

“Right Iliac Fossa Pain”: More Than Meets the Eye

A 29-year-old female Filipino domestic worker presented to the Emergency Department of Changi General Hospital with a 1-day history of right iliac fossa pain and vomiting. The patient was febrile with a body temperature of 38.3°C and she had a raised total white blood cell count of $17.0 \times 10^3/\mu\text{L}$ (4.0-10.0).

What do the computed tomography (CT) scan images (Figs. 1A and 1B) and corresponding post-appendicectomy histological photomicrographs (Figs. 2A and 2B) show? What is the diagnosis?

- A. Eosinophilic appendicitis
- B. Mucinous adenoma of the appendix
- C. Schistosomal appendicitis
- D. Mucinous adenocarcinoma of the appendix
- E. Leiomyoma of the appendix

Findings and Diagnosis

Axial and coronal CT scan images (Figs. 1A and 1B) demonstrate mild thickening of the appendix associated with mural calcification (indicated by the white arrows). Minimal periappendiceal fat stranding was present, suggesting the presence of an early acute appendicitis. There was however, no pneumoperitoneum or localised periappendiceal collection to suggest the presence of gross appendiceal perforation at the time of imaging. The patient also had mural calcification of the rectosigmoid junction (Fig. 3, indicated by the white arrow). Laparoscopic appendicectomy was performed, which revealed an acute appendicitis but there was no evidence of appendiceal perforation. Histopathological evaluation of the resected appendix showed transmural inflammation of the appendix with ulceration of the mucosa (Fig. 2A). There were also numerous ovoid bodies present within the mucosa, muscularis propria and the serosal region, many of which

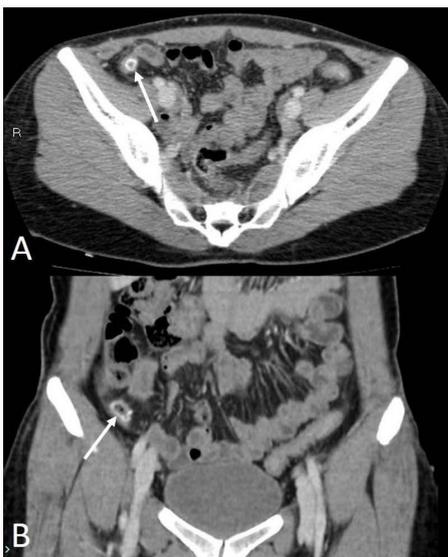


Fig. 1. A) Axial and B) coronal CT images of the pelvis which were obtained at the time of clinical presentation.

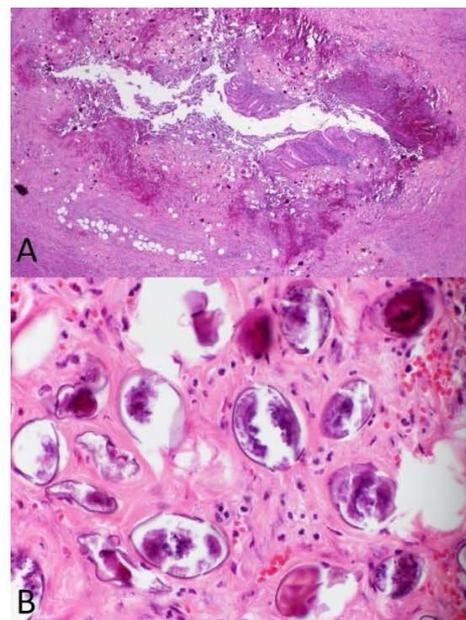


Fig. 2. Histological photomicrographs of the appendicectomy specimen at A) low (x 20) and B) high powered (x 400) magnifications (H&E stain).

Answer: C



Fig. 3. Axial CT image of the pelvis demonstrates mural calcification of the rectosigmoid junction (white arrow).

were calcified and contained coarse purplish granules within the cytoplasm (Figs. 2A and 2B). The diagnosis was schistosomal appendicitis.

