

The Evaluation of 13 Patients with Intrathoracic Extrapulmonary Hydatidosis

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Cases of intrathoracic extrapulmonary hydatid cysts are very rare. We identified 13 patients with intrathoracic extrapulmonary hydatid cysts in our clinic over 12 years. Four patients had extrapulmonary cysts only; nine patients had both intrapulmonary and extrapulmonary cysts. Cysts were identified in the pleural space, extrapleural region, diaphragm and chest wall. Thoracotomy was used in all patients, and extrapulmonary lesions were removed by cyst extirpation from surrounding tissue or by pericystectomy.

In one patient with chest wall involvement, partial rib resections were performed because of rib destruction. In two patients with liver cysts passing through the diaphragm to the thorax, the diaphragm was cut, cysts on the liver roof were removed and then the diaphragm was repaired. There was no mortality, morbidity, or disease recurrence during the post-operative period in any of the 13 patients. We conclude that these rare cases give a new insight into hydatid cyst pathophysiology.

KEY WORDS: HYDATID DISEASE; INTRATHORACIC EXTRAPULMONARY HYDATID CYST; THORACIC HYDATIDOSIS

Introduction

Hydatid disease, which is caused by *Echinococcus granulosus* or *Echinococcus multilocularis* tapeworms, is also known as echinococcosis or hydatidosis.¹ Hydatid disease is the most severe helminthic zoonosis, which has important medical, social and economic effects in Turkey.

Hydatid disease has been described since the times of Galen and Hippocrates. Although liver and lung are the most common sites of the hydatidosis, it can also be found elsewhere in the body. The extrapulmonary location of the disease in

the thorax is very rare, and surgeons might need to employ procedures that are different from those used for pulmonary cysts.²

The aim of this study was to evaluate all patients with thoracic hydatid disease treated in our clinic, to determine how many had intrathoracic extrapulmonary hydatid cysts.

Patients and methods

PATIENT POPULATION

We operated on several patients with thoracic hydatid disease in our department between January 1990 and December 2001, and retrospectively evaluated how many of these patients had intrathoracic

extrapulmonary hydatid cysts. We recorded the age, sex, medical history and the signs and symptoms of disease for each patient. We also performed an indirect haemagglutination test and a radiological examination for each patient.

Ethical approval for the study was not required as all patients were being treated according to routine hospital protocols and no new treatments were being tested. All patients gave informed consent to the surgery.

TREATMENT OF PATIENTS WITH EXTRAPULMONARY HYDATID CYSTS

Thoracotomy under general anaesthesia was used for the evacuation of cysts in all patients. Extrapulmonary lesions were removed by cyst extirpation from surrounding tissue or by pericystectomy. For each patient, one thoracic drain was inserted and the thoracotomy incision was closed without using a patch. Indirect haemagglutination tests were performed in selected patients. The anti-helminthic agent albendazole was given post-operatively to patients treated after March 1996; a regimen of 800 mg/day for three 21-day courses, with 10-day rest periods in between each course of treatment, was used.

Results

In total, 207 patients with thoracic hydatid disease were operated on in our department between January 1990 and December 2001. Intrathoracic extrapulmonary hydatid cysts were found in 13 (6.2%) patients - 12 were male and one was female (Table 1). Patients ranged in age from 20 to 27 years (mean: 21.2 years). Four patients did not have any pulmonary cysts. The most common presenting symptoms were cough and chest pain. We performed an indirect haemagglutination test in 10 of the patients with extrapulmonary hydatid cysts, and findings were positive in seven cases (70%). The 13

patients with intrathoracic extrapulmonary hydatid cysts had intact or ruptured hydatid cysts in the chest wall, diaphragm or pleural space.

A left posterolateral thoracotomy was performed in patient 1, and 43 intact hydatid cysts and 21 ruptured hydatid cysts were found in turbid pleural fluid. We removed the pleural fluid and all of the cysts and performed visceral pleural decortication. No intraparenchymal cysts were found in this patient, there were no complications in the post-operative period and albendazole therapy was given as per the regimen described above.

