Endosonography of a Pulmonary Artery Obstruction in Echinococcosis

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Established Facts

- A minority of echinococcosis cases involves multiple sites, and cysts in the pulmonary artery (PA) are very rare.

Novel Insights

- Echinococcosis may cause severe PA obstruction with sudden thoracic pain and shortness of breath.
- In case of uncertainty of the diagnosis at noninvasive imaging, subsequent minimally invasive endobronchial ultrasound is an excellent diagnostic technique to visualize intra-PA cysts and differentiate it from pulmonary embolism.

Key Words

Pulmonary artery obstruction · Endobronchial ultrasound · Echinococcosis · Hydatidosis · Pulmonary embolism

Abstract

A 44-year-old woman with a history of pulmonary embolism and abdominal echinococcosis complained of sudden thoracic pain and shortness of breath. A D-dimer of 77.5 mg/l (reference ≤ 0.5 mg/l) was found. Chest CT scan revealed obstruction of the right lower and middle lobe pulmonary artery (PA). Anticoagulation therapy was initiated for the presumed diagnosis of recurrent pulmonary embolism. However, due to persistent symptoms of dyspnea, follow-up CT

angiography of the chest was performed 3 months later. A persistent PA obstruction was found and the presumed diagnosis of embolism was questioned. Subsequently, endobronchial ultrasound (EBUS) imaging was performed to support an alternative diagnosis. EBUS imaging showed an inhomogeneous, sharply demarcated, intravascular lesion with round hypoechoic areas compatible with cysts. The diagnosis of embolism was rejected and treatment with albendazole was initiated for pulmonary echinococcosis. Echinococcosis is a parasitic disease and cystic spread in the PA is exceptional. The patient has remained stable for more than 4 years. In case of disease progression, including progressive PA obstruction or life-threatening hemoptysis, surgical resection will be considered.

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Fig. 1. CT angiography. Axial view showing a right PA thrombus involving the right lower and middle lobe (arrow).

Fig. 2. Follow-up CT angiography. Coronal view showing PA obstruction indicative of echinococcal cysts localization. Abdominal echinococcal cysts with typical layers are shown (arrow).

Introduction

Echinococcosis caused by *Echinococcus granulosus*, also known as hydatidosis or hydatid cysts, is a parasitic disease known from the time of Hippocrates [1]. Echinococcosis is prevalent in regions of Eurasia, several South American countries, North and Central America, and Africa. In the Western world, it is predominantly seen as an imported disease in immigrants from endemic countries [2, 3]. Echinococcosis cysts grow slowly (1–30 mm in diameter yearly) and about 80% occur in a single organ. Moreover, echinococcosis may involve any organ including the lungs, but cysts in the pulmonary artery (PA) are very rare [1, 4].

Case Report

A 44-year-old woman with a history of pulmonary embolism, left-sided nephrectomy, and abdominal echinococcosis complained of sudden thoracic pain and shortness of breath. She discontinued anticoagulation 7 years before presentation. Physical examination revealed a normal blood pressure and a saturation of 99%. Due to

Fig. 3. EBUS image demonstrating intra-PA obstruction by echinococcal cysts surrounded by organized thrombus. The hypoechoic aspect and absence of Doppler flow strongly support the cystic nature of the obstruction. The surrounded organized tissue is suspected for in situ thrombosis. C = Cyst.

the sudden onset of symptoms and a history of pulmonary embolism, a D-dimer test was performed, which revealed a D-dimer of 77.5 mg/l (reference ≤ 0.5 mg/l). She was initiated on low molecular weight heparin, and a CT angiography was performed under the suspicion of pulmonary embolism. The axial image on this CT angiography revealed a right lower and middle lobe PA obstruction (fig. 1). The diagnosis of recurrent pulmonary embolism was made and anticoagulation therapy was given. A follow-up CT angiography 3 months later showed persistent PA obstruction, and the presumed diagnosis of embolism was questioned as PA cysts seemed to be present (fig. 2). Importantly, these lesions on the coronal image resemble cysts compatible with echinococcosis. Endobronchial ultrasound (EBUS) that can be used to detect centrally located pulmonary emboli [5–7] demonstrated an inhomogeneous intravascular lesion containing multiple sharply demarcated round hypoechoic areas. In absence of Doppler flow, these lesions are compatible with the diagnosis of cysts (fig. 3). The combination of clinical presentation, follow-up CT angiography, and EBUS imaging made us reject the initial diagnosis of pulmonary embolism and support the diagnosis of PA echinococcosis. No signs of pulmonary hypertension on transthoracic echocardiography were present. In our patient, a prolonged (4 years up to now) treatment with albendazole has been chosen, as radical surgical treatment may lead to serious complications in this stage and is noncurative due to widely disseminated disease. The patient has remained stable for more than 4 years. In case of disease progression, including progressive PA obstruction with the risk of pulmonary hypertension or life-threatening hemoptysis, surgical resection will be considered.

Discussion

Although a minority of echinococcosis cases involves multiple sites, and cysts in the PA are very rare, this case demonstrates that echinococcosis may cause severe PA

obstruction with a presentation of sudden thoracic pain and shortness of breath. In case of uncertainty of the diagnosis at noninvasive imaging, subsequent minimally invasive EBUS is an excellent diagnostic technique to visualize intra-PA cysts and differentiate them from pulmonary embolism. Due to the completely different therapeutic consequences, we decided that we needed more assurance of the diagnosis.

Morbidity in echinococcosis is usually secondary to spontaneous rupture of the cyst (with or without anaphylaxis), infection of the cyst, or dysfunction of the involved organ due to cyst growth. If located in the PA, pulmonary hypertension is a complication which has been described [8–10]. Mortality is secondary to anaphylaxis, systemic complications of the cysts (sepsis, respiratory failure) or perioperative complications. Surgical resection and percutaneous drainage are accepted treatments of echinococcosis cysts if located in the liver, spleen, or subcutaneously [1]. In case of PA, localization surgical resection of echinococcal cysts by arteriotomy with or without the need of pneumonectomy or pulmonary endarterectomy can be considered. Surgical treatment of PA echinococcosis has been reported but is considered high risk [11–13]; therefore, this decision must be taken very carefully. To our knowledge, percutaneous drainage of the PA has not yet been performed.

Financial Disclosure and Conflicts of Interest

The authors have no conflicts of interest to disclose.

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