

CASE REPORT ΕΝΔΙΑΦΕΡΟΥΣΑ ΠΕΡΙΠΤΩΣΗ

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.

Echinococcosis or hydatid disease is caused by the larvae of the tapeworm *Echinococcus*.¹⁻⁴ Six species are recognized, but the vast majority of infestations in humans are caused by *E. granulosus*. *E. granulosus* causes cystic echinococcosis, which has a worldwide distribution and is the commonest type in the Mediterranean countries. Humans are exposed less frequently to *E. multilocularis*, which causes alveolar echinococcosis. *E. vogeli* and *E. oligarthrus* are rare species and cause polycystic echinococcosis.¹⁻⁴ The clinical significance of *E. shiquicus* and *E. felidis* is not known.^{1,2}

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,⁵ 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.⁶

Clinically, most infected individuals remain asymptomatic until the cyst in the lung becomes large enough to cause pressure symptoms.⁷ The most common symptoms are:

ARCHIVES OF HELLENIC MEDICINE 2013, 30(2):225-228
ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2013, 30(2):225-228

G. Michas,¹
E. Varytimiadi,¹
A. Varytimiadi,²
E. Toli,¹
E. Kyriakopoulos,³
M. Michail,³
E. Vryonis^{1,4,5}

¹Department of Internal Medicine, General Hospital of Kalamata, Kalamata

²Third Department of Obstetrics and Gynecology, "Attikon" University General Hospital, Athens

³Department of Radiology, General Hospital of Kalamata, Kalamata

⁴Infection Prevention and Control Committee, General Hospital of Kalamata, Kalamata

⁵North Middlesex University Hospital NHS Trust, London, UK

Εμφάνιση πνευμονικής
εχينوκοκκίασης ως πνευμονική
μάζα με πλευριτικό άλγος, πυρετό
και μη παραγωγικό βήχα

Περίληψη στο τέλος του άρθρου

Key words

Chest pain
Cough
Echinococcosis
Fever
Lung mass

Submitted 18.12.2012

Accepted 10.1.2013

cough (53–62%), chest pain (49–91%), dyspnea (10–70%) and hemoptysis (12–21%).⁸ Other less frequent symptoms include nausea and vomiting and thoracic deformation.⁹ 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.⁸

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 SpO₂, 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/mm³ (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.¹⁰ 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 2005–2009.¹¹

In the majority of patients, as the case presented here,



Figure 1. Chest X-ray in a 16-year-old female, showing massive right-sided lung mass.

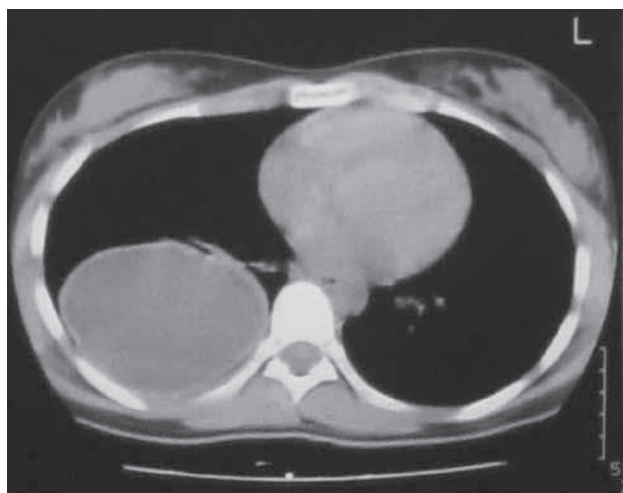


Figure 2. Computed tomography (CT) of the thorax in a 16-year-old female, showing the presence of a right sided mass, with a diameter of 9 cm and the morphological characteristics of a cyst.

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%),^{12,13} while in children, cysts appear more often in the lung.⁶ 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.^{14–16}

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.⁸ 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.¹⁷ 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 larvae tissue, protoscolexes) 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.^{18,19} 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.¹

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.²⁰ 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.²¹ 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.¹⁸ 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 5) are not often used.²²

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.²³ The most frequent complications are pleural infection and prolonged air leakage and the operative mortality rate is less than 2%.²⁴

When surgery is not available or complete removal is not feasible, chemotherapy with benzimidazoles is the preferred treatment.²³ Side effects of benzimidazoles include mild hepatotoxicity, leukopenia, hair loss, and gastric disturbances. Albendazole is preferred to mebendazole because of its better bioavailability.²⁵ 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.²⁶

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.

