

Main Pulmonary Artery Hydatidosis with Secondary Involvement of the Lungs: a Shepherd Boy's Story

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Abstract

Cardiac hydatid cyst (CHC) is a rare disease that was endemic in some regions especially in sheep-raising areas. The most commonly accepted theory for the routes of heart involvement in hydatid cyst was infestation by the hexacanthus embryo through coronary arteries.

Here, we describe a case with the primitive cardiac hydatid cyst located around the pulmonary valve (PV) and main pulmonary artery (mPA) extended to right and left pulmonary arteries and metastasized to both lungs. Could it be possible for the embryo to adhere PV and mPA directly?

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Introduction

Cardiac echinococcosis is not a common lesion, the incidence reported ranges between 0.5 to 2 % of all human echinococcosis, and most cases have been reported from endemic regions like the Mediterranean basin, Australia, South America, Africa.¹

Although many of the Cardiac echinococcosis cases are asymptomatic, chest pain, palpitation, dyspnea have been described.^{1,2} Faintness, angina, arrhythmia, pulmonary or systemic emboli were also reported, rarely.¹⁻⁸

Case report

A 20 years old male patient from central Territory, Iran, was admitted in Tehran Heart Centre due to dyspnea, hemoptysis & syncope, on April 2003.

His coughing had started a year prior to and gradually hemoptysis had developed.

On February 2003, he had a few attacks of syncope & had been visited by a cardiologist, who referred him to our clinic due to detection of mPA cystic lesions in the patient's two dimensional echocardiography (Figure 1).

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Figure 1. The first two dimensional echocardiography revealed multiple cystic lesions in main pulmonary artery

He was a shepherd in his childhood & youth and had been grown up in a rural sheep-raising area. He had been complaining of easy fatigability and some suspicious faintness for several years. But nobody believed in him. Even when he had fainted on the ground.

Quetelet Index (Body Mass Index) was 20.2, BP=100/80 mmhg, HR=110 bpm; T=37.5 Cs (Rising up to 38.2 Cs at evening); Heart auscultation revealed Systolic ejection type murmur 2/6 in upper left sternal border as the only remarkable sign. EKG revealed sinus tachycardia & Right axis deviation (RAD). (Figure 2).

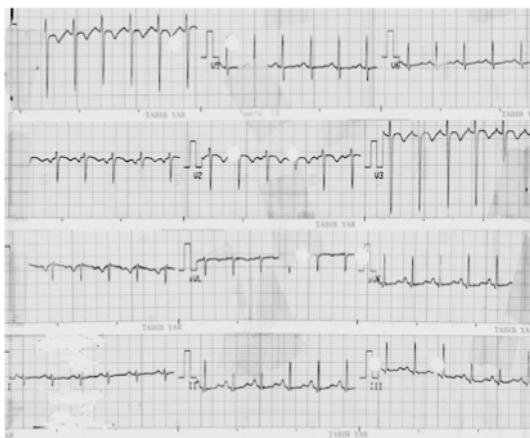


Figure 2. EKG revealed sinus tachycardia and right axis deviation

In chest X-Rays multiple round opacities in both lungs was noted, the largest one, located in the inferior segment of the left lung, had been revealed excavation (Figure 3).



Figure 3. Chest X-ray revealed opacities in the lungs

Lab exams were normal except slight microcytosis (MCV=69), ESR=30 and CRP +++. Sputum direct smears & cultures were negative for BK and any other important microorganism.

Severe chest restriction was reported in pulmonary function test. CT scan revealed multiple round lesions in both lungs which some having signal void area infavor of excavation. It also revealed that the main PA has been dilated, containing thrombosis extended to the left pulmonary artery (Lt PA). (Figures 4 & 5).

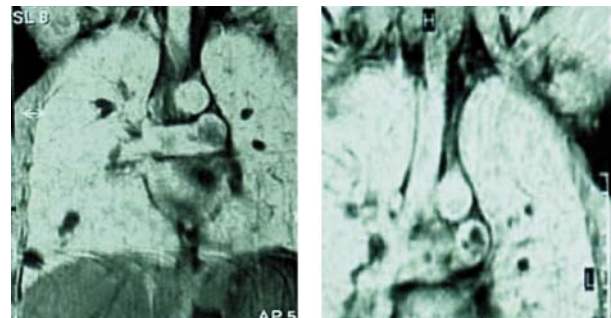


Figure 4. Thoracic CT-scan revealed main and left pulmonary artery involvement

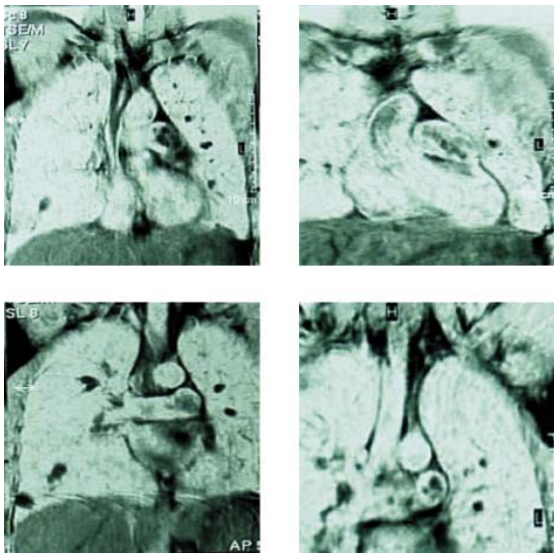


Figure 5. CT scan confirmed the presence of thromboembolic mass around pulmonary valve

Lower limb color Doppler and abdominal sonography was normal. Also, total body CT scan finding was unremarkable.

