Summary

Hydatid disease remains a serious health problem for the Mediterranean countries, such as Turkey. Living in a rural area is an important risk factor for the disease. Hydatid cysts are usually located in the liver, lung, and brain. Mediastinal hydatid disease is very rare that have been only anecdotally in the literature. The objective of this study was to evaluate the clinical and radiographic findings and surgical treatment of this unusual lesion. Between 1985 and 2002, 11 cases with primary mediastinal hydatid cyst were treated surgically at our clinic. Median age was 28.4 and ranged from 19 to 46 years. Symptoms included chest pain in nine patients (82%), and cough in six patients (54%). The cyst was located in the anterior mediastinum in four patients (36%), in the posterior mediastinum in five patients (45%) and in the middle mediastinum in two patients (18%). All cysts were intact except one cyst that ruptured into right intrapleural space. Surgical approach was right thoracotomy in five patients (45%), left thoracotomy in three patients (27%), and median sternotomy in three patients (27%). Total pericystectomy was chosen as the surgical procedure in all patients except four (36%), who had cystectomy and local curettage for cyst located vital structures. There were no complications and mortality postoperatively. Primary hydatid cysts of the mediastinum are distinct clinical entity that must be considered when caring for a patient with a mediastinal mass in endemic regions. Because of surrounding vital structures the cyst should be treated without delay.

Keywords: Mediastinal hydatid cyst; Mediastinal cyst; Surgery; Mediastinal diagnosis

1. Introduction

Hydatid disease is primarily an illness of residents in rural areas who frequently come into contact with carnivores, sheep, and cows. Human hydatid disease caused by the larval form of *Echinococcus granulosus* has a worldwide distribution and is endemic in many countries at the Mediterranean region, the Middle and Far East, and South America [1]. Turkey is also an endemic area to be studied for hydatid disease [2–5]. Hydatid cyst has a predilection to locate in liver, lungs, and brain. Although many uncommon locations have been reported, the disease is rarely present in the mediastinum. Approximately 100 cases have been described in the English literature so far [6–9]. We have treated 11 cases of primary mediastinal hydatid cysts in 17 years period. Although the features of this disease have been described in small series and case reports, to our knowledge no large series defining features of this cysts have been published. In this report the clinical, diagnostic problems and treatment of this rare entity are discussed.

2. Materials and methods

Between January 1985 and April 2002, 427 patients with intrathoracic hydatid cysts were operated at our clinic. Eleven (2.6%) of these patients had mediastinal hydatid cyst. The records of these cases were analyzed for age, sex, signs and symptoms at presentation, location of cyst, type of surgical procedure, hospital stay, and results of operation. The surgery and pathology reports were reviewed to determine lesion location, size, and gross features. The diagnosis of mediastinal hydatid cyst was surgically confirmed in every case.

3. Results

There were six men and five women who lived in a rural area in East Anatolia and were in contact with sheep and sheep dogs. Median age was 28.4 years (range 19–46). Preo-
operative sign and symptoms were present in nine patients (82%), chest pain in nine (82%), cough in six (54%), back pain in two (18%), fever in two (18%), anemia in one (9%), dyspnea in one (9%), and chilling in one (9%). The median duration of symptoms was 4 months (range 0–36 months).

Various imaging techniques were used in the diagnosis of our cases. These included chest roentgenogram and thoracic computed tomograph (CT) in 11, echocardiograph in five, bronchoscopy in four, and esophagoscopy in two. Chest radiographs and CT revealed a mediastinal abnormality in all patients (Figs. 1 and 2). The abnormality was characterized as a mediastinal mass in nine patients, diffuse mediastinal widening in one patient, a mediastinal mass partially obscured by adjacent pulmonary parenchymal consolidation in one patient. Cyst margins were sharply outlined against the adjacent lung in eight lesions. Two masses had a lobulated contour and one mass had a smooth spherical margin. The most frequent CT manifestation of mediastinal hydatid cyst was that of a homogeneous mass containing fluid. Region of interest values in these areas of fluid attenuation, ranged from 16 to 75 HU (mean 32 HU). Only one case had a ruptured cyst, in the other cases cysts were intact. There was no evidence of hydatid disease elsewhere in the body except one patient who had liver cyst.

In seven cases, indirect hemagglutination tests for echinococcosis were obtained in the preoperative period and were positive in five patients. Two cases had eosinophilia (18%).

Of all the patients who had the location of the primary hydatid cyst recorded, four were noted to be in the anterior mediastinal compartment and five were in the posterior mediastinum. Two patients had middle mediastinal cysts. Clinical size of the lesions was recorded in all patients, and ranged from 4 to 9 cm, with a median of 7.2 cm. These sizes were corrected by operations.

