Diagnostic Dilemma of Hydatic Disease: Analysis of 16 Patients

Hidatik Hastalıkta Tanısal İkilem: 16 Hastanın Analizi

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ABSTRACT Objective: Hydatic cyst disease may cause atypical clinical and radiological features and complications that cause difficulties in diagnosis. Misdiagnoses may end up with delay in treatment. We aimed to examine the characteristics and surgical results of hydatic cyst disease with atypical clinical features. **Material and Methods:** Hydatic cyst patients with atypical clinical and radiological features diagnosed in the department of chest surgery, between 2003 and 2007 were retrospectively evaluated according to sex, age, symptoms, radiological diagnosis, surgical procedures applied and postoperative complications. Sixteen patients with atypical clinical features were analyzed for misdiagnoses and challenges in the diagnosis and results of treatment. **Results:** The study group consisted 7 males and 9 females with a mean age of 37.25 ± 15.69 (16-67) years. Thoracic empyema (7 patients), lung cancer (4 patients), mediastinal tumor (1 patient), tuberculous pleurisy (1 patient), chest wall tumor (2 patients) and bulleous lung (1 patient) disease were the misdiagnosed diseases in hydatic cyst disease. All patients were treated surgically. Hydatic disease was confirmed pathologically. There was no mortality among patients. The mean follow-up period was 21 months and no relapse was observed. **Conclusion:** Hydatic disease may imitate other chest diseases when it causes atypical clinical features. In order to reduce the risks of recurrence, which may need a second operation, the disease should be considered especially in endemic regions. If exact diagnosis is not known, surgical intervention should not be avoided.

Key Words: Echinococcosis, pulmonary; cystectomy; thoracotomy; postoperative complications

ÖZET Amaç: Kist hidatik hastalığı tanıda zorluklara neden olan atipik klinik ve radyolojik görünümlere ve komplikasyonlara yol açabilmektedir. Tanısal hatalar tedavide gecikmeye yol açmaktadır. Biz atipik klinik görünümü olan hidatik kistli hastalarda karakteristik özellikleri ve cerrahi sonuçlarını göstermeyi amaçladık. **Gereç ve Yöntemler:** 2003-2007 yılları arasında göğüs cerrahisi kliniğinde teşhis konulan atipik klinik ve radyolojik görünümlü kist hidatik hastaları cinsiyet, yaş, semptomlar, radyolojik teşhis, uygulanan cerrahi girişimler ve postoperatif komplikasyonlara göre retrospektif olarak değerlendirilmiştir. Hastaların atipik klinik görünümlü 16 hasta, tanı sırasında yaşanan hatalar, zorluklar ve tedavi sonuçları açısından analiz edilmiştir. **Bulgular:** Çalışmaya, yaş ortalaması 37.25 ± 15.69 (16-67) yıl olan 7 erkek ve 9 kadın hasta alındı. Kist hidatik hastalığında hatalı tanılar; torasik ampiyem (7 hasta), akciğer kanseri (4 hasta), mediastinal tümör (1 hasta), tüberkülöz plörezi (1 hasta), göğüs duvarı tümörü (2 hasta) ve büllöz akciğer (1 hasta) idi. Tüm hastalar cerrahi olarak tedavi edildi. Hidatik hastalık patolojik olarak doğrulandı. Hastalarda hiç ölüm görülmedi. Ortalama takip süresi 21 ay idi ve hiç nüks olmadı. **Sonuç:** Hidatik hastalık, atipik klinik görünüme neden olduğunda diğer akciğer hastalıklarını taklit edebilmektedir. Başka bir operasyonu gerektirecek nüks riskini azaltabilmek için, hastalık özellikle endemik bölgelerde göz önünde tutulmalıdır. Kesin teşhis belli değil ise, cerrahi girişimden kaçınılmamalıdır.

Anahtar Kelimeler: Ekinokokkozis, pulmoner; kistektomi; torakotomi; postoperatif komplikasyonlar

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Hydatic cyst is a parasitic disease, which is common in agricultural regions and underdeveloped communities where people ignore environmental and medical health precautions. In Turkey, the incidence of hydatic cyst is 14/100.000 and the prevalence is 87-400/100.000.[1-4]

In the literature, hydatic cyst as a benign disease has unexpectedly high morbidity and mortality rates due to previously mentioned severe complications. Localization and size of the lesion determine the clinical approach. Unfamiliar localization may end up with different radiological appearance and clinical outcome. To identify the atypical cases, it is essential to be aware of the characteristics and true or false positiveness of the diagnostic modalities.[5-8] The main principle in surgery is to perform cystectomy and obliteration of residual cavity while saving maximum functional lung tissue.[9]

We aimed to identify the clinical and radiological characteristics, misdiagnoses and surgical results of hydatic cysts disease with atypical clinical features.

