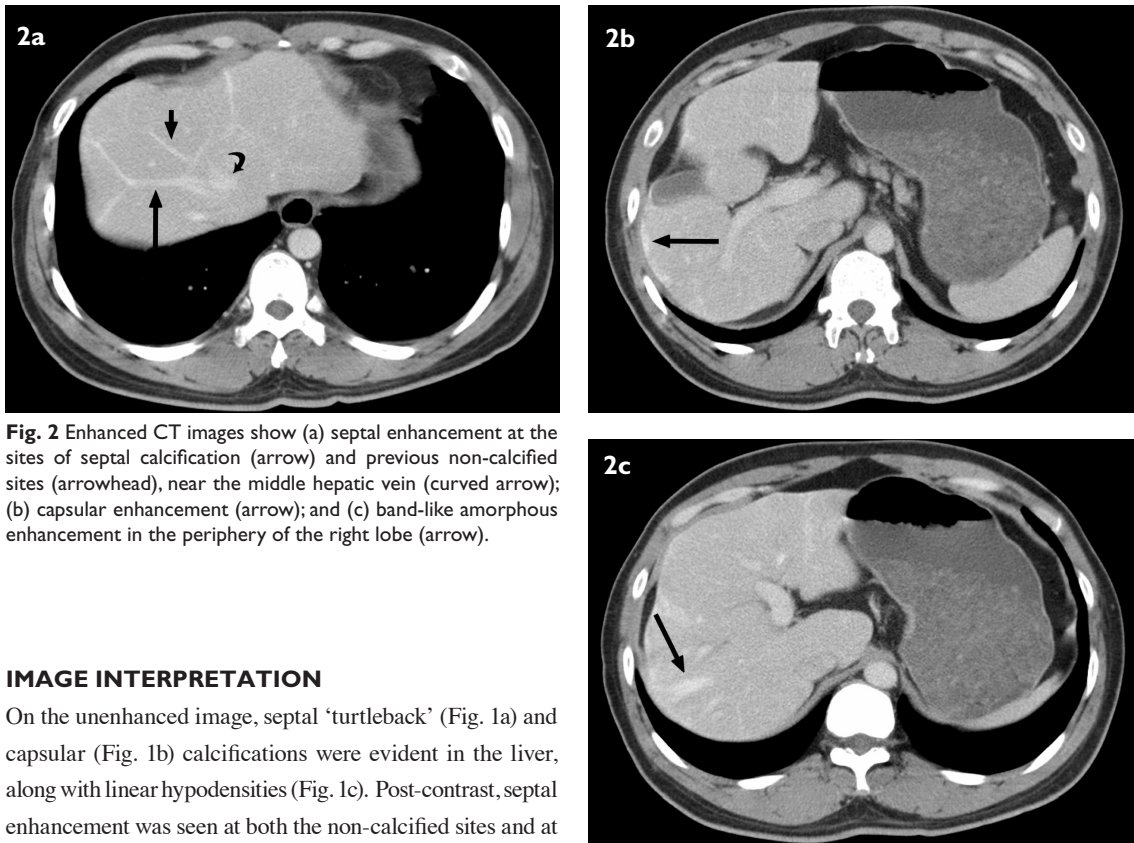


Fig. 2 Enhanced CT images show (a) septal enhancement at the sites of septal calcification (arrow) and previous non-calcified sites (arrowhead), near the middle hepatic vein (curved arrow); (b) capsular enhancement (arrow); and (c) band-like amorphous enhancement in the periphery of the right lobe (arrow).

IMAGE INTERPRETATION

On the unenhanced image, septal 'turtleback' (Fig. 1a) and capsular (Fig. 1b) calcifications were evident in the liver, along with linear hypodensities (Fig. 1c). Post-contrast, septal enhancement was seen at both the non-calcified sites and at the sites of septal and capsular calcifications (Figs. 2a & b). In addition, band-like amorphous enhancement was observed in the periphery of the right lobe (Fig. 2c) on the portovenous phase, showing further enhancement on the delayed phase, which corresponded to the area of linear hypodensity in the unenhanced image.

DIAGNOSIS

Chronic *Schistosoma japonica* infection of the liver.

CLINICAL COURSE

The patient subsequently revealed that he was previously diagnosed with a 'liver parasite' infection several years ago, after being investigated for persistent abdominal pain and diarrhoea, while residing in China. The renal calculi were managed conservatively, as they were small and non-obstructing.

