Three atypical pulmonary hydatidosis lesions mimicking bronchial cancer from Turkey

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SUMMARY

Hydatid disease is endemic in Turkey. *Echinococcus granulosus* causes cystic echinococcosis mostly in the liver and lung. Although pulmonary hydatid cysts can be diagnosed by clinical and radiological findings, atypical or complicated lung lesions may be misdiagnosed. In the present study, three cases with hemoptysis and atypical lung lesions were diagnosed and treated as lung cancer or tuberculosis based on the clinical and laboratory findings along with the imaging data and fiberoptic bronchoscopy evaluation. Eventually, pathological examination of the bronchoscopic biopsy material confirmed the definitive diagnosis as pulmonary hydatidosis. The three patients presented herein emphasize the importance of considering pulmonary hydatid disease in the definitive diagnosis of atypical lung lesions such as bronchial carcinoma in echinococcosis endemic areas.

KEY WORDS: Pulmonary hydatidosis, Atypical lung lesions

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Hydatid disease or Echinococcosis in human is caused by the tapeworm *Echinococcus sp*. *Echinococcus granulosus* causes cystic echinococcosis and accounts for greater than 90% of human cases. The illness is endemic in sheep and cattle raising areas in Turkey, South America, Australia, and New Zealand (Morar and Feldman, 2003; Tor *et al.*, 2001). According to a retrospective study in Turkey, 14,789 cystic echinococcosis cases were diagnosed with cystic echinococcosis between the years 2001-2005. Of all the cases, 13.13%, 16.94%, 16.09%, 38.57%, 5.70%, 6.80%, and 2.75% cases were detected in the Marmara, Aegean, Mediterranean, central Anatolian, Black Sea, eastern Anatolian, and southeastern Anatolian regions, respectively (Yazar *et al.*, 2008).

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In another study, cystic echinococcosis seropositivity was 2.7% in Kayseri Rural Area, central Anatolia, Turkey between years 2000 and 2002 (Yazar *et al.*, 2006).

The adult form of *E. granulosus* lives in the canine intestine and sheds eggs that are eventually ingested by the intermediate host (usually sheep, cattle and accidentally human). Oncospheres are released from eggs in the gastrointestinal tract of intermediate hosts, and penetrate the intestine via the bloodstream.

They are distributed to the entire body but mostly cysts develop in the liver. The lung is the second most common site of involvement. In a study conducted in Hatay, southern Anatolia, Turkey, the rate of pulmonary echinococcosis was found to be 19.23% (Hakverdi *et al.*, 2008). In another study, among the 153 hydatid cases reported in Adana, southern Anatolia, Turkey between the years 2000-2006, lung involvement was detected in 54 cases (35.2 %) (Bal *et al.*, 2008). Pulmonary hydatid cysts typically present in the lower right lung and 75-90% of them are solitary and unilat-

eral. The cysts are composed of three layers in which the avascular outer pericystic layer is formed by host compressed tissue and chronic inflamatous cells; the ectocyst laminated membrane is an acellular laminar mucopolysaccaride layer; the inner endocyst layer is the germinal membrane that gives rise to larval scolices (Hussain *et al.*, 2003; Kennedy and Sharma, 1990; Saygi *et al.*, 1997; Ramos *et al.*, 2001).

Diagnosis of pulmonary hydatid cysts is mainly based on clinical and radiological findings. Clinical findings vary depending on the site of cyst and compression of the adjacent tissues. Complications such as cyst rupture can cause sudden cough, hemoptysis, expectoration of hydatid fluid and membranes or rarely anaphylaxis (Morar and Feldman, 2003; Kennedy and Sharma, 1990; Hussain *et al.*, 2003; Sayek *et al.*, 2004; Kurt *et al.*, 2008; Saygi *et al.*, 1997; Ramos *et al.*, 2001). Pulmonary hydatid cysts have a typical appearance and site of lesion on chest radiograms.

Radiological findings change when the cyst ruptures into the bronchus and the air erodes the layers of the cyst. In countries like Turkey where lung cancer and cystic hydatidosis are common causes of pulmonary diseases, the definitive diagnosis may become more sophisticated with cysts having a solid appearance or atypical localization mimicking bronchial carcinoma (Fidaner *et al.*, 2001; Bozkurt *et al.*, 2004; Yazar *et al.*, 2008). The present study evaluates the diagnostic approach to three uncomplicated atypical pulmonary hydatid lesions mimicking bronchial carcinoma.

