A Case of Pulmonary Hypertension due to Chronic Pulmonary Hydatid Cyst Embolism

Yurdanur Erdoğan[1], Ülkü Yılmaz Turay[1], Pelin Pınar Deniz[1], Çiğdem Biber[1], Pınar Ergun[1], Bülent Çiftçi[1], Aydın Yılmaz[1], Atilla Gökçek[2]

[1]Atatürk Chest Disease and Chest Surgery Hospital, Pulmonary Disease, Ankara, Turkey
[2]Atatürk Chest Disease and Chest Surgery Hospital, Radiology, Ankara, Turkey

Abstract
Hydatid pulmonary embolism is an uncommon condition resulting from the rupture of a hydatid cyst. We report a case of pulmonary hypertension due to chronic pulmonary hydatid cyst embolism. Our patient was 33 year old female. She was admitted to our hospital with complaints of progressive dyspnea and fatigue. Her past medical history revealed surgical excision of hepatic hydatid cyst one year ago and pulmonary artery hydatid cyst embolectomy six month ago. On admission her pulmonary artery systolic pressure was 60 mmHg. Her CT angiography revealed bilateral lower lobe pulmonary arterial thrombus. After the oral anticoagulation and albendazole treatment, repeated echocardiographic evaluations showed PAP as 50, 45, 30 mmHg.

Keywords: hydatid cyst, embolism, pulmonary hypertension

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A 33-year old female patient was admitted to our hospital with the complaints of dyspnea and fatigue. In her past medical history, she was operated as hepatic cyst hydatid 1year ago and pulmonary artery hydatid cyst embolectomy six months ago. It was found that the cyst had ruptured into inferior vena cava and gall bladder in first operation. Albendazole treatment had been started after the operation. Our physical examination showed a blood pressure of 130/70 mmHg, heart rate of 106 bpm, temperature of 36.8°C and respiratory rate of 22/min. Respiratory system examination was normal. Cardiovascular examination revealed accentuated P2, apical 1/6° systolic murmur. Abdominal examination was normal except incision scar belonging to past operation. The result of routine laboratory tests were as follows: Haemoglobin: 12.7mg/dl, leukocyte count: 6620/mm^3, eosinophil: 1040/mm^3. Serum biochemical tests and urinary examination were normal. Serologic test results for hydatid cyst (Casoni and Weinberg) were positive. Her spirometric tests results were as follows; FVC 87%, FEV_1 85%, FEV_1/FVC:102, PEF:87%. Chest radiography showed a 2×2×3cm mass located near the right hilus (Figure 1). Thorax CT revealed a mass located on the right hilus (complicated cyst? lymphadenopathy?) and chronic infiltrative changes. Pulmonary angiography showed a filling defect in the lower lobe pulmonary artery (Figure 2) , systolic, diastolic and mean pressures were 45, 15, 25 mmHg respectively. Through a right thoracotomy, a hydatid cyst was found in the right lower lobe pulmonary artery and enucleated. After the pulmonary embolectomy, the patient was discharged. 5 months later she was admitted to our hospital with the complaints of dyspnea and palpitation again. Chest X-ray showed bilateral pleural effusion and right hilar enlargement. In rheumatologic and immunologic evaluation, rheumatoid factor and IgE levels were found to be normal. Specific Echinococcus granulosus IgG was found to be high by indirect hemagglutination (1/1024). Thrombophilic risk factors were evaluated, no risk factors found. In this admission, echocardiography was performed. Pulmonary artery systolic pressure was 55-60 mmHg, right atrium was mildly dilated, mild mitral and tricuspid regurgitations were detected. In her thoracoabdominal computerized tomography, there was a hypodense area in liver thought to be related to residual hydatid cyst which was located in close proximity with inferior vena cava (Figure 3). Starting from this residual cyst, there was an irregular shaped filling defect protruding to the beginning of right atrium (ruptured hydatid cyst or thrombus?). This image was consulted with gastroenterological surgery and it was thought to be a scar, secondary to operation. Also, bilateral lower lobe pulmonary arterial thrombi were detected. Her lower extremity venous doppler evaluation was normal. Unfractionated heparin infusion was started, later on oral anticoagulation was started, and albendazole was added to treatment. Three months later, repeated thorax CT revealed no thrombi in pulmonary arteries, and echocardiographic measurement of pulmonary arterial systolic pressure was 40 mmHg. In the next admission, she was tachypneic and cyanotic in physical examination. In her control thorax CT, multiple thrombi were found in pulmonary arterial segments. Systolic pulmonary arterial pressure was measured to be 85 mmHg by echocardiography. Control Echinococcus granulosus IgG by indirect hemagglutination