Perfusion ventilation scan revealed multiple segmental perfusion defects throughout both lungs and reported as a high probability scan for pulmonary embolism (Figure 6).

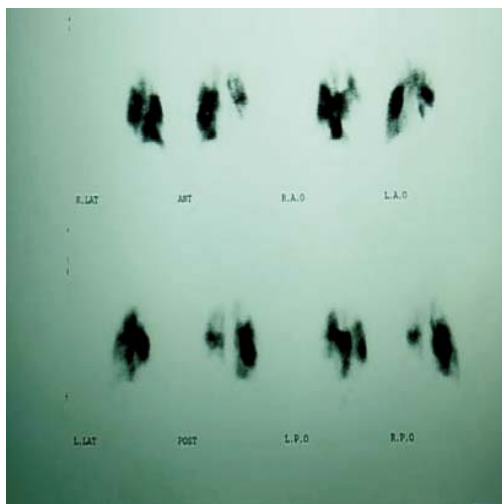


Figure 6. Perfusion scan confirmed the pulmonary embolism

Traneseophageal echocardiography (TEE) revealed mild dilation of RA & RV, mild TR and an echodens mass, extended from pulmonary valve (PV) to right pulmonary artery (Rt PA), (40 mm in size) producing a pressure gradient of 20 mmHg through PV. Normal function was described in left cardiac side (Figure 7).



Figure 7. Traneseophageal echocardiography reported an echodens mass, extended from pulmonary valve (PV) to right pulmonary artery (RT PA), (40 mm in size)

The serum Echinococcus titer measured By Enzyme-linked immunosorbent assay (ELISA) was highly positive (77: normal range <20).

Therefore, with the diagnosis of cardiopulmonary hydatidosis, the patient was transferred to the cardiovascular surgery department and antiscolicidal agent was started (Albendazol, 400mg/BD).

At operation, after primary procedures (including cardiopulmonary bypass and cold cardioplegia) the massive mass of cystic lesions and thrombosis in main, RT and Lt PA were found which were evacuated completely.

A few small cysts were also found in RV ostium and RV free wall, which were dissected completely. Histopathological analysis confirmed hydatid cysts.

Now (February, 2004) the patient is well, and two dimensional follow-up echocardiography revealed that the operation was great (Figure 8).



Figure 8. Two dimensional follow-up echocardiography revealed that the operation was great



Also serial chest X-ray demonstrates that the pulmonary lesions were diminished in size by now.

In our story, the shepherd boy was right and at last, somebody believed in him before the wolf breaks his heart, completely.

Discussion

Although liver and lungs are the primary sites of hydatid cyst in 85- 90% of cases, it should be insisted that the cyst may involve any other organ in the body (including bone, brain, heart etc).² Due to the work of Dévé,^{2,10} it is now commonly accepted that the main route of cardiac infestation with hexacanthus embryo of *Echinococcus Granulosa* was through the coronary Arteries.

Thus it is primitive and often unique lying in myocardial layers. Sometimes, after rupture of primitive hydatid cysts, the secondary cardiac lesions are developed. These secondary cysts can progress to the myocardium, where they tend to be numerous.¹¹

Cardiac hydatidosis through coronary arteries often involves left ventricular freewall. However, questions persist regarding the predominance of left ventricular (LV) locations of CHCs. Several hypotheses have been offered: (1) dominance of the left coronary artery, (2) the more important myocardial mass in the LV offers better condition of development for parasite; and (3) different pressure regimens. All these factors seem to play roles, but none is likely to be a determining one.^{2,11} this report revealed a strange pattern of larva dissemination throughout heart & pulmonary arteries that could be known as a reverse dissemination of hydatid cyst (heart to lungs).

As reported, involvement started from a strange location, PV & mPA, extended throughout PV and developed pulmonary artery mass, then ruptured and made a mess of thrombosis and cystic lesions in this case. Then after daughter cysts were embolized through pulmonary arteries, lungs getinvolved. Therefore, there is a point. How could it be possible? & how could it be explained by Deve's theories? Moreover, could the embryo be transmitted by blood stream to adhere endothelium of the PV and mPA directly and resist against blood flow?

Although in this case we do not have any clues to case histories and could not reject the possibility of PV congenital malformations; we think that, the routes of hytatid cyst dissemination must be re-evaluated.

In this case although two dimensional echocardiography was the golden tool for detection and follow up, severity of dissemination was demonstrated by TEE and CT scan, concomitantly. Each one of them revealed some aspects of patient's cardiopulmonary involvement. TEE revealed right PA and CT scan reported left PA involvement and neither one could demonstrate right ventricular free wall involvement (maybe due to small size).

Although there is some controversy regarding CHCs`

treatment,¹² we believe, in our experience, that the surgical approach concomitant with the medical treatment is the best one.

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