A left anterolateral thoracotomy was performed on patient 2. In thoracotomy, intrathoracic extrapulmonary multiple cystic lesions with many daughter vesicles were seen, especially in the anterior and lateral regions. The hydatid cysts and daughter vesicles were localized in the upper portion of the intrathoracic space between the endothoracic fascia and parietal pleura, and they had pushed the lung and parietal pleura towards the inferior. The multiple cystic lesions and daughter vesicles were removed, and 13 multilocular intact hydatid cysts and multiple ruptured cysts that had invaded the ribs and muscles in the chest wall were examined. Intact hydatid cysts extended into the cervical region throughout the pectoralis major muscle and into the back to the inferior of the scapula, so they were extending into the hemithorax passing the ribs. Ruptured cystic lesions had caused destruction of the second, third and fourth ribs, and damage to some areas of the latissimus dorsi muscle, serratus anterior muscle and intercostal muscles. Cysts inside the hemithorax were located in the extrapleural region, however, and there were no pulmonary parenchymal lesions. All cystic structures in the chest wall and in the cervical region were removed, and partial rib

Patients with intrathoracic extrapulmonary hydatid cysts who were operated on for

TABLE 1:
Clinical data for 13 patients (12 male, one female) with intrathoracic extrapulmonary hydatid cysts who were operated on for thoracic hydatid disease between January 1990 and December 2001

Patient number	Sex and age (y)	Medical history	Signs and symptoms	Radiological examination	Indirect haemagglutination
1	Male, 20	An operation for a cyst in the liver 13 years ago	Cough, dyspnoea and pain in the left hemithorax	Multiple cystic lesions and pleural fluid in the lower half of the left hemithorax	+
2	Male, 20	An operation for a cyst in the cerebral frontal lobe 8 years ago	Cough, chest pain and masses on the neck and left side of the chest	Multiple cystic lesions in the cervical region, upper one-third and middle regions of the left hemithorax. Destruction of the second, third and fourth ribs	+
3	Male, 20	-	Chest pain	Multiple extrapleural cysts in the right hemithorax and a solitary hepatic cyst	+
4	Male, 21	An operation for a cyst in the liver 4 years ago	Dyspnoea	Solitary cyst in the left chest wall	-
5	Male, 21	-	-	One cystic lesion in the lung and a cystic lesion in the liver on the right side	+
6	Male, 20	-	Cough	Two cystic lesions in the lung and one cyst above the diaphragm on the right side	-

TABLE 1 (continued):
Clinical data for 13 patients (12 male, 1 female) with intrathoracic extrapulmonary hydatid cysts who were operated on for thoracic hydatid disease between January 1990 and December 2001

Patient number	Sex and age (y)	Medical history	Signs and symptoms	Radiological examination	Indirect haemagglutination
7	Male, 21	-	Cough, chest pain	One cystic lesion in the lung and one cyst above the diaphragm on the right side	
8	Female, 27	-	Cough, dyspnoea	Cystic lesions in the lung and pleural space, on the left side	+
9	Male, 21	-	Cough, expectoration	Cystic lesions in the lung and pleural space, on the left side	
10	Male, 22	-	Cough, dyspnoea	Cystic lesions in the lung and pleural space, on the left side	+
11	Male, 21	-	Chest pain, cough	Cystic lesions in the lung and pleural space, on the left side	+
12	Male, 20	-	-	Cystic lesions in the lung and pleural space, on the right side	-
13	Male, 21	-	Cough, expectoration	Cystic lesions in the lung and pleural space, on the right side	

resections were performed on the three affected ribs. Damaged areas of the muscles were debrided. Albendazole therapy was given in the post-operative period.

Standard posterolateral thoracotomy was performed in patient 3. Pleural adhesions, intrapleural cysts or pulmonary parenchymal cysts were not observed during the surgical operation. Multiple cysts were located just underneath the parietal pleura in a chain-like fashion, in the line of internal mammarian vessels which ascended towards the apex, throughout the paravertebral area. The diaphragm had two lobular enlargements with a large mass under them. We removed the smaller of the two cysts on the diaphragm by enucleation and performed cystotomy on the larger cyst on the diaphragm. In doing so we reached a large liver cyst, approximately 8 cm in diameter in the shape of a dumbbell. We cut the diaphragm and removed more than 340 daughter vesicles from the liver cyst, then performed capitonage on this large liver cavity and closed the diaphragm in a standard manner. Twelve extrapleural intrathoracic cysts between the parietal pleura and endothoracic fascia were removed by enucleation without causing any ruptures. Albendazole therapy was given post-operatively.

Patient 4 had one secondary cyst in the left chest wall without any intrapulmonary cysts. Mini lateral thoracotomy was performed and this cystic lesion was removed by cyst extirpation from the surrounding tissue.