DISCUSSION

Schistosomiasis is a parasitic disease caused by several species of fluke of the genus *Schistosoma*. The three main species infecting humans are *Schistosoma haematobium* (typically infecting the urinary tract), *Schistosoma japonicum* (typically infecting the liver and bowel) and *Schistosoma mansoni* (typically infecting the liver and bowel). Of these, *Schistosoma japonicum* is the most common in Eastern Asia, particularly China, Indonesia and the Philippines.⁽¹⁾ Its intermediate host is a tropical

freshwater snail, *Oncomelania hupensis quadrasi*.⁽²⁾ The eggs are excreted in the urine and faeces of affected animals. Upon maturity, the eggs release miracidia, which in turn infect the freshwater snails. The miracidia then mature into multiple cercariae within the snail, which are excreted into the water. The cercariae then infect new hosts by penetrating unbroken skin or gastrointestinal mucosa.

The clinical presentation is typically mild. Acute symptoms include a nonspecific rash at the site of entry, followed by constitutional symptoms such as fever, myalgia, lymphadenopathy and diarrhoea. Diagnosis of acute infection is typically made by microscopic identification of eggs in either urine or stool; if stool or urine examinations are negative, rectal biopsy may demonstrate eggs.⁽³⁾ Subsequently, the disease progresses to a chronic stage, with the liver being the most commonly affected organ. The adult worms reside in the mesenteric veins. Eggs laid in the mesenteric veins are transported to the liver via the portal veins and lodged in terminal branches of the intrahepatic portal veins, causing ischaemia and inflammation, followed by fibrosis.⁽⁴⁾ The fibrous tissue can form within the liver parenchyma or along the hepatic capsule in broad fibrous septa. On plain CT images, the fibrous septa can be visualised as hypodense areas.⁽⁵⁾ These areas of fibrosis can subsequently undergo dystrophic calcification.

The typical pattern of calcification consists of linear septal and curvilinear capsular calcification, giving rise to a polygonal 'turtleback' appearance.⁽⁶⁾ Septal calcification is seen more frequently in the right lobe and the peripheral aspect of the liver.⁽⁷⁾ Occasionally, a notch of the hepatic contour (junctional notch) is present at the junction of the capsule and septal calcification, giving the liver a pseudolobar appearance. Calcification within the portal tributary walls is rare, with only one case reported in the study by Araki et al.⁽⁷⁾

Three distinct types of enhancement have been described – septal, amorphous and capsular.⁽⁷⁾ Enhancement is typically progressive on multiphase CT, a feature of fibrotic tissue. Septal enhancement is linear and extends from deep within the liver parenchyma to the surface. Amorphous enhancement appears as an area of non-uniform enhancement. This is thought to be due to enhancement of broad fibrous septa, which lie parallel to the imaging plane. Capsular enhancement occurs along the liver surface. In areas of heavy calcification, enhancement may be difficult to visualise.

Complications of *Schistosoma japonicum* infection include hepatic fibrosis and portal hypertension, potentially resulting in life-threatening gastrointestinal haemorrhage.⁽⁸⁾ There is also an increased incidence of hepatocellular carcinoma and colorectal cancer in patients with schistosomiasis japonica.^(9,10) Migration of ova to other organs can occur; for example, ectopic *Schistosoma japonicum* eggs in the brain and pulmonary vessels may result in cerebral granulomatous disease and pulmonary hypertension, respectively.⁽³⁾

While no definitive microbiological evidence of *Schistosoma japonicum* was obtained in this patient, all the classical CT findings of hepatic schistosomiasis japonica infection were present. The septal and capsular calcifications with the characteristic 'turtleback' pattern, as well as septal, capsular and amorphous enhancement post contrast administration, are pathognomonic of the disease entity. By presenting this case, we would like to highlight the fact that while schistosomiasis japonica is extremely rare in our local population, both clinicians and radiologists should nonetheless be aware of this disease, particularly in a population that is becoming more well-travelled, with increasing numbers of immigrants from endemic countries.

ABSTRACT

A 37-year-old man from China was initially seen at the urology clinic for symptoms of urinary colic. Plain computed tomography (CT) imaging of the kidneys, ureters and bladder was performed, which (in addition to demonstrating renal calculi) revealed incidental findings of 'turtleback' septal and capsular calcifications, features pathognomonic for schistosomiasis japonica. Other classical features were demonstrated on the triphasic hepatic CT imaging that was subsequently performed. The clinical course, radiological features and complications of schistosomiasis japonica are discussed.