CASE 1

A 45-year-old male farmer was admitted to the hospital with hemoptysis. There was no history of fever, chest pain, respiratory distress, cyanosis, chest trauma or foreign body inhalation. Family or personal history did not reveal any significant information.

The physical findings upon examination were normal. Laboratory findings were as follows; ESR: 177 mm/hour, white blood cell count was 9500 without eosinophilia. Tuberculin test showed an induration of 5 mm in diameter and acid resistant bacillus (ARB) was not detected in sputum using direct smear and culture techniques. Chest radiograph revealed a left hilar mass lesion with 3×4cm dimensions, suggesting a malignant lesion (Figure 1A). Abdominal ultrasound was normal.

Fiberoptic bronchoscopy evaluation of the lesion showed an obliterating vegetative tumoral lesion located in the bronchus of the left lower lobe superior segment indicative of necrotic bronchial carcinoma.

The pathological examination of brochoscopic biopsy material revealed cuticular membrane. Serological assays (Indirect hemagglutination test: IHA; Enzyme-Linked ImmunoSorbent Assay: ELISA IgG; Indirect immunofluorescent assay test: IFAT IgG) were positive for *E. granulosus*. Surgical excision confirmed the presence of hydatid disease and provided the cure. After surgery, chemotherapeutic regimens were not used and long term follow-up did not show any recurrence.

CASE 2

A 17-year-old male student had symptoms of fatigue, anorexia, weight loss, dyspnea and hemoptysis for one year. Family and personal history were unremarkable.

The physical examination and laboratory findings were normal. Tuberculin test showed an induration of 9 mm in diameter and ARB was not detected in sputum.

Chest radiograph revealed a heterogen paranchymal infiltration with air bronchograms in right paratracheal zone resembling pulmonary tuberculosis or pneumonia (Figure 1B). Abdominal ultrasound showed a cystic lesion in the right hepatic lobe. Fiberoptic bronchoscopy evaluation of the lesion displayed a white lesion in the right upper lobe orifice indicative of necrotic bronchial carcinoma.

The pathological examination of bronchoscopic biopsy material resulted as cuticular membrane. Serological assays were negative for *E. granulosus*. After surgery, pathological examination of the pulmonary parenchyma around the cyst showed fibrocytes and chronic inflammatory changes. Recurrence was not detected during long-term follow-up without using any chemotherapeutic agents.

