ORIGINAL ARTICLE

ANALYSIS OF PRIMARY INTRATHORACIC EXTRAPULMONARY HYDATID CYSTS A RARE CLINICAL ENTITY

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ABSTRACT

Objective: To evaluate the anatomic location of cysts, operative characteristics of intrathoracic extrapulmonary hydatid cyst and to determine the outcome of aggressive surgical interventions.

Study Design: Case series.

Place and Duration of Study: Thoracic Surgery Unit Lady Reading Hospital Peshawar from 1st July 2008 to 30th June 2011.

Methodology: All patients admitted to Thoracic unit from July 2008 to June 2011 with intra thoracic hydatid cysts were evaluated prospectively as to age, sex, symptoms, diagnostic procedures, anatomic location of cysts, surgical procedures, complications, and outcomes. Chest radiography, computed tomography, and thoracic and abdominal ultrasonography had been performed preoperatively in all of them. Bronchoscopy and spirometry was also performed in all patients for assessment and operability. Echocardiography had been used in 2 patients to determine the contiguity of the cyst to the pericardium. Cystectomy and wide resection were the chief operative procedures. Most of the patients were having cysts in the pulmonary parenchyma only 10 patients had intrathoracic cysts in extrapulmonary locations. This group of patients was included in the study. We excluded patients who had a parenchymal cyst that had perforated to the pleura, myocardial hydatid and patients who had experienced transdiaphragmatic transmission.

Results: Total of 149 patients were operated for hydatid cystectomy, out of these 139 patients had pulmonary hydatid and 10 patients had intrathoracic extrapulmonary hydatid cysts. These 10 patients constitute our study group; out of these 7 were men and 3 women whose mean age was 39.14 ± 16.8 years (range, 16–69 years). Eight (80%) of these were symptomatic, most commonly with chest pain, two patients were asymptomatic. There were 2 (20%) mediastinal hydatid, 2 (20%) diaphragmatic, 2 (20%) pericardial, 2 (20%) oblique fissure, and 1 (10%) each in chest wall and pleural hydatid in our study. Albendazole (10 mg/kg) was prescribed to all patients for as long as 3 months postoperatively. No complication, recurrence, or death occurred during the follow up period of 13 ± 15.4 months (range, 2–36 months).

Conclusion: The extrapulmonary location of Hydatid cysts within the thorax is very rare. This rarity may cause difficulties in diagnosis. To avoid recurrence, it is necessary to resect the affected tissues completely and an anthelmintic medical regimen post operatively.

Keywords: Albendazole, Extrapulmonary, Hydatid cyst, Intraparenchymal cyst.

INTRODUCTION

Hydatid cyst disease, or echinococcosis, is a parasitic disease that has been known since the time of Hippocrates. The physicians and surgeons worldwide may encounter the disease sporadically because of increased travel and immigration[1,2]. It is caused by the parasite, Echinococcus granulosus, which is a cestode that lives in the small intestine of dogs and other canines. Humans are accidental intermediate hosts. The larvae cross the intestinal wall and via the portal system reach the hepatic sinusoids where they develop into cysts. Some larvae are not filtered in the liver, but remain in the blood to reach the next station, the lungs. In addition, some may pass through the pulmonary

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circulation and travel to other organs. Larva transported in the mesenteric lymphatics are carried to the cisterna chili, the thoracic duct, and into the general circulation, ending up in a variety of distant sites[1,3].

Although the liver and the lungs are the usual sites of the disease, cysts can also form elsewhere in the body[4,5]. It is not difficult to diagnose typical pulmonary or hepatic hydatid cysts. Conversely, when cysts appear intrathoracically but in extrapulmonary locations, crucial diagnostic difficulties may occur, with atypical clinical and radiologic signs. Cysts in such sites can lead to fatal complications, such as bronchial rupture, fistulas to the pleural and pericardial cavities, and severe bleeding[6,7].

An intrathoracic yet extrapulmonary cyst is defined as a cyst that is found in intrathoracic extrapulmonary tissues with no involvement of the pulmonary parenchyma and with no transmission of disease from the abdomen to the thorax.

Intrathoracic yet extrapulmonary locations are infrequent, with an occurrence rate of 7.4%[4,6]. Pulmonary sites are the most common site of intrathoracic hydatid cyst development; there, they are called pulmonary hydatid cysts. Conversely, cysts in the diaphragm, pleura, mediastinum, pericardium, myocardium, fissures, and chest wall are called intrathoracic extrapulmonary cysts[6,7] and they can cause a variety of symptoms.

The precise location of an intrathoracic extrapulmonary cyst is usually confirmed during surgical intervention. The corrective surgical procedures, which necessarily differ from those that are performed to treat a pulmonary or hepatic hydatid cyst, should be chosen in accordance with the site of the cyst. To evaluate the anatomic location of cysts, operative characteristics of intrathoracic extrapulmonary hydatid cyst and to determine the outcome of aggressive surgical interventions.

METHODOLOGY

This study was carried out in thoracic surgery unit Lady Reading Hospital Peshawar, from 1st July 2008 to 30th June 2011. Patients admitted to thoracic unit with intra thoracic hydatid cysts were evaluated prospectively as to age, sex, symptoms, diagnostic procedures, anatomic location of cysts, surgical procedures, complications, and outcomes. Total of 139 patients were having cysts in the pulmonary parenchyma only 10 patients had cysts in intrathoracic extrapulmonary locations, this group of patients are included in our study. An intrathoracic yet extrapulmonary cyst was defined as a cyst that was found in intrathoracic extrapulmonary tissues with no involvement of the pulmonary parenchyma and with no transmission of disease from the abdomen to the thorax. We excluded from our study patients who had a parenchymal cyst that had perforated to the pleura, myocardial hydatid and patients who had experienced transdiaphragmatic transmission. Most patients were symptomatic, most commonly with chest pain. Chest radiography, computed tomography, thoracic and abdominal ultrasonography had been performed preoperatively in all of them. Bronchoscopy and spirometry was also performed in all patients for assessment and operability. In 2 patients echocardiography was performed to determine the contiguity of the cyst to the pericardium.