Keywords: liver, parasite, *Schistosoma japonicum*, schistosomiasis, turtleback

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SINGAPORE MEDICAL COUNCIL CATEGORY 3B CME PROGRAMME

Multiple Choice Questions (Code SMJ 201109B)

	True	False
Question 1. With regard to <i>Schistosoma japonicum</i> :		
(a) The three main species of flukes of the genus <i>Schistosoma</i> are <i>S. haematobium</i> , <i>S. japonicum</i> and <i>S. mansoni</i> .	<input type="checkbox"/>	<input type="checkbox"/>
(b) <i>Schistosoma japonicum</i> is the most commonly found species of fluke (of the genus <i>Schistosoma</i>) in Eastern Asia.	<input type="checkbox"/>	<input type="checkbox"/>
(c) The intermediate host of <i>Schistosoma japonicum</i> is a freshwater snake.	<input type="checkbox"/>	<input type="checkbox"/>
(d) Cercariae excreted by the intermediate host infect humans by penetrating the skin or gastrointestinal mucosa.	<input type="checkbox"/>	<input type="checkbox"/>
Question 2. Concerning acute infection by <i>Schistosoma japonicum</i> :		
(a) Initial clinical presentation is typically nonspecific.	<input type="checkbox"/>	<input type="checkbox"/>
(b) Diagnosis of acute infection is typically made by microscopic identification of eggs in blood.	<input type="checkbox"/>	<input type="checkbox"/>
(c) The liver is the most commonly infected organ in humans.	<input type="checkbox"/>	<input type="checkbox"/>
(d) Adult flukes reside in the mesenteric arteries.	<input type="checkbox"/>	<input type="checkbox"/>
Question 3. Concerning hepatic calcification in chronic <i>Schistosoma japonica</i> infection:		
(a) The typical pattern of hepatic calcification is a polygonal 'turtleback' appearance.	<input type="checkbox"/>	<input type="checkbox"/>
(b) Septal calcification is seen more frequently in the left hepatic lobe.	<input type="checkbox"/>	<input type="checkbox"/>
(c) Capsular calcification is a typical finding.	<input type="checkbox"/>	<input type="checkbox"/>
(d) Calcification within the portal vein walls is common.	<input type="checkbox"/>	<input type="checkbox"/>
Question 4. Concerning the pattern of enhancement in chronic <i>Schistosoma japonica</i> infection of the liver:		
(a) Three distinct types of enhancement have been described.	<input type="checkbox"/>	<input type="checkbox"/>
(b) The pattern of enhancement follows that of fibrous tissue.	<input type="checkbox"/>	<input type="checkbox"/>
(c) Amorphous enhancement appears as an area of uniform enhancement.	<input type="checkbox"/>	<input type="checkbox"/>
(d) Capsular enhancement may be difficult to visualise when calcification is heavy.	<input type="checkbox"/>	<input type="checkbox"/>
Question 5. Regarding complications of chronic <i>Schistosoma japonica</i> infection:		
(a) Hepatic fibrosis and portal hypertension are rare complications.	<input type="checkbox"/>	<input type="checkbox"/>
(b) There is also an increased incidence of hepatocellular carcinoma and colorectal cancer.	<input type="checkbox"/>	<input type="checkbox"/>
(c) Pulmonary hypertension is a known complication.	<input type="checkbox"/>	<input type="checkbox"/>
(d) There have been no reported cases of central nervous system involvement.	<input type="checkbox"/>	<input type="checkbox"/>

Doctor's particulars:

Name in full: _____

MCR number: _____ Specialty: _____

Email address: _____

SUBMISSION INSTRUCTIONS:

(1) Log on at the SMJ website: <http://www.sma.org.sg/cme/smj> and select the appropriate set of questions. (2) Select your answers and provide your name, email address and MCR number. Click on "Submit answers" to submit.

RESULTS:

(1) Answers will be published in the SMJ November 2011 issue. (2) The MCR numbers of successful candidates will be posted online at www.sma.org.sg/cme/smj by 24 October 2011. (3) All online submissions will receive an automatic email acknowledgment. (4) Passing mark is 60%. No mark will be deducted for incorrect answers. (5) The SMJ editorial office will submit the list of successful candidates to the Singapore Medical Council.

Deadline for submission: (September 2011 SMJ 3B CME programme): 12 noon, 17 October 2011.