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A Girl with Disseminated Thoracic Echinococcosis

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Abstract

We present an unusual case of extensively disseminated pulmonary and thoracic echinococcosis in a 10 year old girl. Thoracic echinococcosis should be considered in any child from an endemic country with cystic lesions on chest imaging. Confirmation of the diagnosis can be difficult and is important before surgery to reduce the risk of further dissemination and anaphylaxis.

Key words: echinococcus, multiple lung cysts

Introduction

Human echinococcosis is caused by larval forms of the echinococcus tapeworm, found in the small intestine of carnivores. The highest prevalence of cystic echinococcosis is in the entire Mediterranean littoral, southern South America, southern and central parts of the former Soviet Union, central Asia, China, Australia and parts of Africa. In endemic areas the annual incidence of new cases ranges from 1 to 200 per 100,000 inhabitants. Most primary infections in humans consist of a single cyst. Twenty to 40% have disseminated disease with multiple cysts and multiple organ involvement. The most common site is the liver followed by the lungs and less frequently the spleen, kidneys, heart, bone and central nervous system. Infections acquired during childhood may not be diagnosed until adulthood because of the slow growing nature of the cyst.^{1,2}

Patient

A 10 year old girl from Iraq was routinely screened in 2008 for tuberculosis after immigration to the Netherlands. She reported chronic cough and chest pain during the past year and mentioned limited exercise tolerance. A plain radiograph and computed tomography (CT) scan of the chest showed a solitary cystic lesion. Thoracic and abdominal ultrasound examination excluded cystic lesions in the liver and spleen, and showed no evidence of multiple cysts in the chest.

Differential diagnosis at that time included infected congenital cystadenomatoid malformation with pleural adhesions, tuberculosis and echinococcus cyst. Tuberculosis was excluded by a negative purified protein derivative test and echinococcus serology was negative. Eosinophilia in combination with exercise-induced dyspnea was interpreted as suggestive of asthma.

A second CT scan performed a few months later, showing the cyst unchanged and, as a new finding, some small pleural nodules. Flexible bronchoscopy showed no abnormalities on inspection. Bronchoalveolar lavage cultures and tests for galactomannan in bronchoalveolar lavage were negative. Bronchial mucosal biopsies showed a thickened basal membrane consistent with a diagnosis of asthma. She was treated with a course of antibiotics. Her chest pain had disappeared and she felt well. She reported having coughed up salty fluid with some white matter recently on a single occasion, but this was not considered alarming. Surgery was scheduled to remove the cyst, which was considered to be a congenital cystadenomatoid malformation. There was a considerable delay of surgery due to noncompliance. One year later, a magnetic resonance imaging scan (MRI) before surgery showed a massive amount of cystic lesions, occupying a large part of the left hemithorax (figure 1). At that time, repeated echinococcus serology was positive, and the diagnosis of disseminated thoracic echinococcosis was made. After albendazole pretreatment and with precautions during surgery, including irrigation with hypertonic saline as a scolicidal agent to prevent complications, a left lower lobectomy and resection of a large number of cysts was performed. Pathologic examination of the resected tissue confirmed the diagnosis of multiple echinococcus cysts. We treated her for 6 months with albendazole (10mg/kg per day). Follow-up by MRI 3 months after surgery showed no recurrence of cysts.

Discussion

The clinical manifestations of cystic echinococcosis are variable.¹⁻⁵ Cysts are often tolerated well until they reach a size that causes complaints because of their volume. In the lungs, ruptured cysts can be evacuated via the bronchi or spill into the pleural space, and dissemination of protoscolices can result in secondary cysts.¹ Cyst rupture can precipitate reactions from mild

allergy to severe anaphylaxis. Detection of echinococcus cysts in the lung is possible using plain chest radiograph or, preferably, CT scan, and cysts in other organs can be found using CT, MRI and ultrasonography.² Antibody assays can confirm the diagnosis, although most patients have no detectable immune response in uncomplicated cases because the antigen is hidden from immune surveillance. A ruptured cyst is associated with a higher immune response than an intact cyst. Eosinophilia is present in <25% of infected patients.¹

The presented case had a delay in diagnosis, and a long delay in surgical treatment due to an unfortunate combination of diagnostic uncertainty and noncompliance. Could this diagnosis have been made earlier? Retrospectively the salty taste and the expectorated white matter were a sign of endobronchial cyst rupture and probably the pleural nodules on the CT were small daughter cysts instead of pleural adhesions after infection. It is important to realize that a negative serology by no means excludes active echinococcus infection.^{3,4} Repeating the serology immediately after the second CT might have revealed the diagnosis sooner.

In this case, delay in diagnosis resulted in an extensively disseminated pulmonary and thoracic echinococcosis for which lobectomy had to be performed.

We believe that thoracic echinococcosis should be considered in any patient from an endemic country who has cystic lesions on chest imaging. Confirmation of the diagnosis can be difficult, but should be attempted before surgery to reduce the risk of dissemination and anaphylaxis.

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Figure legends

Figure 1

Magnetic resonance imaging (MRI) scan prior to surgery, showing the left hemithorax largely occupied by cysts.

ACCEPTED

Figure 1

