tropical lung disease

An Unusual Presentation of Hydatid Disease of the Lungs*

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In man, hydatid disease is caused by the larval stage of a small tapeworm of dogs and other canines. Most human infections are from Echinococcus granulosus and are associated with the raising of sheep and cattle, the presence of domestic dogs, and the lack of proper health controls. The lungs are involved in 20 percent of the patients. The following is an uncommon presentation of pulmonary hydatid disease.

**CASE REPORT**

One of us (O.P.S.) was called to see a 39-year-old sheep herder from Mildura, Australia. The man came to Southern California to surf and stayed in a hotel for a week before the incident.

On this particular evening, the patient developed a paroxysm of violent cough and expectorated a cupful of blood-tinted watery fluid, containing thin membranous particles. He denied having any fever, chest pain, weight loss, or trauma. He did not smoke, drink excessively, or abuse drugs.

The findings from a physical examination were normal. Microscopic examination of the expectorated fluid revealed many scolecites with hooklets and suckers consistent with the diagnosis of hydatid disease (Fig 1). The patient admitted that four years ago, he had been diagnosed as having a hydatid cyst in his right lung; however, he never had any symptoms.

A chest roentgenogram obtained about two hours after this rather unusual clinical presentation was essentially normal, except for a few linear scars in the right upper lung field (Fig 2).

The patient was instructed to take mebendazole (100 mg twice per day) for one month. He returned to Australia and has remained asymptomatic.

In an endemic area, pulmonary hydatid cysts often remain asymptomatic. The solid looking shadows often calcify. Occasionally, a cyst may rupture, causing anaphylactic reaction and sudden death. The patient described herein was extremely lucky, as he expectorated the entire cyst without any complication. Mebendazole was prescribed to kill any of the remaining cysts.

**FIGURE 1.** Scolex (head) of *E. granulosus* showing rostellum (A), multiple hooklets arranged in two rows (B), and suckers (C) (H and E, original magnification × 150).

**FIGURE 2.** Normal chest roentgenogram two hours after patient had coughed up hydatid cyst. Arrow indicates only linear stranding and increased markings.
**DISCUSSION**

Human hydatid disease is caused by larval tapeworms of the genus, Echinococcus. *Echinococcus granulosus* and *E. multilocularis* are the tapeworms that lead to hydatid disease in man. *Echinococcus granulosus*, accounting for greater than 90 percent of the human cases, lives as an adult worm in the canine intestine. The adult worm sheds eggs that eventually are ingested by the intermediate host, usually sheep, but also pigs and cattle, among others. The eggs develop into a larval stage in the intermediate host, which humans can accidently become, and then penetrate the intestine and most commonly come to reside in the liver. The lung is the second most common site, but also the brain, peritoneal or pelvic cavities, bone, and kidney can be involved. The cycle is completed when the dog ingests remains of slaughtered sheep. *Echinococcus granulosus* is associated with the raising of sheep and cattle in climatic conditions varying from tropical to subarctic. A high incidence of the disease is observed in East Africa, the Mediterranean littoral, South America, the Middle East, Australia, India, and the Soviet Union. *Echinococcus multilocularis* is distributed in Canada, central Europe, Siberia, northern Japan, Alaska, and the north central United States. *Echinococcus multilocularis* has a life cycle similar to *E. granulosus*, but the definitive host is usually the wolf and the intermediate host the mouse. Disease caused by *E. multilocularis*, called alveolar hydatid disease, is distinctly different. More commonly the liver is involved, and the disease is more advanced at presentation.

Clinical features of the disease depend on the organ involved and the size of the hydatid cyst. About 70 percent of the cysts develop in the liver, 20 to 30 percent in the lungs, and the rest in other sites. The structure of the cyst consists of an outer pericyst layer that is composed of the host's tissues. The inner layer is the germinal membrane that is responsible for development of daughter cysts. In between is the acellular laminated membrane layer generated by the germinal membrane.

