

Primary Hydatid Disease of the Spleen: A Case Report

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ABSTRACT

Primary hydatid disease of the spleen is rarely reported. We describe the first report of primary hydatid disease of the spleen which was diagnosed by ultrasonography-guided fine needle aspiration (FNA).

A 25-year-old female from Fars province, Iran, presented with left flank pain one week prior to admission. Abdominal ultrasonography showed a large cystic lesion, 8 cm in diameter at the splenic hilum. A provisional diagnosis of organized hematoma, epidermoid cyst, cystic neoplasm or cystic infection was made, and ultrasonography-guided FNA was performed. Cytologic examination showed scolices, protoscolices, scattered hooklets and microcalcified material. Echinococcosis was diagnosed. Splenectomy was then performed and histopathological examination confirmed the diagnosis. After two-year follow-up the patient is completely well. (*J Infect Dis Antimicrob Agents* 2004;21:93-7.)

INTRODUCTION

Echinococcosis (hydatid disease) primarily affects the liver, however secondary involvement due to hematogenous dissemination may be seen in almost any anatomical locations. Isolated hydatid disease of the spleen is very rare.¹⁻⁵ Hydatid disease is caused by the larval tapeworm of *Echinococcus granulosus*, *E.*

multilocularis, *E. vogeli* or *E. oligarthrus*. The disease is prevalent in South America and in the Mediterranean, parts of northeastern Africa, and parts of eastern Europe. Humans are the intermediate hosts of the larval form or hydatid cyst, whereas the definitive host usually is a dog that passes eggs in its feces. *E. granulosus*, which produces unilocular cysts,

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is prevalent in Australia, Africa, eastern Europe, the Mediterranean and the Middle east. *E. multilocularis*, a causative agent of alveolar echinococcosis, is widespread across arctic, sub-arctic and temperate zones of Asia from Turkey to Japan.⁶ It is also prevalent in Europe and North America. *E. oligarthrus* and *E. vogeli*, causative agents of polycystic echinococcosis, are restricted to South and Central America.⁶ When the hexacanth embryo is ingested, it fixes to and penetrates the intestinal wall, enters the venous capillaries in the portal system or lymphatic vessels and disseminates to the liver and lungs. Human hydatid disease can involve the liver (66%), lung (5-15%) and spleen (less than 2%).¹⁻⁵ Most hydatid cysts are diagnosed by a combination of history and clinical features. Ultrasonography and computed tomography are the best complementary explorative tests for diagnosis.^{1,2,7} Cystic splenic lesion can also be due to lymphangioma, epidermoid cyst, splenic abscess, pseudocystic and cystic neoplasms.^{1,3-5,7} Serologic tests including immunoelectrophoresis, immunofluorescence and enzyme-linked immunosorbent assays are also available.⁸⁻¹⁰ This is the first report of a primary hydatid disease of the spleen diagnosed by ultrasonography-guided fine needle aspiration (FNA).

CASE REPORT

A 25-year-old woman from a farming family in Fars province, Iran, presented with left flank pain one week prior to admission. Due to vague abdominal pain, the patient has previously been treated as peptic ulcer. Abdominal examination revealed no organomegaly. Laboratory tests showed a hemoglobin of 13 g/dl, white blood cells of $21 \times 10^3 / l$, blood urea nitrogen of 4 mg/dl, creatinine of 0.8 mg/dl and suger of 126 mg/dl. Abdominal ultrasonography showed a large cystic lesion, 7.9×7.4 cm in size at the splenic hilum (Figure 1). A clinical diagnosis of epidermoid cyst, hematoma, cystic

neoplasm or infectious cyst was included. Ultrasonography-guided FNA of the splenic lesion was performed with a 0.7 mm (22 gauge) needle, and yielding about 200 ml of clear fluid. Direct and centrifuged deposit smears were made and fixed in 95 percent alcohol, and some were air dried. Papanicolaou, hematoxylin-and-eosin and Wright-Giemsa stains were done. The cytologic study of concentrated sediment showed various morphologic stages of *Echinococcus* including intact and crushed protoscolices with rows of hooklets, scolices and isolated hooklets (Figure 2 A and B). The cyst wall was laminated and microcalcified. No serologic test was performed. Splenectomy was then performed, and special care was taken to avoid intraoperative rupture and spillage leading to dissemination of infectious scolices. During operation there was a large splenic unilocular cyst, 14×7 cm in size. It contained thin wall and clear fluid. The patient had an uneventful post-operative period. She received mebendazole (40 mg per kg per day) 4 days before operation, and continued it for 3 months. Histopathological examination showed laminated membrane with