Discussion

Schistosomiasis is a tropical disease caused by blood-dwelling fluke worms of the genus *Schistosoma*, with approximately 200 million people being affected worldwide.¹ The transmission cycle requires contamination of surface water by excreta, specific freshwater snails as intermediate hosts, and human water contact. The main schistosomes infecting human beings are: *S. mansoni*, which causes intestinal and hepatic schistosomiasis in Africa, the Arabian peninsula and South America; *S. haematobium* which causes urinary schistosomiasis in Africa and the Arabian peninsula; and *S. japonicum* which causes intestinal and hepatosplenic schistosomiasis in China, the Philippines, Thailand and Indonesia.¹ *S. japonicum* is a zoonotic parasite that infects a wide range of animals including cattle, dogs, pigs and rodents. *S. mansoni* is also found in rodents and primates, but human beings are the main host.

Adult *S. japonicum* worms often reside and produce ova in the mesenteric veins of their human hosts, in particular the inferior mesenteric vein. While some ova penetrate the intestinal wall and are excreted along with faecal matter, the remaining ova either become implanted within the intestinal wall (which includes the appendix), or migrate upstream to the liver via the portal vein.² Schistosomiasis as a cause of acute appendicitis is rarely seen in developed countries, and if so encountered are commonly found in those who have travelled or emigrated from endemic areas.³⁻⁷ Terada and his colleagues from Japan reported an incidence of only 0.32% in their series of 311 appendicectomies.⁷ There are 2 mechanisms involved in the pathogenesis of schistosomal appendicitis. The first is an immunological granulomatous reaction to newly deposited ova causing tissue necrosis

and eosinophilia which may occur in the early phase of the infection. The second is related to the obstruction of the appendiceal lumen in the late stage of the infection (after several months to years) by chronic inflammation and fibrosis surrounding the dead ova, thereby increasing the risk of superadded infection.⁸

Lee et al correlated the imaging finding of intestinal mural calcification seen on CT in cases of *S. japonicum* infection, with the calcification of ova deposited within the intestinal wall including that of the appendix.⁹ CT scans are now frequently performed to evaluate patients who present with an acute abdomen in order to exclude a surgical cause such as an acute appendicitis.¹⁰ Since there is no pathognomonic clinical or operative finding, careful evaluation for the presence of intestinal or appendiceal mural calcification on CT will therefore alert the clinician to the possibility of underlying schistosomal infection, and the potential need for treatment with antihelminthic drugs, as seen in the case reported by Tang and his colleagues.¹¹

Colonic calcification seen with schistosomiasis infection should be differentiated from other causes such as phlebosclerotic colitis, renal failure, hyperphosphataemia (including treatment with lanthanum), mucinous adenocarcinomas, and leiomyomatous tumours of the colon and rectum. Phlebosclerotic colitis usually involves the right-sided colon as compared with distal colonic involvement in schistosomiasis. The characteristic CT imaging features of this rare form of colitis includes calcification of the veins of the colonic wall and adjacent mesentery, collateral formation, thickening and oedema of the colonic wall, and increased density of the surrounding mesenteric fat.¹² Varying degrees of colonic calcification have also been reported in patients with renal failure and hyperphosphataemia, as well as in patients who were treated with lanthanum for the latter condition.¹³⁻¹⁵ Distinguishing patients with these conditions from those with suspected intestinal schistosomal infection can however, be readily made based on clinical grounds. Mucinous adenocarcinomas and leiomyomatous tumours of the colon and rectum may also demonstrate intratumoural calcification. These tumours can be differentiated from cases of intestinal schistosomal infection by the presence of colonic wall thickening (often eccentric), associated with bulky, heterogeneously enhancing mural masses which may show areas of hypoattenuation or cystic degeneration.^{16,17}

Conclusion

Although rarely encountered in developed countries, schistosomiasis should be suspected as a cause of an acute appendicitis in patients who have emigrated or are travelling from endemic areas, and in the presence of appendiceal mural calcification on CT.

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