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Endobronchial Echinococcosis Presenting as Non-Resolving Pneumonia

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Summary

Hydatid disease of the lungs is caused by larval cysts of the *Echinococcus* tapeworm. Pulmonary cysts may occasionally invade bronchi or pleura as a result of coughing, trauma or elevated intra-abdominal pressure. We present the case of a patient evaluated for non-resolving pneumonia whose radiographic and bronchoscopic findings were strikingly similar to those seen in pulmonary tuberculosis with endobronchial invasion; he was ultimately diagnosed with pulmonary echinococcosis. This case underscores the importance of considering unusual diagnoses even when typical features of more common conditions are present.

Keywords

Hydatid cyst; hydatid disease; pulmonary echinococcosis; tuberculosis; albendazole

CASE REPORT

A previously healthy 17-year-old male of Bedouin Arab descent began to complain of cough, generalized weakness and intermittent fever, with subsequent gradual weight loss. Four months after onset of symptoms, chest x-ray demonstrated right upper lobe pneumonia; he received a 10 day course of roxithromycin without improvement. Five weeks later, he began to experience hemoptysis; two days later he presented to the Emergency Department. Physical examination revealed reduced breath sounds over the upper left hemithorax. Complete blood count showed 18,700 white blood cells/mm³ with 72% granulocytes and 2.6% eosinophils. Erythrocyte sedimentation rate was 75 mm/hr. Chest X-ray demonstrated a left upper lobe infiltrate in the apical segment, without lymphadenopathy – unchanged in comparison to the previous x-ray. Treatment was initiated with intravenous penicillin G, while concurrently evaluating for tuberculosis. Purified protein derivative (PPD) skin testing was negative. Sputum and gastric aspirate were sent for mycobacterium culture and Ziehl-Neelsen acid-fast staining. One of the six specimens was positive for Ziehl-Neelsen staining. Computed tomography of the chest (figure 1a) revealed destructive left upper lobe

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consolidation with abscess formation and mediastinal lymphadenopathy. The scan also revealed a large liver cyst (figure 1c), with a maximal diameter of seven centimeters, interpreted to be most consistent with a hydatid cyst.

Flexible fiber-optic bronchoscopy revealed what appeared to be an endobronchial mass suspicious for tuberculosis in the apical segment of the left upper lobe. Multi-drug antituberculosis therapy was immediately begun (isoniazid, rifampin, pyrazinamide, and ethambutol) along with prednisone, pending results of bronchoalveolar lavage and culture. Polymerase chain reaction (PCR) testing for tuberculosis and atypical mycobacterium in the lavage fluid was negative. Echinococcus ELISA test (R-Biopharm) was found to be positive. Treatment was changed to albendazole and antituberculosis therapy, pending mycobacterial culture results.

Seven weeks after the initial hospitalization, he complained of continued weakness and weight loss; chest X-ray revealed lobar pneumonia of the left-upper lobe. He underwent flexible fiber-optic bronchoscopy which revealed mucoid substance occluding the left-upper lobe bronchi; after the bronchoscope was withdrawn, the patient expectorated large quantities of the white substance (figure 2a), and the material was sent for pathological examination.

Microscopic examination of the expectorated material revealed laminated germinal layers characteristic of echinococcal cysts, as well as echinococcal hooklets. Examination of hematoxylin & eosin – stained sections also revealed echinococcal hooklets (figure 2b). All mycobacterial cultures were negative.

He was treated with albendazole for six months. Eight months after presentation, follow-up ultrasonographic study demonstrated reduction in cyst size (from 70 to 12 mm) and chest CT (figure 1b) showed complete resolution of the previous findings. 18 months after presentation, the patient is entirely asymptomatic and has no exercise limitation.

DISCUSSION

Hydatid disease, or cystic echinococcosis, of the lungs is caused by larval cysts of the *Echinococcus* tapeworm – generally by the *Echinococcus granulosus* species. Infection results from inadvertent ingestion of tapeworm eggs in food or drink or through close contact with dogs or other infected carnivores (1,2). The cysts may be primary or secondary and form most commonly in the liver, followed by the lungs, and rarely in other organs, including the spleen, kidney, orbit, heart, brain and bone. In children, the lungs are the most common location of disease (3,4).