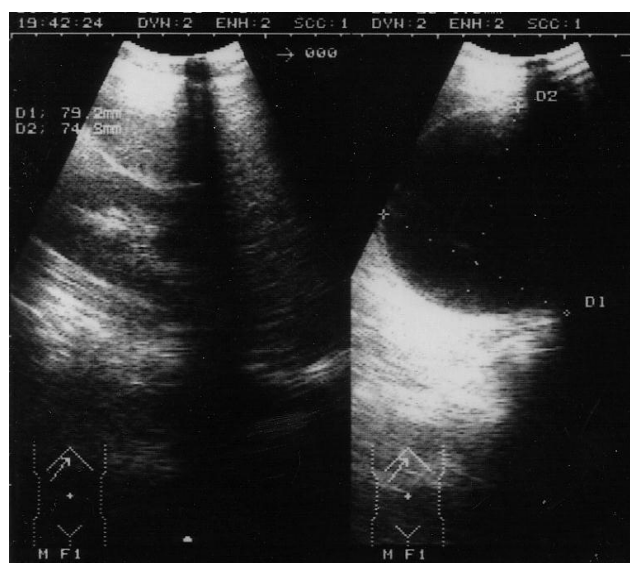


Figure 1. Abdominal ultrasonography showed a cystic lesion, 7 cm in diameter at the splenic hilum.

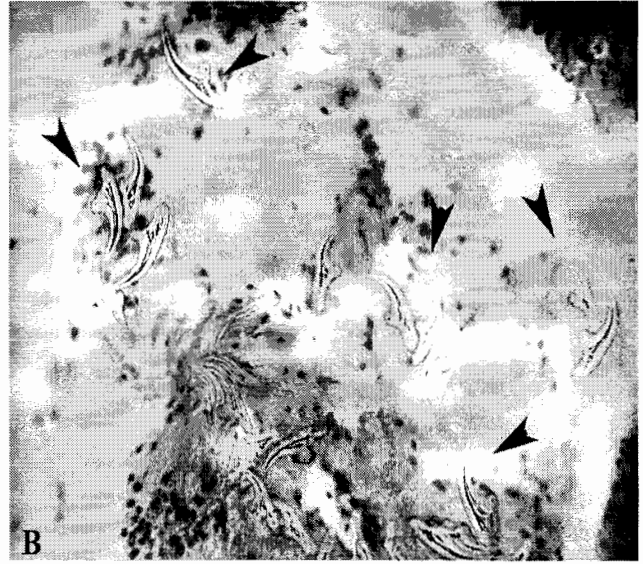
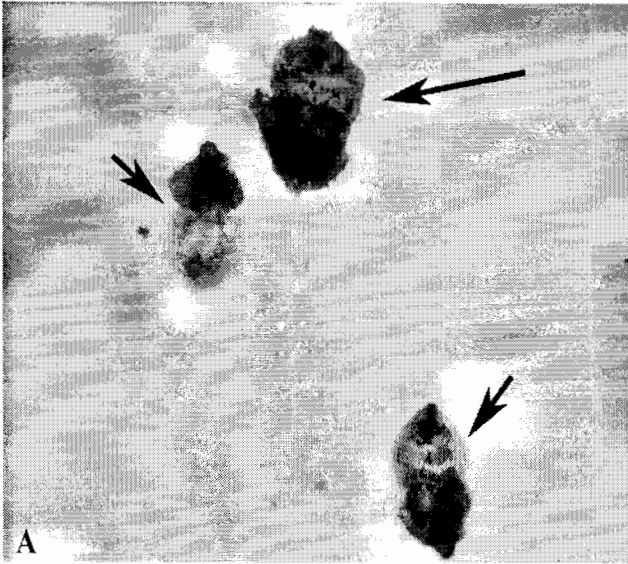


Figure 2A and 2B. Cytologic study showed protoscolices (long arrow), scolices (short arrow) and hooklets (arrowhead). (H&E, Papanicolaou, 400 x 400, respectively).

