OPEN ACCESS – CASE REPORT

Ectopic Schistosomiasis Presenting as Ruptured Appendicitis with Periappendiceal Abscess Formation: An Alternative Pathogenetic Perspective

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ABSTRACT

Schistosomiasis is still a public health burden in the Philippines. Chronic infection with *Schistosoma japonicum*, the only species endemic in the Philippines, clinically manifests itself in a wide variety of pathologies usually correlated with the anatomical site of adult worm activity and deposition of eggs. One of the documented ectopic sites for Schistosoma ova is the appendix. A rare sequela of this is acute appendicitis and an even rarer consequence is progression to appendiceal rupture leading to acute peritonitis. We present a case of a 27-year-old Filipino residing in Davao City but born in Agusan Province who initially complained of right lower quadrant abdominal pain but presented at the emergency room with generalized abdominal tenderness with signs of peritoneal irritation. Exploratory laparotomy with an infraumbilical incision revealed ruptured appendicitis with periappendiceal abscess formation and appendectomy was subsequently done. Schistosoma infection of the appendix was subsequently established by histopathological analysis. Furthermore, features observed suggest an atypical pathogenetic process contrary to the putative pathogenesis of most cases of acute appendicitis.

Key words: schistosomiasis, Schistosoma japonicum, acute appendicitis, periappendiceal abscess

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INTRODUCTION

Schistosomiasis is still a public health problem, especially in endemic areas in the Philippines. The risk for *S. japonicum* infection spans approximately 12 million people living in 28 provinces located across 12 different geographical regions.[1] These regions have no distinct dry season and comprise predominantly rice-growing areas. Transmission continues to be high because climatic conditions and current rice farming methods maximize contact between freshwater snails and humans. Furthermore, a 2011 study done in Northern Samar showed that bovine and water buffaloes play a major role in the transmission of schistosomiasis with an infection prevalence of approximately 90% making control efforts even harder.[2]

Without intervention, *Schistosoma* typically survives in the host body for up to 5 years but there have been reports of chronic infections for up to 40 years.[3] Long-term infection or repeated reinfection with *S. japonicum* causes two types of morbidities: those with subtle clinical manifestations and those with end-organ complications. Subtle clinical manifestations are caused by elevated inflammatory cytokines induced by ova or the parasite itself. Chronic anemia, growth retardation, impaired cognitive abilities, and malnutrition have been documented in children with *S. japonicum* infection.[4,5] The end-organ complications are sequelae of granuloma formation with subsequent tissue injury and fibrosis. Several ectopic localizations for various species of Schistosoma have been documented before.[6,7] The first-ever known case of appendiceal schistosomiasis was reported by Turner in 1909 and has been described in many reports, especially in endemic areas but to our knowledge, there is only one other reported case that progressed to ruptured appendicitis and subsequent

periappendiceal abscess formation.[8,9] We present an unusual case of appendiceal Schistosomiasis wherein the patient had a ruptured appendix with localized periappendiceal abscess formation.

CASE

A 27-year-old Filipino female residing in Davao City (4 years) but originally from Agusan Province of the Philippines, came into our institution with a 2-day history of sudden onset of epigastric pain which later migrated to the right lower quadrant of the abdomen. There was no fever, anorexia, vomiting, or dysuria. She had no comorbidities or previous surgeries. The patient decided to undergo ultrasonography of the whole abdomen as an outpatient but yielded unremarkable results. An increase in the severity of the pain prompted consult in our institution.

At the emergency room, a physical examination of the abdomen revealed tenderness on all quadrants upon light palpation, with note of muscular guarding. Vital signs remained normal and the patient was afebrile. Other physical examination findings were unremarkable.

A pregnancy test done was negative. Complete blood count revealed only a slight increase in white blood cell count of 14.58 x 10^9/L with normal hemoglobin levels and no eosinophilia. There was no pyuria in the urinalysis and coagulation studies and serum creatinine was normal. However, blood chemistry revealed hypokalemia at 3.2 mmol/L.

The preliminary diagnosis of this case was acute abdomen probably from ruptured appendicitis. The patient was immediately started on intravenous Ampicillin-Sulbactam, intravenous correction of potassium, and prepared for surgical intervention. The team decided on exploratory laparotomy with an infraumbilical midline incision. Intraoperatively, the appendix measured 7 cm x 2 cm and was gangrenous with a perforation near the base and the antimesenteric area. Packets of pus were noted around the appendix, both at the left and right paracolic gutters and at the pelvic gutter. Excision of the appendix with lysis and suctioning of all packets of pus was done. Adequate peritoneal lavage was ensured. An appendix was sent to surgical pathology for microscopic examination.