MATERIAL AND METHODS

Patients with a diagnosis of hydatic cyst disease with atypical clinical and radiological features between 2003 and 2007 were retrospectively evaluated according to sex, age, symptoms, radiological diagnosis, surgical procedures applied and postoperative complications. Sixteen patients with atypical clinical features of hydatic cyst disease were included in the study.

Chest radiography, ultrasonography (USG), computerized tomography (CT) and magnetic resonance imaging (MRI) were used in the radiological diagnosis. Specific serologic tests and eosinophil counts were assessed in some patients. Clinical and radiological misdiagnoses were recorded.

Patients were postoperatively followed-up monthly by chest radiography and biochemical blood tests including liver enzymes, within the first three months and then the follow-up continued every three months until the end of postoperative year one by chest radiography. They were reassessed for complications and recurrences.

RESULTS

The mean age of 16 patients was 37.25 ± 15.69 (16-67) years. There were 7 male and 9 female patients. Symptoms were chest pain (n=8, 50%), cough (n= 8, 50%), dyspnea (n= 6, 38%), fever (n= 5, 31%), night sweat (n= 1, 6%) and diaphragmatic paralysis (n= 1, 6%).

Initial radiological assessment supported the diagnosis of lung cancer and transthoracic fine needle aspiration revealed benign cytology in four patients. Thoracic empyema was misdiagnosed mostly (Table 1). Non-specific diagnostic interventions, such as thorax MRI (12.5%), transthoracic fine needle biopsy (25%), bronchoscopy (18.8%) were used in the diagnostic process**.** Thoracentesis was performed to achieve diagnosis in 8 patients with a presumptive diagnosis of empyema/pleural effusion and in 4 patients with a presumptive diagnosis of lung cancer. No intraoperative complication developed in patients who had undergone transthoracic fine needle aspiration and thoracentesis.

Diagnostic thoracotomy was performed in all patients to determine the correct etiology and treatment. Care was taken to save as much lung parenchyma as possible in patients undergoing resection.

Surgical procedures performed were cystotomy and capitonage in 8 (50%) patients, wedge resection in 4 (25%) and cystectomy in 4 (25%) patients (Table 2). Wedge resection was preferred in patients 9, 10, 11 and 16 with peripheral localization (Figure 1).

TABLE 1: Misdiagnosed diseases in the patients.

Disease	Patients (number)	%
Thoracic empyema	7	43.8
Lung cancer	4	25
Mediastinal mass	1	6.2
Bullous lung disease	1	6.2
Tuberculous pleurisy	1	6.2
Undetermined disease	2	12.6
Total	16	100

TABLE 2: General parameters of the patients.

