Acute Appendicitis Caused by an Echinococcal Brood Capsule Unmasks an Asymptomatic Hepatic Hydatid Cyst

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ABSTRACT
Hydatidosis is an endemic helminthic disease in the cattle-grazing regions of Asia. It is usually caused by the cestode *Echinococcus granulosus*. Internal organs, particularly the liver and lungs, are predominantly affected, but the appendix is only rarely involved with the formation of characteristic hydatid cyst. We present a unique case of appendiceal hydatidosis in an 18-year-old woman with acute appendicitis. Her preoperative abdominal ultrasound revealed an asymptomatic hydatid cyst in the liver. An echinococcal brood capsule was detected postoperatively within the appendicular lumen under microscope.

INTRODUCTION
Hydatid disease is caused by the larval form of cestode *Echinococcus granulosus*, *E. multilocularis*, and *E. oligarthrus*, with the first being the most common species. Hydatid disease mainly involves the liver (70%) and lungs (15%), followed by the spleen (4%), kidneys (3%), central nervous system (2%), and heart (0.02–2%).1,2 Relatively unusual sites include the head and neck region, breasts, mediastinum, peritoneum, thyroid, and appendix. Appendiceal hydatidosis has been reported only a few times in the literature.

CASE REPORT
An 18-year-old woman presented with excruciating right periumbilical colicky pain, preceded by an episode of vomiting. On physical examination, muscle guarding with rebound tenderness was elicited at right iliac fossa. Preoperative complete blood count yielded neutrophilia and mild eosinophilia. With a high clinical suspicion of acute appendicitis, an abdominal ultrasonography (US) revealed an aperistaltic dilated appendix without fecolith and a coexistent 7.2/C2 6.7 cm hydatid cyst in the liver (Figure 1). She subsequently underwent an appendectomy, while excision of the hepatic hydatid cyst was scheduled for 6 weeks after the appendectomy.

Grossly, the appendix appeared inflamed. On a cut section, the dilated lumen contained only mucoid debris. Histopathologically, the appendix was found to be infiltrated transmurally up to its serosa with many inflammatory cells, including eosinophils, and the mucosa was focally ulcerated. Its lumen contained scant necroproteinaceous exudate. Within that debris, a distorted but yet viable brood capsule of the *Echinococcus* species was recognized. It was a vesicle-like structure, filled with a thin proteinaceous fluid and sheathed by a thick acellular cuticular membrane. The capsule contained a single central protoscolex, with three distinct parts: the broad globular scolex, the soma or body, and a constricted part in between known as the neck. This protoscolex was still immature as there was no evidence of hooklets or suckers that would produce a hydatid cyst (Figure 2). On the basis of these findings, the patient was diagnosed with acute appendicitis due to impaction of the echinococcal brood capsule.

In similar conditions, some patients may present with hypersensitivity-like symptoms in response to immunogenic hydatid fluid leakage, but our patient did not report such symptoms.13 Her liver function test was within normal
limits. She was promptly instituted upon oral albendazole (800 mg daily for 3 months). Six weeks after appendectomy, the hepatic cyst was surgically removed. The characteristic laminated membrane of the hydatid cyst was identified under microscope (Figure 3). The patient was free of relapse at a 7-month follow-up visit.

DISCUSSION

Echinococcosis or hydatid disease is a form of zoonoses caused by the larvae from *Echinococcus* species, particularly *E. granulosus*. Its usual life-cycle circulates between carnivorous definitive hosts such as dogs or wolves and intermediate hosts such as sheep, goats, and cattle. The adult worm lays eggs within the gastrointestinal tract of definitive hosts, which pass into their feces. Humans are the incidental intermediate hosts, infected through the ingestion of contaminated food.4

The eggs hatch in the human intestine and disseminate via hematological routes to the liver, lungs, and other organs to develop into hydatid cysts. The appendix is rarely invaded with hydatidosis.1-3 In 1902, Lyon reviewed 241 cases of hydatid disease in North America, and the intestine was involved only once.5 A 2013 review reported that only one of 463 published cases of hydatid cyst involved the appendix.3

The appendix is generally involved in cases of extensive multiorgan-disseminated hydatidosis.6,7 De novo appendiceal invasion occurs infrequently.8-10 Regardless of its initial presentation, the appendiceal hydatidosis usually features the characteristic laminated cyst containing “hydatid sands,” i.e., the brood capsules. The cysts are localized within the appendicular wall, inside its lumen, mesoappendix, and sometimes even distorts the appendix enough to make it almost nonexistent.6,8-10 In our case, the appendix was secondarily involved after the asymptomatic rupture of a preexisting hepatic hydatid cyst. In contrast to earlier literature, the hydatid cyst was absent in the appendix. To our knowledge, this is the first
case of such a unique presentation of appendicular echinococcosis in the literature.

We hypothesize that the luminal presence of the brood capsule triggered an inflammatory response. Passage of hydatid fluid from an incipient rupture of the capsule could be the likely cause. A hepatic hydatid cyst usually ruptures into the biliary system and only rarely ruptures within the peritoneal cavity. The intrabiliary leakage can be a frank rupture, which occurs in 5–17% of cases and is readily recognizable under conventional radiology, or the leakage may be occult, as seen in 10–37% of cysts. Patients who suffer from a frank rupture almost always present with severe abdominal symptoms and allergic reactions. Whereas those with an occult leakage, like our patient, remain relatively asymptomatic with or without mildly elevated liver enzymes. Surgical excision followed by postoperative antimicrobial therapy using antihelmintics is the treatment of choice for hydatid disease. Our case was successfully managed in this way.

Appendiceal echinococcosis in absence of characteristic hydatid cyst formation is extremely rare but should be considered when diagnosing acute appendicitis in endemic regions. Surgical removal is the best treatment option. Basic education on personal hygiene and hand-washing is the most effective prevention.

DISCLOSURES

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