Histopathological analysis of the appendix revealed dense neutrophilic infiltration in the mucosa, submucosa, muscularis propria, serosa, and mesoappendix (Figure 1). There is pus in the appendiceal lumen with large areas of necrosis. Infiltration of Schistosoma ova with granuloma formation in the submucosa and muscularis propria is noted (Figure 2). Praziquantel was started at 40 mg/k/day at 2 divided doses and sent home improved.

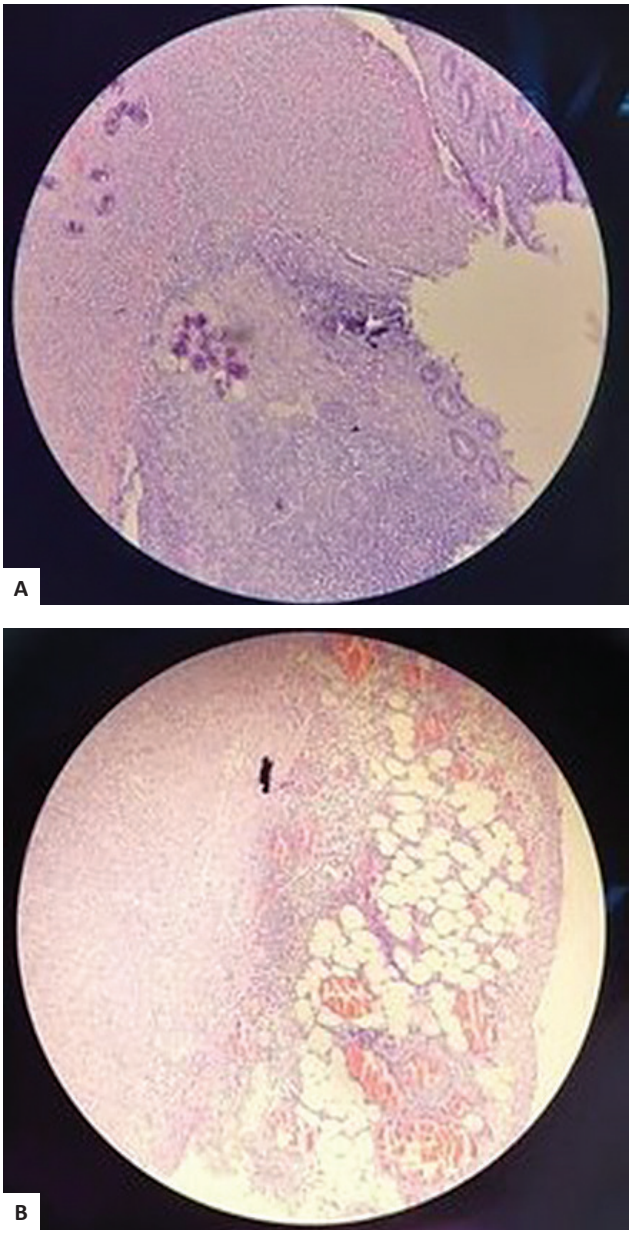

Figure 1. (A) Dense neutrophilic infiltration in the mucosa, submucosa, muscularis propria, serosa and **(B)** mesoappendix (H&E, 4x).

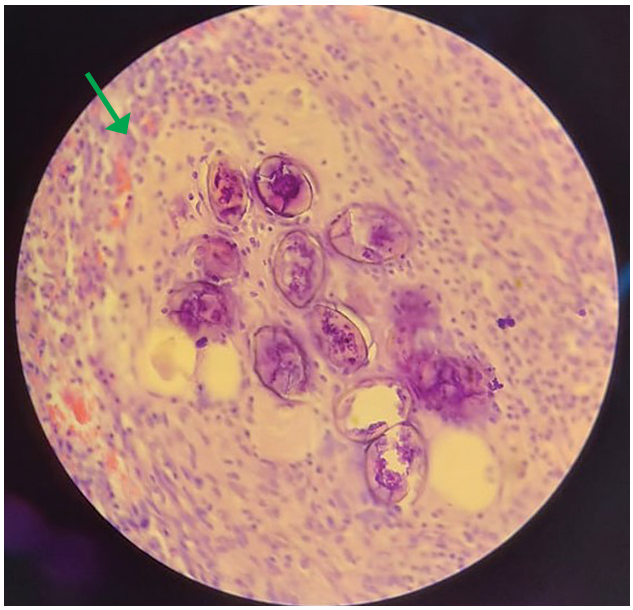

Figure 2. Schistosoma ova with granuloma formation *(green arrow)* found in submucosa layer (H&E, 40x).