Lung cysts are usually caused by transphrenic spread of larvae through hepatic sinusoids (1) or by hematogenous spread of metacestodes following rupture of eggs in the stomach. They may also result from trans-lymphatic spread. Endobronchial disease may be primary, but more often results from secondary intrapulmonary spread: up to 30% of pulmonary cysts may rupture and invade bronchi or pleura as a result of coughing, trauma or elevated intra-abdominal pressure (2).

Patients with pulmonary hydatid cysts are initially asymptomatic (1). Symptoms, caused by enlargement or rupture of cysts, include fever, fatigue, chest pain, anorexia, and expectoration of cyst fragments. Patients may also present with complications such as asphyxia, pneumothorax, empyema or death resulting from invasion of the cardiovascular system.

Signs of pulmonary hydatid cysts are often apparent in a plain chest radiograph. Typical computerized tomography findings include the meniscus sign and the water-lily sign. Immunodiagnostic testing provides supportive evidence of pulmonary echinococcosis, although false-positive as well as false-negative reactions may occur. Direct microscopic examination of cyst fragments confirms the diagnosis, although percutaneous aspiration of lung cysts has generally been considered too risky to be used as a routine diagnostic tool (1).

Therapy

Spontaneous resolution may occur after expectoration of the cyst, although symptoms may nonetheless persist due to residual fragments or additional cysts. Treatment of choice is surgery, but pharmacotherapy with benzimidazoles (mebendazole or albendazole) for uncomplicated pulmonary echinococcosis has become more accepted as studies have been published establishing their efficacy, particularly in small and young cysts (5).

Treatment by percutaneous aspiration-injection-reaspiration (PAIR) is effective in liver cysts (6), but the technique shows more complications in pulmonary cysts and is therefore not usually performed (1). Surgical options include enucleation, wedge resection, segmentectomy, lobectomy and pneumonectomy. Accidental intraoperative spillage of cyst contents may result in anaphylaxis and/or dissemination with later cyst recurrence. Common complications of surgery include atelectasis, hemothorax and pneumothorax. Recommended follow-up consists of annual abdominal ultrasonography for five years, as well as a plain chest x-ray and computerized tomography after two to three years and again at five years (2).

At this point, a number of questions remain unanswered. First, the duration and choice of medical therapy: although albendazole has been traditionally administered for 3–6 months, optimal duration of therapy in patients who receive only medical treatment is unclear (7,8). Praziquantel has also been used for echinococcal disease; the combination of praziquantel and albendazole may be more effective than either drug alone (9,10). The optimal surgical intervention is also the subject of debate. While PAIR is rapidly gaining acceptance for treatment of liver cysts, reports of high complication rates have prevented its widespread use for pulmonary cysts. As previously mentioned, a number of surgical options exist; the role of video – assisted thoracoscopic surgery (VATS) has yet to be defined (11).

Our Patient presented a case of non-resolving pneumonia ultimately diagnosed as hydatid disease, simultaneously involving both the liver and lung – including bronchial involvement.

His disease may have resulted either from direct transphrenic spread, or from two separate primary hematogenous processes. He was unique in that clinical as well as radiographic findings (plain chest x-ray, chest CT) were highly suggestive of pulmonary tuberculosis, and one of the gastric fluid samples actually demonstrated acid-fast bacilli – a finding which ultimately turned out to be a “red-herring.” The case underscores the importance of considering echinococcosis in the differential diagnosis of non-resolving pneumonia (12) – even when the findings clearly point toward tuberculosis; it reminds us that we must make use of multiple tools, including fiber-optic bronchoscopy, in order to actively and aggressively seek out the diagnosis of hydatid disease.

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Figure 1.
Chest CT before (a) and after (b) treatment; abdominal CT before (c) and after (d) treatment

Figure 2.
Macroscopic (a) and microscopic (b) findings