A further three patients (numbers 5 - 7) had a total of four pulmonary cysts and three diaphragmatic cysts. One of the diaphragmatic cysts had developed due to penetration of the diaphragm by a liver dome cyst. The two other cysts were above the diaphragm and did not involve the liver as they were secondary to ruptured pulmonary cysts. Posterolateral thoracotomy was performed and a hydatid cyst was

removed in each patient. In a patient with a liver cyst that passed through the diaphragm to the thorax, the diaphragm was cut, the cysts on the liver roof were removed and the diaphragm was repaired. Albendazole therapy was given in one patient.

In six patients (numbers 8 - 13), there were ruptured intrapulmonary cysts and a total of seven pleural cysts. Secondary cysts had probably developed as a result of intrapulmonary cyst rupture into the pleura. Posterolateral thoracotomy was performed and then pulmonary and extrapulmonary hydatid cysts were removed. Albendazole therapy was given in three patients with pleural cysts.

There were no major complications, mortality or morbidity in any of the 13 patients during the post-operative period. We followed the patients for a mean of 15.2 months (range: 6 - 19 months). We did not observe recurrence of hydatidosis in any of the patients.

Discussion

Extrapulmonary, intrathoracic hydatid cysts are very rare and are found in chest wall, mediastinal, pericardial, myocardial, lobar fissure and pleural locations. To date, pleural hydatid cysts were reported to be the most common forms of extrapulmonary intrathoracic cysts.² Pleural hydatid cysts are always secondary to the rupture of hydatid cysts located in an organ adjacent to the pleura.³ In 1988, Vivó *et al.*³ reviewed 1841 patients who had been operated on since 1947 for thoracic hydatidosis, including 77 patients who had pleural complications. In our study, seven of the 13 cases had pleural cysts. We suggest that these hydatid cysts were secondary to the rupture of pulmonary or hepatic hydatid cysts. There were diaphragmatic cysts in four patients. We think that these diaphragmatic hydatid cysts were secondary to the rupture of pulmonary

hydatid cysts in two patients and hepatic cysts in the other two patients.

In a study published in 1988, only one chest wall hydatidosis was reported among 842 patients with thoracic hydatidosis who had been treated over a 30-year period.⁴ Ozdemir *et al.*⁵ reported that thoracic wall involvement was seen in six patients (0.09%) in a series of 6500 patients who underwent 7995 operations for hydatid disease in their department. Jlidi *et al.*⁶ reported that the incidence of chest wall hydatid cysts was about 0.9 – 2%. We found chest wall hydatid cysts in two patients who had a history of having had an operation for hydatid cysts 4 and 13 years previously. In patient 2, there was destruction of the ribs and muscle damage. The gold standard for therapy is radical removal of the cysts or the chest wall that is involved. We removed intact and ruptured cysts and employed partial rib resections and muscle debridement in this patient.

We found extrapleural hydatid cysts between the parietal pleura and endothoracic fascia in our third patient. We have hypothesized a novel dissemination theory in this patient, which proposes that the liver dome cyst had opened into the diaphragm and caused two diaphragmatic cysts. The scoleces were probably transported by the diaphragmatic lymphatics from the liver and then by parasternal lymphatics and intercostal lymphatics to the chest in this patient.⁷

Surgery is the treatment of choice for patients

with intrathoracic hydatid disease and thoracotomy was used in all patients. Radical procedures, such as pneumonectomy, lobectomy and segmentectomy, were not used in any patient with a intrapulmonary hydatid cyst. Extrapulmonary lesions were removed by cyst extirpation from the surrounding tissue or by pericystectomy. In one patient with chest wall involvement, partial rib resection was done because of rib destruction. In two patients with liver cysts passing through the diaphragm to the thorax, the diaphragm was cut, the cysts removed from the liver roof and the diaphragm repaired.

Medical therapy with albendazole has been proposed as adjunct therapy to surgical procedures and as a primary medical therapy in patients who cannot have complete cyst removal by surgery. We used post-operative albendazole treatment in seven patients who had multiple intrathoracic cysts. In our patients, there was no mortality, morbidity, or recurrence of hydatid disease during the post-operative period.

We conclude that because cases of intrathoracic extrapulmonary hydatid cysts are very rare, the discussion of our cases will give a new insight into hydatid cyst pathophysiology.

Conflicts of interest

No conflicts of interest were declared in relation to this article.

• Received for publication 15 October 2004 • Accepted subject to revision 2 November 2004

• Revised accepted 15 December 2004

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