Corresponding Author: Bülent Çiftçi, Atatürk Chest Disease and Chest Surgery Hospital, Pulmonary Disease, Ankara, Turkey, Phone: +90 312 4271010, E-Mail: bciftci@superonline.com

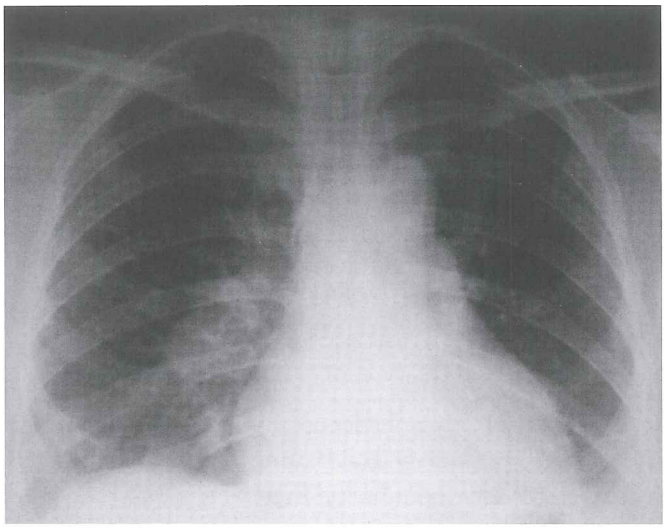

Figure 1.

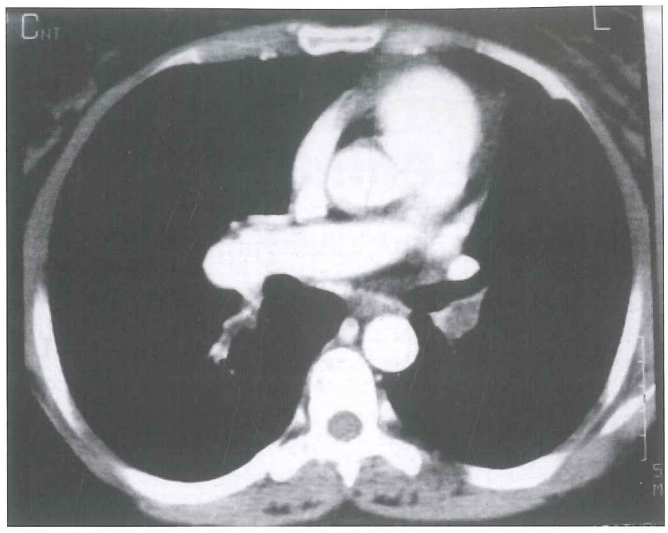

Figure 2.

was higher. (1/1280). It was detected that oral anticoagulation and albendazole treatment were not used in order. After the treatment was put in order, the patient was discharged. With 3 months interval, repeated echocardiographic evaluations showed pulmonary arterial pressures as 50, 65, 45, 30 mmHg. Repeated thorax CT showed no mass in inferior vena cava (Figure 4). She is still under the treatment of oral anticoagulation with albendazole for one year .

DISCUSSION

Echinococcosis is an infection of humans caused by the larval stage of echinococcus granulosus. It occurs most commonly in Africa, Australia, Argentina, Chile, South America, Middle East, Mediterranean and New Zealand. The hydatid tapeworm (*E. granulosus)* requires two hosts to complete its life cycle. Dogs (and other canines) are the definitive host and a variety of species of warm-blooded vertebrates (sheep, cattle, humans etc.) are the intermediate host. Humans are accidental hosts and do not play a role in the biological cycle. The adult worm inhabits the small intestine of the definitive host, is usually 2-7 mm long, is attached to the mucosa by a double row of hooklets contained in its scolex and has at least three proglottids, which contain numerous eggs. The eggs pass out in the faeces of the dog and stick to the animal's fur or to the grass. Intermediate hosts ingest eggs when grazing on contaminated ground and the embryos are released after hatching in the small intestine. Embryos penetrate the intestinal mucosa, enter the portal circulation, and are carried to the other organs, most commonly the liver (60-75%) and lungs (15-25%). It can also locates bone, brain and mediastinum (10-15%) (1, 2).