Patient number	Sex, age	Radiologic evaluation					Preoperative diagnosis	Surgical procedure
		Chest X-Ray	Thorax CT	Thorax USG	Abdomen USG	Thorax MRI		
1	M, 35	Hydropneumothorax on the right	Pulmonary collapse and hydropneumothorax	-	Two cystic lesions of the liver	-	Thoracic empyema	Cystotomy + capitonage Decortication
2	F, 20	Loculated empyema on the right	Pleural thickness + pleural fluid	-	NPF	-	Thoracic empyema	Cystotomy + capitonage Decortication
3	M, 35	Diaphragmatic elevation on the left	Mediastinal mass and diaphragmatic hernia	-	NPF	Mediastinal mass and diaphragmatic paralysis	Mediastinal tumor, diaphragmatic hernia	Cystectomy + diaphragmatic plication
4	F, 67	Hydropneumothorax on the right	Thoracic invasion of the liver cyst	-	Cystic lesion of the liver	-	Thoracic empyema	Cystotomy + capitonage Phrenoraphy + decortication + middle lobectomy
5	F, 16	Homogeneous density on the right	Massive pleurisy	-	Cystic lesion of the liver	-	Tuberculosis empyema	Cystotomy + capitonage + Decortication
6	F, 16	Homogeneous density on the right	Massive pleurisy	-	Two cystic lesions of the liver	-	Thoracic empyema	Cystectomy + Decortication + Phrenotomy + phrenoraphy
7	F, 19	Hydropneumothorax on the right	Massive pleurisy	-	-	-	Thoracic empyema	Cystotomy + capitonage Decortication
8	F, 40	Homogeneous density on the right	Cystic lesion and pleurisy on the right	Loculated empyema on the right	-	-	Thoracic empyema	Cystotomy + capitonage Decortication
9	M, 41	Nodular density at the left upper lobe	Nodule on left upper lobe (2 x 1 cm)	-	NPF	-	Lung cancer	Wedge resection
10	M, 62	Nodular density at the left lower lobe	Nodule on left lower lobe (2.5 x 1 cm)	-	NPF	-	Lung cancer	Wedge resection
11	F, 39	Nodular density at the right upper lobe	Nodule on right upper lobe (2 x 1 cm)	-	NPF	-	Lung cancer	Wedge resection
12	M, 40	Non homogenous density of between 3 and 5 costae on the right	Costal destruction and two cystic lesions on the chest wall	-	-	-	Chest wall tumor	Cystotomy + 4th and 5th costae resection + phyrenoraphy
13	M, 58	Multipl hidroaeric level and bullaeous lesion	Hydropneumothorax and septation	-	Two cystic lesions of the liver	-	Bullous lung disease	Cystotomy + capitonage
14	F, 46	Non homogeneous density on the left	Costal destruction and cystic lesions between the intercostal space	Semisolide lesion of the chest wall	NPF	Mass of the intercostal space	Chest wall tumor	Costal resection and reconstruction
15	F, 35	Homogeneous density on the right	Massive pleurisy	Massive pleural fluid	NPF	-	Thoracic empyema	Cystotomy + capitonage Decortication
16	M, 27	Nodular density at the left upper lobe	Nodule on left upper lobe (2 x 2 cm)	-	NPF	-	Lung cancer	Wedge resection

M: Male, F: Female, NPF: No pathologic finding, CT: Computed tomography, USG: Ultrasonography , MRI: Magnetic resonance imaging.

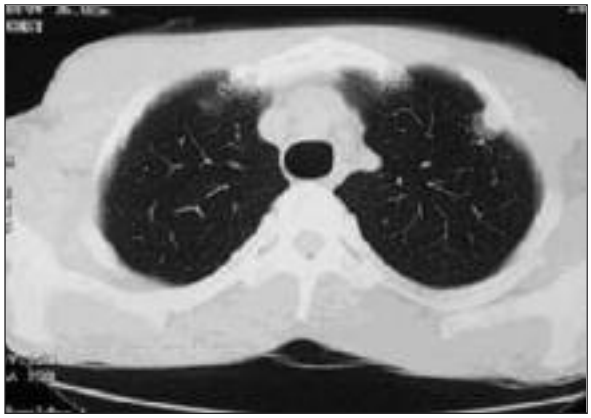

FIGURE 1: Hydatic cyst mimics peripheral tumoral lesion.

The disease was complicated in patient 4 and 6 with disease originating from the abdomen and invading the thorax by diaphragmatic destruction. Decortication, middle lobectomy and primary diaphragmatic repair was performed in patient 4. Patient 6 was treated with decortication and primary diaphragmatic repair. Costal resection was performed in patient 14, who had solid images of membrane in radiological examination without any sign of cystic structure. MRI failed to confirm chest wall tumor and she underwent diagnostic thoracotomy. Diaphragmatic resection and decortication was added to costal resection in patient 12, who was had *Echinococcus alveolaris* (Figure 2). In this patient, thoracotomy was required because of complex clinical feature with tumor like lesions near the chest wall and diaphragm with lobulated components. Plication of the diaphragm was necessary after cystectomy in patient 3 mimicking mediastinal tumor or diaphragmatic hernia (Figure 3). In patient 13, chest radiography revealed bullous lung disease and pneumothorax. Urgent thoracotomy was performed because of serious dyspnea. Perforated hydatic cyst with an intraparenchymal localization was detected during thoracotomy.

Patient 5 had a diagnosis of tuberculosis pleurisy without microbiological confirmation. Patient had no clinical or radiological response after tuberculosis treatment for six months. We performed a diagnostic thoracotomy with the suspicion of hydatic cyst.