ΠΕΡΙΛΗΨΗ

Εμφάνιση πνευμονικής εχινοκοκκίασης ως πνευμονική μάζα με πλευριτικό άλγος, πυρετό και μη παραγωγικό βήχα

Γ. ΜΙΧΑΣ,¹ Ε. ΒΑΡΥΤΙΜΙΑΔΗ,¹ Α. ΒΑΡΥΤΙΜΙΑΔΗ,² Ε. ΤΟΛΗ,¹ Ε. ΚΥΡΙΑΚΟΠΟΥΛΟΣ,³ Μ. ΜΙΧΑΗΛ,³ Ε. ΒΡΥΩΝΗΣ^{1,4,5}

¹Τμήμα Εσωτερικής Παθολογίας, Γενικό Νοσοκομείο Καλαμάτας, Καλαμάτα, ²Τμήμα Μαιευτικής και Γυναικολογίας, Πανεπιστημιακό Γενικό Νοσοκομείο «Αττικόν», Αθήνα, ³Ακτινολογικό Τμήμα, Γενικό Νοσοκομείο Καλαμάτας, Καλαμάτα, ⁴Επιτροπή Νοσοκομειακών Λοιμώξεων, Γενικό Νοσοκομείο Καλαμάτας, Καλαμάτα, ⁵North Middlesex University Hospital NHS Trust, Λονδίνο, Ηνωμένο Βασίλειο

Αρχία Ελληνικής Ιατρικής 2013, 30(2):225–228

Η εχινοκοκκίαση ή υδατίδωση προκαλείται από τις προνύμφες της ταινίας *Echinococcus*. Αν και είναι ενδημική νόσος στην Ελλάδα, σπανίως εμφανίζεται σε νέους που κατοικούν σε αστικές περιοχές και δεν αναφέρουν επαφή με τα ζώα. Το αναφερόμενο ενδιαφέρον περιστατικό αφορά σε ένα 16χρονο κορίτσι που παρουσιάστηκε με πλευριτικό άλγος, πυρετό και μη παραγωγικό βήχα και διαγνώστηκε με πνευμονική εχινοκοκκίαση. Η ασθενής παραπέμφθηκε σε θωρακοχειρουργό και υποβλήθηκε σε χειρουργική επέμβαση για την εκτομή της μάζας. Η μετεχειρητική πορεία της ήταν ομαλή και εξήλθε με πρωτόκολλο θεραπείας αλβενδαζόλης. Η διάγνωση της πνευμονικής εχινοκοκκίασης πρέπει πάντα να περιλαμβάνεται στη διαφορική διάγνωση ενός νεαρού ασθενούς που παρουσιάζεται με πνευμονική μάζα συνοδευόμενη από πλευριτικό άλγος, πυρετό και μη παραγωγικό βήχα.

Λέξεις ευρητηρίου: Βήχας, Εχινοκοκκίαση, Πλευριτικό άλγος, Πνευμονική μάζα, Πυρετός