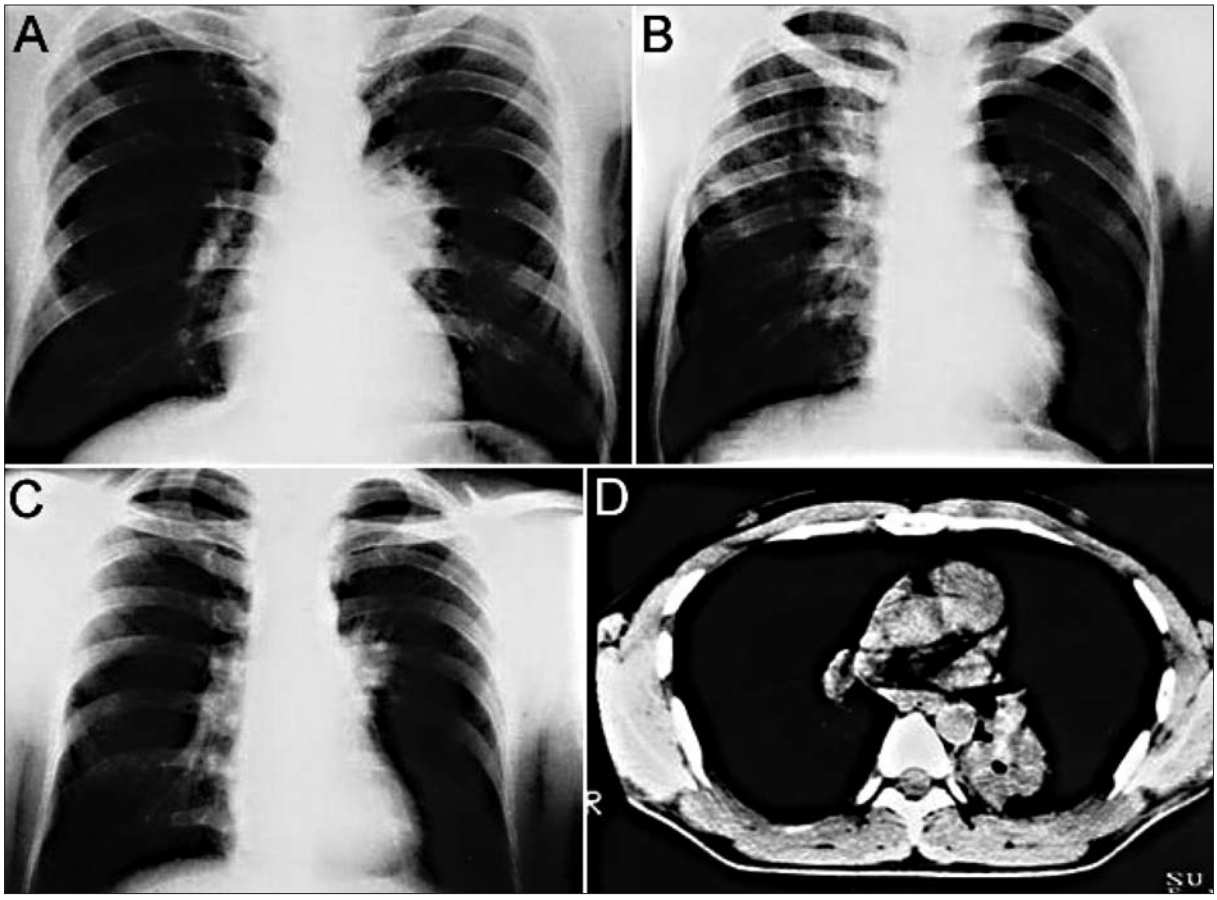

FIGURE 1 - *The radiological findings of three atypical pulmonary hydatid disease cases. (A-C) Chest radiographs of cases 1 and 3 showing left hilar mass lesions resembling malignant tumors, respectively. (B) Chest radiograph of case 2 demonstrating heterogeneous paranchymal infiltration with air bronchograms in right paratracheal zone suggestive of pulmonary tuberculosis or pneumonia. (D) Chest CT of case 3 showing cavitary solid tissue lesion with perifocal consolidation signifying bronchogenic carcinoma.*

CASE 3

A 34-year-old male worker was admitted to hospital with symptoms of cough, sputum, hemoptysis, left lateral chest pain, fatigue, anorexia and weight loss for 2-3 months. Family and personal history were unremarkable. The findings from a physical examination were normal. Laboratory findings were normal except ESR (34 mm/hour). Chest radiograph revealed a left hilar mass lesion with 3 cm diameter (Figure 1C). Chest computed tomography (CT) showed a cavitary solid tissue lesion with perifocal consolidation and reported as bronchogenic carcinoma (Figure 1D). Abdominal ultrasound was normal. Serological assays were negative for *E. granulosus*. Fiberoptic bronchoscopy evaluation of the lesion displayed a necrotic, soft white tumoral lesion growing into the lumen of a subsegmental bronchiole of the left lower lobe apical segment indicative of necrotic bronchial carcinoma. The pathological examination of forceps biopsy material resulted as cuticular membrane. In the next few days, the patient expectorated watery hydatid fluid, containing membrane particles. Albendazole treatment was initiated. Recurrence was not detected during long term follow-up.

The majority of pulmonary hydatid cyst cases can be diagnosed by the clinical findings and evaluation of data obtained from imaging techniques and serological methods. The diagnosis of pulmonary echinococcosis relies mostly on chest radiograph. Cysts appear as round or oval homogenous masses with smooth borders surrounded

by normal lung tissue and calcification is rare. Large cysts sometimes shift the mediastinum, cause atelectasis or pleural effusion. During enlargement, the cyst can erode into the bronchus and air can enter between the pericyst and endocyst leading to the thin crescent (meniscus) sign. As the air continues to enter this space, the cyst ruptures and air fills the endocyst. The air fluid level in the cyst and air like onion peel between pericyst and endocyst is called the Cumbo sign. After the contents of the cyst are partially expectorated, collapsed membranes inside the cyst form the serpent sign.