Pleural liquid analyses, performed in three patients with pleurisy, revealed *Pseudomonas aeruginosa* infection in patient 8, and *Staphylococcus aureus* infection in patients 6 and 15. Postoperative complication was detected in two patients. Bile-leakage was detected at the abdominal drain during the postoperative period of patient 4 and it was cured by medical treatment. Postoperative wound infection in patient 12 was treated with daily local dressing**.** Histopathological examinations revealed hydatic cyst in all patients. There were no recurrences and major complications in the follow up period of 21 months. There was no mortality either.

Surgery not only provided definitive diagnosis but also treated the disease. Except in 4 patients

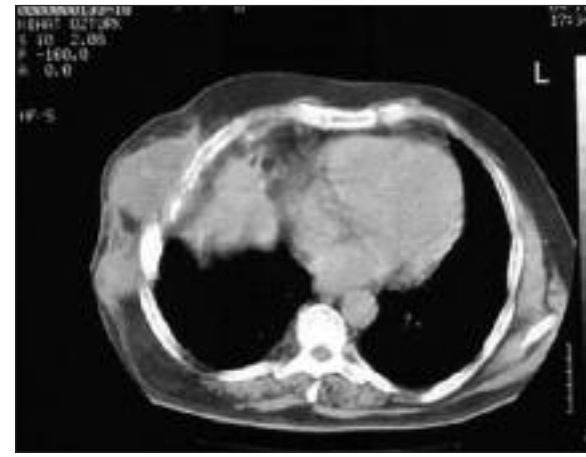

FIGURE 2: Hydatic disease located at the chest wall, mimicking chest wall tumor.

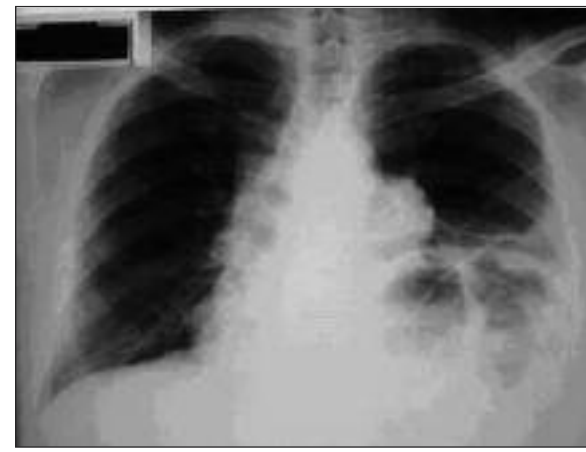

FIGURE 3: Diaphragmatic elevation mimicking a diaphragmatic hernia and mediastinal mass in postero-anterior chest radiography.

treated with wedge resection, albendazole (10 mg/kg/day) was used for all patients for at least 2 months in order to prevent probable recurrence. Age, sex, symptoms, radiological diagnosis, operation and postoperative complications of the patients were summarized in Table 2.

DISCUSSION

Although the prevalence of hydatic disease has been low in developed countries, an increase in the frequency can be expected in this part of the world recently with the increase in mobility and migration of people.[10] The countries considered to be highly infested include Uruguay, Argentina, Chile, Algeria, Tunusia, Australia and New Zealand.[11] It is also still a serious health problem in Turkey, especially in the regions of raising livestock.[1]

Although the most common site of involvement is the liver and the lungs, brain, heart, kidney, spleen, uterus, fallopian tubes, mesentery, diaphragm and muscles may also be affected.[12] Intrathoracic but extrapulmonary localizations are infrequent with a prevalence of 7.4%.[5,13] Uncommon localizations may cause rather unusual symptomatology. Serologic tests are often useful, but their potential to give false negative and positive results has limited their value in the diagnosis. Eosinophilia is another typical finding of the disease occurring in 20-30% of patients but since it may be present in many other pathologic conditions, the value of this test as a diagnostic tool is small.[10] Thus, these tests were not used for the definitive diagnosis of our patients. Radiological evaluation gains significance in such a complicated case. Surgery, supported by a sufficient radiological evaluation, provides an effective and accurate solution.