Operation was the primary treatment modality in all patients. In preoperative period no patients received medical therapy. Surgical approach was right posterolateral thoracotomy in three patients, right anterolateral in two patients, left posterolateral thoracotomy in two patients, left anterolateral in one patient, and median sternotomy in three patients. Cystotomy and total pericystectomy was carried out in seven cases (64%), in the other cases partial pericystectomy was performed. There were no complications and mortality in postoperative period. The postoperative hospital stay ranged between 5 and 10 (mean 7.3) days. The hospital stay of patients was not correlated with the type of operative approach. Albendazole or mebendazole was administered after the operation to all our patients. The patients were followed for a period of 1 month to 11 years and no recurrence was detected.

4. Discussion

The adult Echinococcus granulosus is a worm that resides in the jejunum of dogs and other canines and produces eggs that are passed in the stool. Eggs ingested by intermediate hosts (cows, sheep, human) liberate an embryo in the duodenum, which passes through the intestinal mucosa to enter portal circulation. Most of these embryos are trapped in the liver; the rest pass through the liver and scatter to other organs and develop into hydatid cysts. Hydatid cysts are rarely present in the mediastinum.

Cystic lesions account for up to one-fourth of all mediastinal masses identified incidentally or during workup for symptomatic mediastinal abnormalities [10]. Cysts of bronchogenic, pleuroperticardial, thymic, intramural esophageal, lymphangiomata, anterior meningocele and enteric origin as well as other rare types may be found in the mediastinum of adults and children. In subsequent years, Thameur and associates [8] identified mediastinal hydatid cysts in 8 of 1619 intrathoracic hydatid cysts (0.5%). Echinococcus of the mediastinum was seen (0.5%). Rakower and Milwidsky [11] recorded more than 23,000 patients with hydatid disease in various large series; only 25 cases (0.1%) of the hydatid cysts were reported in the mediastinal compartment and para-vertebral sulcus. Our region is an endemic area for hydatid disease [3–5]. In our clinic 427 patients with thoracic hydatid...

disease were treated surgically in 17 years period and of these only 11 cases had primary mediastinal hydatid cyst (2.6%).

In general, mediastinal echinococcosis is neither clinically nor radiologically distinguishable from other mediastinal cystic lesions [9,11]. Diagnosis can be obtained through the combined assessment of clinical, radiological, laboratory, and historic data of patient as was seen in our cases. Currently, chest radiograph, CT and magnetic resonance imaging (MRI) facilitate diagnosis. CT is considered essential and is important for displaying morphology, density, and limits of these lesions. It often accurately defines the relationship of the lesion with the adjacent structures. The differential diagnosis in such cases can only be made by surgery. In all our cases, diagnosis was made by surgery. In our clinic, the most commonly employed serological test for diagnosis of cyst hydatid was indirect hemagglutination test that was observed to be positive in five cases. Casoni and Weinberg tests were not routinely used because of their high rates of false positive or false negative results.

Rakower and Milwidsky [11] collected 74 cases and reported in the literature that more than 55% of primary cysts occur in paravertebral sulcus, 36% occur in anterior mediastinum, and less than 8% occur in visceral compartment. In our cases the cyst was located in the anterior mediastinum in four patients (36%), in the posterior mediastinum in five patients (45%) and in the middle mediastinum in two patients (18%).

Symptoms and complications of cyst depend on size, location and involvement of neighboring structures [12,13]. If symptoms related to the cyst were present, they were of compression or erosion of adjacent structures. They ranged from retrosternal or parasternal pain, cough, dysphagia to dyspnea or severe compression of the trachea and superior vena cava [6,9]. In reviewing clinical presentation of mediastinal hydatid cysts, we found that most patients are asymptomatic. Only two patients (18%) were asymptomatic and were to have a cyst by incidental chest films. Marti-Bonatti et al. [12] reported one case in which the cyst ruptured into the aorta. Only one of our patient had a ruptured cyst into pleural space.

The gold standard for therapy is radical removal of the germinative membrane and pericyst through the appropriate thoracic incision [6,9,11]. When the localization of the cyst and invasion to vital structures prevent the total excision, partial pericystectomy is the treatment of choice after the removal of germinative membrane. We performed total excision in 64% of all our patients. It has been proposed that better results are obtained by combining surgical procedures with albendazole for postoperative prophylaxis, and to reduce the incidence of recurrence [2,3]. Postoperative albendazole or mebendazole was administered to all our patients and recurrence was not seen in a long period.

In summary, although rare, hydatid cyst should be considered in the differential diagnosis of mediastinal cystic lesions especially in endemic regions. CT scan of the thorax is the most efficient method of diagnosing these lesions. Surgical resection was successful in all cases and remains the treatment of choice for mediastinal hydatid cyst; additional adjuvant medical therapy is essential to avoid recurrence.

References