Pulmonary symptoms and signs result either from the compression of the surrounding tissue by an enlarging cyst or due to complications, such as rupture of infection. The cysts may rupture into a bronchus, the pleural cavity, blood vessels, mediastinal structures, or even the chest wall. Usually the diagnosis is either first suspected or decided upon by radiographic studies. In endemic areas, hydatid cysts are often the most common cause of well-defined, solid, single or multiple round densities. Rarely, these densities may calcify when a cyst ruptures into a bronchus and empties its contents only partially, leaving the ruptured membrane floating on a fluid level. This radiographic finding, often considered to be pathognomonic of pulmonary echinococcosis, is called the "water lily sign." When the air is trapped between two layers of the ruptured cyst wall, the crescent or meniscus sign can be demonstrated. Other radiologic signs that have been described are the double arch sign, mass migrating sign, solar eclipse sign, daughter cyst sign, and the ring-within-a-ring sign. A spontaneous remission, with coughing out of the hydatid laminated membrane, may leave only a few fibrotic streaks, or a patch of bronchiectasis on a chest roentgenogram. Spontaneous cures are thought to be unusual but may be more common than previously thought. Rupture can lead to sudden death from an anaphylactic reaction or tracheal obstruction. Residual cavities should be excised because of the risk of a residual germinal membrane creating a recurrent cyst or the residual cavity becoming secondarily infected. Pleural thickening, effusion, and pyopneumothorax occur in 15 to 20 percent of the cases. Computed tomography and ultrasound are used most commonly to evaluate for hepatic disease. All suspected hydatid cysts of the lung should have a CT of the chest, abdomen, and pelvis to evaluate for metastatic disease which may not be uncovered by other means. Computed tomography is also recommended for following a patient's clinical course.

Surgery should still be considered the treatment of choice, but chemotherapy does have a place in some patients' management. The surgical approach should try to preserve as much lung tissue as possible. The two main surgical options are aspiration or enucleation. In either case, the pericystic lining should be removed and the remaining cavity drained or, more preferably, closed by capottonage (suturing of the cavity walls). The surgical mortality is less than 2 percent. The spreading of daughter cysts during surgery is significantly reduced, by experienced surgeons, by injecting scolecisidal agents such as hypertonic saline solution into the cyst after aspiration and by meticulous packing around the cyst and thorough washing of the surgical field prior to closure. Recurrence rates range from 0.4 percent to 14 percent. Wedge or segmental resection, lobectomy, or pneumonectomy should only be considered for very large cysts or when surrounding parenchymal tissue is devitalized. Conserving lung tissue is important, since recurrent cysts of the lung can occur, which may require extensive surgery.

Mebendazole is the drug most thoroughly evaluated for hydatid disease. It is an oral anthelmitic previously used for parasites limited to the intestines. High-dose therapy, up to 40 mg/kg/day, for a prolonged period, up to several years has been used for unresectable, metastatic, and recurrent cyst disease and in inoperable patients and has had variable
responses. This is most likely due to its poor and variable absorption and inconsistent penetration into cysts. Mebendazole used as preoperative therapy sterilizes cysts and decreases the recurrence of cysts from surgical spread.

Albendazole has a more limited history of use and has shown promise but cannot be recommended as an adequate alternative to surgery at this time.

The diagnosis of pulmonary echinococcal disease is mainly circumstantial. There are no really useful tests apart from the presence of a cyst on a chest roentgenogram and, sometimes, eosinophilia. The intradermal test first developed by Casoni in 1911 has low specificity and sensitivity. The ELISA test has been recommended for large-scale screening, indirect hemagglutination for routine diagnosis, and the complement fixation test for evaluation for reinfection or continued infection, but further clinical trials need to be performed. Any obscure pulmonary mass or cyst in an individual from an endemic area should raise the suspicion of hydatid disease of the lungs.

REFERENCES

2 Leroux BT. Pulmonary hydatid disease. Thorax 1972; 27:365-75
5 Beggs I. The radiology of hydatid disease. AJR 1985; 145:639-48