Pulmonary cysts are usually solitary in 72% of cases affecting one lobe. Hydatid cysts are usually asymptomatic. Symptomatic patients present with cough, dyspnea and chest pain. On the chest radiography, the lesions vary in diameter from 1 to 20 cm. Sometimes cyst is surrounded by an area of pneumonitis or atelectasis(1).

Complications of the hydatid cyst include pressure effects from enlarging cyst, secondary bacterial infection and cyst rupture. Three types of cyst rupture are recognized: contained, communicating and direct. Unlike contained rupture, communicating and direct ruptures are symptomatic forms. Direct rupture refers to disruption of the cyst wall and spilling of the cyst contents into cavities which it has ruptured (3,4). Our case is an example of direct rupture into inferior vena cava, and resultant pulmonary hydatosis. She had symptoms related to pulmonary hypertension most probably due to hydatid cyst embolization. Although hemoptysis is the most frequent sign of hydatid pulmonary embolism, our case did not show this symptom (5,6). In her past liver operation, hydatid cyst had been found to be ruptured into the inferior vena cava. Also, thoracoabdominal CT showed a filling defect in inferior vena cava which was most probably belonging to ruptured hydatid cyst.

Hydatid pulmonary embolism has been classified according to three types of outcome: 1) acute fatal embolism, 2) subacute embolism and pulmonary hypertension and death in less than a year, 3) chronic pulmonary hypertension (2, 7, 8). In our case, thorax CT evaluations showed repeated emboli to the pulmonary arteries, and continuously changing pulmonary arterial pressures were consistent with the embolizations. So, this form of repeated embolizations and pulmonary artery pressure is compatible with the chronic pulmonary hypertension in hydatid cyst.

In the diagnosis of pulmonary embolization, pulmonary angiography is the gold standard technique which is invasive procedure and has morbidity and mortality although rare (9). Multidetector computed tomography an-

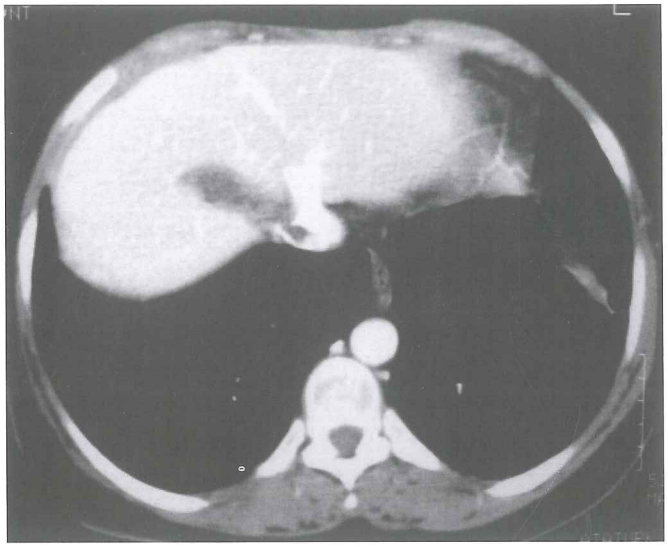

Figure 3.

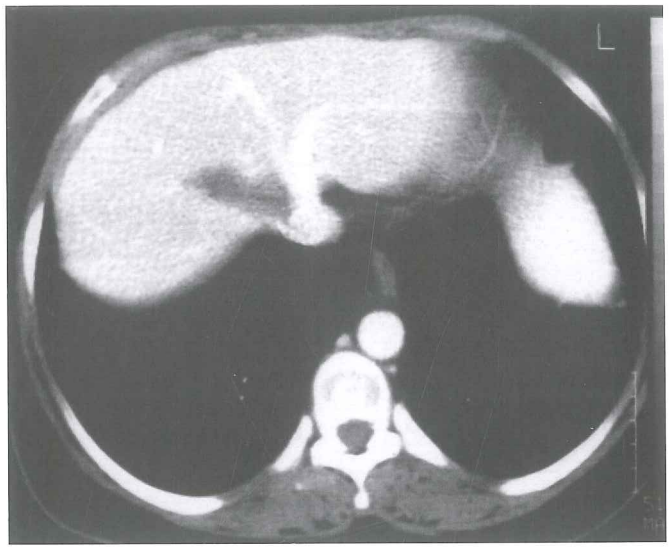

Figure 4.