DISCUSSION

In chronic schistosomiasis, clinical manifestations depend on the organ involved. These are classified into hepato-intestinal, hepatosplenic, pulmonary, cerebral, and ectopic forms.[5] Although rare, ectopic schistosomiasis has been reported in the heart, ovaries, uterus, fallopian tubes, ureters, urinary bladder, and the appendix.[6]

The preferred anatomic location for residence and egg deposition varies by species. *S. japonicum* in particular often resides in the inferior and superior mesenteric veins. After the eggs are deposited into the vascular lumen, digestive compounds are utilized by the schistosome so that around half of the ova can penetrate the blood vessel wall and enter the bowel including the appendix. The rest of the ova proceeds to the liver where they are filtered from the circulation.[10]

In our patient, *S. japonicum* ova were identified in the appendix making it the most obvious cause of appendicitis. However, the role of the Schistosoma ova in the pathogenesis of acute appendicitis and the subsequent rupture of the said organ in our patient remains unclear. Obstruction followed by infection from fecal contaminants is thought to be an important mechanism in the pathogenesis of acute appendicitis. However, using histopathological criteria, Satti et al., reported that there are 2 distinct histopathologic features of acute Schistosomal appendicitis: obstructive and granulomatous. Obstructive schistosomal appendicitis results from obstruction of the appendiceal lumen seen in chronic infection due to fibrosis around calcified eggs which increases the risk of other infections. On the other hand, granulomatous schistosomal appendicitis is caused by active granulomatous inflammation with eosinophilic necrosis around the ova. This was used as a marker for active schistosomal infection and is more congruent in the histopathological examination of the appendix in our patient.[11] In this case, there is the presence of histopathologic features that suggest that appendicular obstruction played no role in the pathogenesis of acute appendicitis and subsequent rupture. We hypothesize that the cause of appendicitis and subsequent rupture of the appendiceal wall, in this case, was caused by granulomatous tissue destruction. We contrast this to the only other report of ruptured appendiceal schistosomiasis with abscess formation by Al-Waheeb in 2008 wherein histopathological analysis of the sections from the appendix and omentum yielded no granulomatous response. Both cases led to rupture of the appendix and subsequent abscess formation but through different mechanisms.[9]

The gold standard for the diagnosis of schistosomiasis is the detection of parasite ova in fecal specimens. We performed direct microscopy on three separate stool samples to look for evidence of adult worm activity, but all yielded negative for parasites or ova. This is the reason why we had to rely on histopathologic analysis to clinch the diagnosis of schistosomiasis in this case. A possible explanation for this finding is that ova seen in the appendix of the patient may be old and adult worms that have already died out. However, this is contradicted by our histopathologic findings that suggest an ongoing infection. Nonetheless, we committed to an ongoing infection and decided to administer Praziquantel. Praziquantel is the recommended treatment for all species and all forms of schistosomiasis at 40 mg/kg, which is highly effective in approximately 91.7% of treated individuals.[12]

CONCLUSION

Putative pathogenesis of most cases of acute appendicitis involves luminal obstruction commonly by fecalith, lymphoid hyperplasia, fecal debris, true calculus, or tumor. In our case, we lack evidence of obstruction which suggests a possible existence of a non-obstructive form of appendicitis predominated by a granulomatous process.

Furthermore, in the absence of pathognomonic clinical preoperative and intraoperative findings, histopathologic diagnosis of patients with appendicitis is required for proper intervention.

Recent epidemiological and zoological studies seem to show that the national prevalence of schistosomiasis in the Philippines may have been initially underestimated leading to the relaxation of control measures. Schistosomiasis remains a public health burden and doctors practicing in endemic areas should be aware of the possibility of seeing atypical presentations of this parasitic disease.

ETHICAL CONSIDERATION

Patient consent was obtained before submission of the manuscript.

STATEMENT OF AUTHORSHIP

All authors certified fulfillment of ICMJE authorship criteria.

AUTHOR DISCLOSURE

All authors declared no conflict of interest.

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