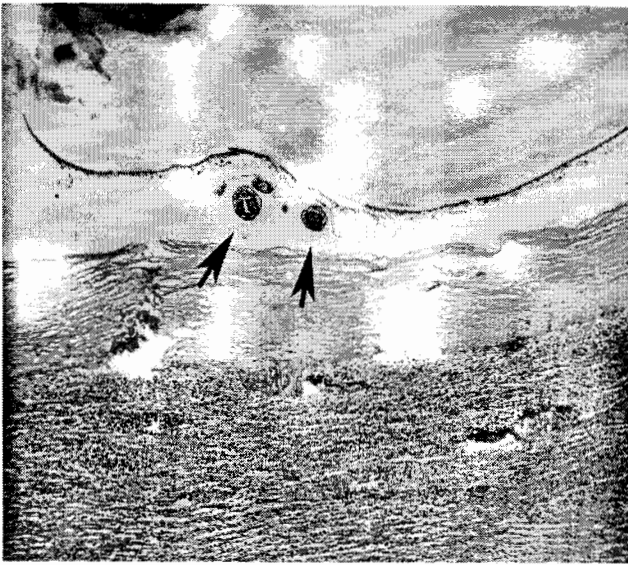


Figure 3. Histopathological examination showed laminated membrane with attached and free protoscolices (arrow). (H&E, x 360)

attached and free protoscolices and hooklets. (Figure 3) The patient was completely well after two years of splenectomy.

DISCUSSION

A human can be an intermediate host of *Echinococcus*. After ingesting the eggs, the embryo

penetrates the intestine and can involve almost any organ, but mostly the lung or liver.^{1,2,11} Primary splenic involvement is very rare.¹⁻⁵ Secondary hydatid disease of the spleen can be caused by the spontaneous or iatrogenic rupture of the hydatid disease of the liver, and spillage leading to dissemination of scolices to the spleen. The common presentations of hydatid disease of the spleen usually include pain and pressure effect from enlarging cyst. Imaging characteristics of hydatid disease are multivesicular cysts, calcification and intracystic membrane.⁷ Ultrasonography in our case did not show any of these characteristic findings. A new ultrasonographic classification for cystic echinococcosis (CE) is proposed. There are seven types including simple, multiple, detached endocystic, mixed-type, heterogeneous, hyperechoic and calcified CE. This classification appears more correlated with the natural history of CE and permits a better differential diagnosis and more suitable treatment.¹² Ultrasonography-guided FNA can be the most rapid and safe preoperative diagnostic test, although there remains a concern of the risk of anaphylactic reaction or dissemination from

accidental rupture and spillage of infectious scolices.^{9,13,14} In literature reviews, ultrasonography-guided FNA appears to be a safe diagnostic approach for evaluation of suspected hydatid disease, and adverse events are uncommon.^{3,8-10,13-17} A use of thin (22 gauge) needle for FNA will ensure a lower rate of such events.^{3,14} Cases with atypical clinical presentations with non-characteristic radiologic features; FNA might be the appropriate initial diagnostic test. The appearance of scolices, detached hooklets and fragments of multilayered laminated chitinous membrane are diagnostic cytological clues of hydatid disease.^{8,10,14,15} Until recently, surgery is the treatment of choice for most of hydatid disease. However, the current recommendation by the World Health Organization is percutaneous puncture under sonographic guidance, aspiration of cystic fluid, injection of a protoscolicidal agent and reaspiration of cyst content (PAIR).¹⁸ Recently some authors have used assisted laparoscopy to treat splenic hydatid disease.² Praziquantel and albendazole are drug treatments of choice.¹⁸ However, mebendazole was used in our case due to no availability of both mentioned drugs. Cytologic diagnosis using FNA has been described in the hepatic, nephrogenic and pancreatic hydatid disease.^{3,8,19,20} In our case, cytologic diagnosis permitted an immediate diagnosis and helped establish the best surgical approach with minimal manipulation.

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