The radiographic pleural manifestation in the acute stage of rupture of the cyst varies from loculated hydropneumothorax to nonloculated partial, complete or tension hydropneumothorax as in our patient.[13] Computed tomography greatly added to the diagnosis of hydatic disease of the lung, particularly to the early discovery of coexistent small cysts and of pending or existing rupture of the cyst.[10] Computed tomography scans and the less costly USG are valuable to confirm the diagnosis of a cystic lesion. If the diagnosis of hydatic cyst is suspected, needle aspiration is contraindicated because of the risk of pleural contamination[14,15] like our approach to avoid thoracentesis or needle biopsy especially when hydatic cyst is suspected. However, we performed thoracentesis in 8 patients with a presumptive diagnosis of empyema/pleural effusion and in 4 patients with a presumptive diagnosis of lung cancer.

According to the localization, echocardiography and especially MRI scanning methods may be of value.[16,17] Intact or simple hydatic cysts of the lung produce no characteristic symptoms. Their clinical manifestation depends on the site and size of the cyst. Small, peripherally located cysts are usually asymptomatic, whereas large cysts may manifest with symptoms of compression to adjacent organs. When the hydatic cyst ruptures into the pleural space or bronchus, the symptoms are usually severe and complex. They consist of vigorous or dry coughing, expectoration of large amount of sputum, frothy blood, chest pain, moderate dyspnea, generalized malaise and fever. The symptoms of the intrapleural rupture of the hydatic cyst are accompanied by the physical findings of localized or generalized hydropneumothorax as is the case in seven of our patients.[15] The initial diagnosis of these seven patients was thoracic empyema. Treatment for thoracic empyema was initiated. Radiological features suggested hydatic cyst and patients underwent thoracic surgery. One patient who was diagnosed with tuberculosis pleurisy without microbiological confirmation was administered antituberculous treatment by a chest disease specialist for six months. The suspicion of hydatic cyst radiologically and no response to tuberculosis treatment directed us to perform diagnostic thoracotomy.

Differential diagnosis is essential since the disease can mimic other benign thoracic diseases such as abscess, empyema or malignancy. Segmental or wedge resection is usually required in these patients. The large bronchopleural fistula (BPF) that results from the rupture of hydatic cyst to the pleura contributes to the evolution of an empyema, if

the correct treatment is delayed[15] as in patient 4 in our study. This patient had been managed with closed chest tube drainage formerly. After this stage, exploratory thoracotomy was performed and upon the detection of a large BPF during the operation, middle lobectomy was added to the operative procedure due to diffuse destruction of the lobe and chronic fistula.

The chest wall localization is a rare one, but may be seen especially in endemic regions like Turkey. Of 2 patients with chest wall localization, one had single chest wall localization and the other diaphragmatic localization. Surgical interventions performed required atypical resections and reconstructions rather than conventional methods.

An uncommon presentation is the hydatidothorax. This results from massive leakage of hydatic fluid into the pleura. It is necessary to clean the pleural cavity with scolicidal solution.[18] All five patients with hydatidothorax in our study had free germinative membrane in the thoracic cavity. It was a sign of intrapleural perforation. Pleural cavities were cleaned with scolocidal solutions. Pulmonary cyst that had ruptured to the pleural cavity were removed.

Another rare feature of hydatic disease is thoracic extension of a liver cyst via transdiaphragmatic invasion. Toole, et al. reported an incidence of only 1 percent in 1250 liver cysts.[18] Cystic lesion that invades the diaphragm may expand to the thoracic cavity and affect the pulmonary parenchyma, which causes an intense inflammatory reaction and a sinus track that establishes a communication between the biliary and bronchial systems like in our patient 4. In most patients, it is possible to treat this complication with thoracotomy. The associated surgical procedures depend on the intraoperative findings. Pulmonary decortication and closing the small BPF are usually sufficient. Sometimes a minor resection, and rarely a lobectomy are necessary. Chemotherapy can be added to the pre- or post-operative process to prevent the disease. The primary aim is to reduce recurrence (still reported 10%), morbidity and mortality.[19] There was no recurrence in any patient in our series. Complications may delay the diagnostic process and are associated with the risk of recurrence and morbidity. Although surgery ensures complete treatment, medical treatment will help to prevent the recurrences.

As a result, hydatic cyst disease may imitate other diseases of the thorax. Misdiagnosis because of atypical clinic and radiological appearance in thoracic hydatic cyst disease may cause a delay in surgical treatment. Early surgical intervention is the major treatment. In order to reduce the risks of recurrence, which may need another operation, the disease should be taken into consideration especially in endemic regions.

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