giography is a noninvasive diagnostic procedure in pulmonary embolism, and also in differential diagnosis of pulmonary embolism from other cardiac, infectious, pulmonary and pleural disorders having the same symptomatology (10, 11). The diagnostic investigation should also include echocardiography, and MRI. Echocardiography is the imaging modality of choice of locating hydatid cyst in the heart and pericardium (12). We used spiral CT and angiography as diagnostic procedure, and followed our patient with spiral CT angiography for repeated embolizations and echocardiography for pulmonary artery hypertension. Routine blood tests are usually nondiagnostic. Serologic tests for hydatid cyst may be present to confirm diagnosis in patients without previous history of hydatid cyst. As in our patient seroconversion is important for following. Eosinophilia is uncommon except for cyst rupture (13).

These patients have clinical and radiological findings similar to pulmonary thromboemboli, but prognosis and treatment protocols are different in hydatid cyst emboli. The definite treatment of pulmonary embolization of hydatid cyst is surgical. The role of medical treatment is not clearly defined (14). Albendasole has shown some promise (15) We performed pulmonary embolectomy for cyst embolus, also surgery is not suitable for multiple embolism albendasole was given and for pulmonary hypertension we used anticoagulant treatment.

CONCLUSION

Hydatid cyst is still a problem in endemic areas. Pulmonary embolization of cyst may be acutely fatal or progressive pulmonary disease may occur causing pulmonary hypertension though it is rare. Our case is interesting for being a cause of chronic pulmonary hypertension resulting in serious morbidity.

REFERENCES

1. Morar R, Feldman C. Pulmonary echinococcosis. Eur Respir J 2003; 21: 1069-77.
2. Smith G, Irons S, Schelleman A. Hydatid pulmonary emboli. Australasian Radiology 2001; 45: 508-511.
3. Lewall DB, mcCorkell SJ. Rupture of echinococcal cysts: Diagnosis,classification and clinical implications. AJR 1986; 146:391-4.
4. Pedrosa I, Saiz A, Arrazola J et al. Hydatid disease.Radiologic and pathologic features and complications. Radiographics 2000; 20: 795-817.
5. Lahdhili H, Hachicha S, Ziadi M, Thameur H. Acute pulmonary embolism due to rupture of a right ventricle hydatid cyst. Eur J Cardiothorac Surg 2002;22:462-4.
6. Arıbaş OK, Kanat F, Türk-Arıbaş E et al. Embolisation of hydatid cysts in the pulmonary artery presenting with haemoptysis. Neth J Med 2007; 65(3): 109-11.
7. Glisanz V, Campo C, Cue R et al. Recurrent pulmonary embolism due to hydatid disease of heart.Br Heart J 1977;39:553-8.
8. Lioulias A, Kotoulas C, Kokotsakis J et al. Acute pulmonary embolism due to multiple hydatid cysts. Eur J Cardio-thorac Surg 2001; 20:197-99.
9. Kauczor HU, Heussell CP, Thelen M. Update on diagnostic strategies of pulmonary embolism. Eur Radiol 1999;9:262-275.
10. Tercan F, Kacar N, Kılıç D et al. Hydatid cysts of the bilateral pulmonary arteries and left ventricle wall; computed tomography and magnetic resonance imaging findings. J Comput Assist Tomogr 2005; 29(1): 31-3.
11. Odev K, Acikgözoglu S, Gormüs N et al. Pulmonary embolism due to cardiac hydatid disease: imaging findings of unusual complications of hydatid cyst. Eur Radiol 2002; 12: 627-33.
12. Desnos M, Brochet E, Cristofini P et al. Polyvisceral echinococcosis with the cardiac involvement imaged by two-dimensional echocardiography and nuclear magnetic resonance imaging. Am J Cardiol 1987; 59:383-384.
13. Kardaras F, Kardara D, Tselikos D et al. Fifteen year surveilance of echinococcal heart disease from a referral hospital in Greece. Eur Heart J 1996;17:1265-1270.
14. Bousnina S, Racil H, Maghraoui O, et al. Hydatid pulmonary embolism. Seven cases. Rev Pneumol Clin 2005; 61: 31-6.
15. Morris DL, Dykes PW, Marriner S, et al. Albendazol-objective evidence of response in human hydatid disease. JAMA 1985;253:2053-7.