Another pathognomonic sign is the water lily sign which occurs after the endocyst detaches completely and the layer caves into the cyst cavity, floating freely on the cyst fluid (Morar and Feldman, 2003; Kennedy and Sharma, 1990). Chest computed tomography (CT) scan of unruptured cysts is homogeneous and often indistinguishable from some pulmonary lesions unless daughter cysts attached to the inner layer endocyst are detected or cyst rupture. However, CT scan can display the cystic appearance of a pulmonary mass lesion and help localize the cystic lesion for surgical purposes (Morar and Feldman, 2003; Koul *et al.*, 2000).

Laboratory findings are non-specific. In case of leakage from the cyst, eosinophilia can be detected in 15% of cases (Morar and Feldman, 2003). Serological assays support the diagnosis but they are positive in only ~50% of patients with pulmonary hydatidosis compared to 90% of cases with liver cysts (Morar and Feldman, 2003; Hussain *et al.*, 2003). In addition, false positive results can be seen due to nonspecific cross reactivity with other helminthes, cancer and immune disorders (Hussain *et al.*, 2003). False negative results can be detected depending on the cyst integrity and site of the lesion in lungs (Hussain *et al.*, 2003). Thus, a negative serology does not rule out hydatid disease.

Pulmonary hydatid disease has characteristic radiological findings, but as the cyst turns into a complicated or atypical cyst, the definitive diagnosis is delayed or hampered resulting in higher morbidity and mortality (Turgut *et al.*, 2007; Pedrosa *et al.*, 2000). Kurt *et al.* presented a case who was diagnosed as breast cancer and at the same time had bilateral pulmonary multiple masses of different size and irregular shape which were considered metastases to lung according to the results of chest radiograph and chest CT. Regardless of the increased uptake of fluoro-deoxy-glucose (FDG) by breast cancer and its metastatic lymph nodes, the pulmonary lesions did not take up FDG during positron emission tomography (PET). The result of FDG-PET excluded metastasis and indirect hemaglutination test confirmed pulmonary echinococcosis (Kurt *et al.*, 2008). Tor *et al.* evaluated 13 complicated patients using fiberoptic bronchoscopy and diagnosed pulmonary hydatid disease in three patients (Tor *et al.*, 2001).

Saygi *et al.* diagnosed 24 patients with pulmonary hydatid disease after thoracotomy 14 of whom were preoperatively diagnosed by fiberoptic bronchoscopy evaluation and subsequent cytological and histopathological examination of the biopsy material (Saygi *et al.*, 1997).

In the present study, 116 patients were diagnosed as pulmonary hydatid disease in which three cases were initially evaluated as pulmonary malignancy based on radiological mass lesion findings by chest radiograph/chest CT. In addition, fiberoptic bronchoscopy evaluation of each lesion was assessed as necrotic bronchial carcinoma. However, bronchoscopic biopsy demonstrated pulmonary hydatid disease caused by *E. granulosus*. Serology was positive for *E. granulosus* only in case 1. Case 2 was evaluated as tuberculosis or pneumonia based on chest radiograph and white undefined lesion by fiberoptic bronchoscopy.

The patient received 10 days of non-specific chemotherapy and 35 days anti-tuberculosis therapy. Clinical, radiological and laboratory findings and fiberoptic bronchoscopy evaluation did not facilitate the definitive diagnosis of the patients. However, the pathological examination of lung biopsy material dispersed doubts as to the patients' diagnosis.

Lung cancer and pulmonary hydatidosis are both prevalent diseases in Turkey. The lesions presented herein were initially considered bronchial carcinomas based on the radiological appearance but diagnosed as pulmonary hydatidosis eventually. In addition, perforated pulmonary hydatidosis should be ruled out in hilar lesions, especially in young patients. In conclusion, three patients presented herein show the importance of considering pulmonary hydatidosis in the defin-

itive diagnosis and timely treatment of patients presenting with uncomplicated atypical lung lesions mimicking lung cancer and tuberculosis in echinococcosis endemic areas.

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