References

- McMANUS DP, ZHANG W, LI J, BARTLEY PB. Echinococcosis. *Lancet* 2003, 362:1295–1304
- CRAIG PS, McMANUS DP, LIGHTOWLERS MW, CHABALGOITY JA, GARCIA HH, GAVIDIA CM ET AL. Prevention and control of cystic echinococcosis. *Lancet Infect Dis* 2007, 7:385–394
- MORAR R, FELDMAN C. Pulmonary echinococcosis. *Eur Respir J* 2003, 21:1069–1077
- ROMIG T, DINKEL A, MACKENSTEDT U. The present situation of echinococcosis in Europe. *Parasitol Int* 2006, 55(Suppl):S187–S191
- SIRACUSANO A, TEGGI A, ORTONA E. Human cystic echinococcosis: Old problems and new perspectives. *Interdiscip Perspect Infect Dis* 2009, 2009:474368
- ARROUD M, AFIFI MA, EL GHAZI K, NEJJARI C, BOUABDALLAH Y. Lung hydatid cysts in children: Comparison study between giant and non-giant cysts. *Pediatr Surg Int* 2009, 25:37–40
- TORGERSON PR, DEPLAZES P. Echinococcosis: Diagnosis and diagnostic interpretation in population studies. *Trends Parasitol* 2009, 25:164–170
- SANTIVANEZ S, GARCIA HH. Pulmonary cystic echinococcosis. *Curr Opin Pulm Med* 2010, 16:257–261
- ARINC S, KOSIF A, ERTUGRUL M, ARPAG H, ALPAY L, UNAL O ET AL. Evaluation of pulmonary hydatid cyst cases. *Int J Surg* 2009, 7:192–195
- EUROPEAN CENTRE FOR DISEASE PREVENTION AND CONTROL. *Annual epidemiological report. Reporting on 2009 surveillance data and 2010 epidemic intelligence data*. ECDC, Solna, Sweden, 2011 (available at: www.ecdc.europa.eu)
- HELLENIC CENTER FOR DISEASE CONTROL AND PREVENTION. *Epidemiological data on echinococcosis in Greece, 2005–2009*. KEELPNO, Athens, 2011
- SCHANTZ P. Echinococcosis. In: Guerrant RL, Walker DH, Weller PF (eds) *Tropical infectious diseases: Principles, pathogens and practice*. WB Saunders, Philadelphia, 1999:1005–1025
- KING CH. Cestods (tapeworms). In: Mandell GL, Bennett JE, Dolin R (eds) *Principles and practice of infectious diseases*. Churchill Livingstone, New York, 1995:2544–2553
- GÓMEZ R, MORENO E, LOINAZ C, De LA CALLE A, CASTELLON C, MANZANERA M ET AL. Diaphragmatic or transdiaphragmatic thoracic involvement in hepatic hydatid disease: Surgical trends and classification. *World J Surg* 1995, 19:714–719; discussion 719
- JERRAY M, BENZARTI M, GARROUCHE A, KLABI N, HAYOUNI A. Hydatid disease of the lungs. Study of 386 cases. *Am Rev Respir Dis* 1992, 146:185–189
- ARIBAS OK, KANAT F, GORMUS N, TURK E. Pleural complications of hydatid disease. *J Thorac Cardiovasc Surg* 2002, 123:492–497
- DOPCHIZ MC, ELISSONDO MC, ANDRESIUK MV, MAIORINI E, GUTIÉRREZ AM, MUZULIN PM ET AL. Pediatric hydatidosis in the south-east of the Buenos Aires province, Argentina. *Rev Argent Microbiol* 2009, 41:105–111
- TURGUT AT, ALTINOKT, TOPÇU S, KOŞAR U. Local complications of hydatid disease involving thoracic cavity: Imaging findings. *Eur J Radiol* 2009, 70:49–56
- DZIRI C, HAOUET K, FINGERHUT A, ZAOUCHE A. Management of cystic echinococcosis complications and dissemination: Where is the evidence? *World J Surg* 2009, 33:1266–1273
- HERNÁNDEZ-GONZÁLEZ A, MURO A, BARRERA I, RAMOS G, ORDUÑA A, SILES-LUCAS M. Usefulness of four different *Echinococcus granulosus* recombinant antigens for serodiagnosis of unilocular hydatid disease (UHD) and postsurgical follow-up of patients treated for UHD. *Clin Vaccine Immunol* 2008, 15:147–153
- BALIKIAN JP, MUDARRIS FF. Hydatid disease of the lungs. A roentgenologic study of 50 cases. *Am J Roentgenol Radium Ther Nucl Med* 1974, 122:692–707
- GOTTSTEIN B, REICHEN J. Hydatid lung disease (echinococcosis/hydatidosis). *Clin Chest Med* 2002, 23:397–408
- BRUNETTI E, KERN P, VUITTON DA; Writing Panel for the WHO-IWGE. Expert consensus for the diagnosis and treatment of cystic and alveolar echinococcosis in humans. *Acta Trop* 2010, 114:1–16
- SHEHATHA J, ALIZZI A, ALWARD M, KONSTANTINOV I. Thoracic hydatid disease; a review of 763 cases. *Heart Lung Circ* 2008, 17:502–504
- STAMATAKOS M, SARGEDI C, STEFANAKI CH, SAFIOLEAS C, MATTHAIPOULOU I, SAFIOLEAS M. Anthelmintic treatment: An adjuvant therapeutic strategy against *Echinococcus granulosus*. *Parasitol Int* 2009, 58:115–120
- GAVIDIA CM, GONZALEZ AE, LOPERA L, JAYASHI C, ANGELATS R, BARON EA ET AL. Evaluation of nitazoxanide and oxfendazole efficacy against cystic echinococcosis in naturally infected sheep. *Am J Trop Med Hyg* 2009, 80:367–372

Corresponding author:

E. Vryonis, North Middlesex University Hospital NHS Trust, Sterling Way, London N18 1QX, UK
e-mail: evryonis@gmail.com