Pulmonary echinococcosis presenting as a pulmonary mass with chest pain, fever and non-productive cough

Echinococcosis or hydatid disease is caused by the larvae of the tapeworm Echinococcus. As it occurs only rarely in young people who reside in urban areas and do not report contact with animals the case is reported here of a 16 year-old girl who presented with chest pain, fever, and non-productive cough, in whom the diagnosis of pulmonary cystic echinococcosis was made. The patient underwent surgery for excision of the mass and was discharged on oral albendazole treatment. The diagnosis of pulmonary cystic echinococcosis should always be included in the differential diagnosis of a young patient presenting with a lung mass, chest pain, non-productive cough, and fever.

In cystic echinococcosis, the liver and the lungs are the most frequently involved organs. In adults, cysts occur more often in the liver, followed by the lungs, whereas in children, the lung is the predominant site for cyst detection. The lung allows for the faster growth of the cyst due to its compressible nature, rich vascularization, and negative pressure.

Clinically, most infected individuals remain asymptomatic until the cyst in the lung becomes large enough to cause pressure symptoms. The most common symptoms are:
cough (53–62%), chest pain (49–91%), dyspnea (10–70%) and hemoptysis (12–21%). Other less frequent symptoms include nausea and vomiting and thoracic deformation. Complications, such as cyst rupture or aggregated infection, may change the clinical presentation.

Diagnosis is made by chest X-ray or computed tomography (CT), and supported by serological testing. On chest X-ray, the cysts appear well defined as a rounded mass of uniform density, occupying a part of one or of both hemithoraces. The better imaging definition of CT is useful to exclude alternative differential diagnoses and recognize complicated cysts.

CASE REPORT

A 16-year-old girl presented at the General Hospital of Kalamata with a week's history of vague chest pain in the right hemithorax, fever, and non-productive cough. She was not a smoker and reported no previous health problems. The medical history was free of chronic diseases or surgery. The patient was resident in an urban area and reported no travel abroad or contact with animals.

On physical examination, the respiratory sounds were reduced over the right lung field. The blood pressure, heart and respiratory rate, temperature, and SpO\textsubscript{2} were normal. Laboratory tests showed a small increase of C reactive protein (CRP): 2.98 mg/dL (normal value <0.5 mg/dL), a slight decrease in Hct: 28%, Hgb: 8.4 g/dL (MCV: 70.7, MCH: 21.2, MCHC: 30). WBC: 9,590/mm\textsuperscript{3} (68.2% neutrophils and 20.1% lymphocytes). Blood glucose level and renal and liver function tests were normal, as was the urine examination. Chest X-ray revealed a mass on the right lung (fig. 1). CT scan of the chest confirmed the presence of the mass, which had a diameter of 9 cm and the morphological characteristics of a cyst (fig. 2). Abdominal ultrasound (US) showed no pathology in the liver or spleen or any other abnormality.

The patient was referred to a thoracic surgeon at a tertiary care hospital in Athens, where she underwent surgery for the excision of the mass. A right thoracotomy was performed, followed by lower lobectomy, including the mass. The histopathological report established the diagnosis of an echinococcus cyst. The postoperative course was uneventful and the patient was discharged home on oral albendazole treatment.

DISCUSSION

The incidence of echinococcosis is relatively low in Greece, as in most European countries. Specifically, epidemiological data for echinococcosis show that the annual incidence of disease in Greece was 0.14 cases/100,000 population/year, for the years 2005–2009.

In the majority of patients, as the case presented here, a single organ is involved and a solitary cyst is formed. In adults, cysts appear more often in the liver, followed by the lung (10–30%) and other organs (10%), while in children, cysts appear more often in the lung. Twenty to forty percent of patients with pulmonary cysts also have liver cysts, and almost 60% of pulmonary cysts are located in the right lung.

Most pulmonary cysts are discovered incidentally on a routine X-ray examination. It appears that most persons with small lung cysts remain asymptomatic for 5 to 20
years after infection, until the cyst enlarges sufficiently to cause symptoms. The most symptoms are caused by the pressure of the cyst on the surrounding tissues. Children and teenagers may be asymptomatic despite having a large cyst, because of the higher elasticity of their lung parenchyma. The most common symptoms are cough, chest pain, dyspnea and hemoptysis. The patient reported here presented with the triad of chest pain, non-productive cough and fever. The clinical presentation changes when a complication arises, such as cyst rupture or infection. Lung cysts may rupture spontaneously or as a result of trauma or infection, and their contents (fragments of larval tissue, protoscoleces) may be transferred to either the bronchial tree, causing cough, chest pain and hemoptysis, or the pleural cavity, giving rise to pneumothorax, pleural effusion and empyema.

Cyst rupture may also lead to the release of antigenic material causing immunological reactions which produce generalized symptoms, such as fever and acute hypersensitivity reactions. The diagnosis of cystic echinococcosis is based on imaging and serological methods. The most commonly used imaging examination for the evaluation of a pulmonary cyst is chest X-ray. On chest X-ray, the cyst appears well defined as a rounded mass of uniform density, with a diameter of 1–20 cm, surrounded by normal lung tissue. Calcification is not common in pulmonary cysts. The better imaging definition provided by CT scanning offers more information about the cystic nature and is particularly useful in case of a complicated cyst.

Immunodiagnostic tests are sometimes used to support the diagnosis. The sensitivity of serological tests in the case of a pulmonary cyst is low (only 50% of patients have a positive test); thus, tests for specific antibodies and antigens (immunoglobulin G1 or G2, antigen B, antigen S) are not often used.

Surgery is the main therapeutic approach, because excision of the cyst ensures complete removal of the parasite and the patient is cured, although surgery does not always prevent recurrence. The most frequent complications are pleural infection and prolonged air leakage and the operative mortality rate is less than 2%.

When surgery is not available or complete removal is not feasible, chemotherapy with benzimidazoles is the preferred treatment. Side effects of benzimidazoles include mild hepatotoxicity, leukopenia, hair loss, and gastric disturbances. Albendazole is preferred to mebendazole because of its better bioavailability. Albendazole given for several months at a dosage of 400 mg twice a day is efficacious for pulmonary cysts. A newer benzimidazole compound, oxfendazole, has been studied in animal models and preliminary results suggest that it might be a more effective compound.

In conclusion, echinococcosis is a disease endemic in Greece that presents with varied symptomatology. The diagnosis of pulmonary cystic echinococcosis should always be included in the differential diagnosis of a young patient presenting with a lung mass, chest pain, non-productive cough, and fever.
References

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