Cystectomy and wide resection were the chief operative procedures. There were mediastinal hydatid, diaphragmatic, pericardial, oblique fissure, chest wall and pleural hydatid in our study. Albendazole (10 mg/kg) was prescribed to all patients for as long as 3 months postoperatively. Data has been analyzed using SPSS version 15. Descriptive statistics were used to describe the results.

RESULTS

This study included 10 patients with intrathoracic yet extrapulmonary hydatid cysts, out of these 7 (70%) were men and 3 (30%)

women whose mean age was 39.14 ± 16.8 years (range, 16–69 years). Eight (80%) of these were symptomatic, most commonly with chest pain as the main complaint, two (20%) patients were asymptomatic. Bronchoscopy and spirometry were also performed in all patients for assessment and operability. Echocardiography had been used in 2 patients to determine the contiguity of the cyst to the pericardium. Magnetic resonance imaging had been used in 2 patients with suspected mediastinal hydatid cyst. In all of these patients cystectomy and wide resection were the main operative procedures that were performed. The location of 10 extrapulmonary hydatid cysts is shown in table. One of the patients with pericardial hydatid was a redo case, this young lady 28 years old had thoracotomy done in other hospital 14 months ago and only cystectomy was done with no wide resection ended up with recurrence. In most patients, the preoperative diagnosis was not identical to the postoperative diagnosis. Cystectomy and wide resection were the chief operative procedures. Costal resection was performed in the patient who had cysts of the chest wall. Pleurectomy was done in patient with pleural hydatid, and the two patients with diaphragmatic hydatid had cystectomy and resection done and diaphragmatic repair done in the end, pericardial resection after identifying the phrenic nerves was done in two pericardial hydatid cysts. Albendazole (10 mg/kg) was prescribed to all patients for as long as 3 months postoperatively. No complication, recurrence, or death occurred during the follow up period of 13 ± 15.4 months (range, 2–36 months).

Table: Anatomic location of hydatid cysts (n=10).

Location	Frequency	Percentage
Mediastinum	2	20%
Diaphragmatic	2	20%
Pericardium	2	20%
Oblique Fissure	2	20%
Chest Wall	1	10%
Pleura	1	10%

DISCUSSION

Echinococcosis remains a substantial public-health problem in many countries, such as Turkey[6,9] where the incidence is 1 in 2,0009. Our country is also a developing country and the disease is not uncommon. Although the liver and the lungs are the usual sites of the disease, cysts can also form elsewhere in the body[7,8]. The cyst may be symptomless for a considerable time or may be associated with slight pain, and may only be discovered when a radiograph of the chest is taken as in our study patients were either having slight chest pain or were asymptomatic. Pulmonary hydatid may be associated with cough. Primary hydatid cysts very rarely form in intrathoracic yet extrapulmonary sites. Hydatid cysts can be found in various tissues[7,8,9]. Intrathoracic yet extrapulmonary locations are infrequent, with an occurrence rate of 7.4%[4,6]. Pulmonary sites are the most common site of intrathoracic hydatid cyst development; there, they are called pulmonary hydatid cysts. Conversely, cysts in the diaphragm, pleura, mediastinum, pericardium, myocardium, fissures, and chest wall are called intrathoracic extrapulmonary cysts[6,13] and they can cause a variety of symptoms. Except myocardial hydatid every type of extrapulmonary site was present in our study. Although hydatid cysts usually produce various symptoms, they can be asymptomatic[12]. Intrathoracic extrapulmonary cysts may produce compression symptoms in surrounding vital structures[13]. Our patients chiefly experienced chest pain (in 80%). Imaging techniques are useful and are usually accurate in the diagnosis of hydatid disease. Although serologic tests are also used, they have lesser value in diagnosis because of false negative and false-positive results[11,14]. We found it possible to establish satisfactory and reliable diagnoses with the use of conventional radiography in concert with computed tomographic scanning and ultrasonography. In addition, the radiologic findings are helpful in the accurate planning of surgery, when that time arrives. Cystotomy and capitonnage are the standard treatments for

typical hydatid disease. Conversely, in the case of intra thoracic extra pulmonary cysts, preoperative diagnostic methods are not always reliable. Difficulties in diagnosis may lead to an incorrectly conceived initial surgical approach. Precise diagnosis usually occurs during surgical intervention. When an intrathoracic extrapulmonary cyst is identified, the surgeon should be aware that cystotomy and capitonnage are too conservative. More aggressive procedures, such as wide resections and reconstruction of surrounding tissues, can achieve complete extirpation without spillage and subsequent recurrence.

We performed cystectomy and resection of the adjacent pericystic structures in all 10 of our patients who had intrathoracic extrapulmonary cysts. The rate of overall bone involvement is less then 2%, which suggests that location in the thoracic cage is not common[15]. Despite this, thoracic-cage involvement occurred in one of our patients. Costal destruction was present in that patient which was removed.

Mediastinal hydatid disease has rarely been reported[16,17]. Mediastinal cysts create pressure symptoms, such as dysphagia, because of esophageal compression[6]. None of the two mediastinal hydatid patients in our study had compression symptoms. The diaphragm is another uncommon site for hydatid cysts[18]. Two of our patients had intradiaphragmatic cysts. After the cysts were removed, the defects were repaired primarily with prolene number 1/0.

There was one patient in our study with primary pleural hydatid, In this patient, many cystic structures were drained, and complete decortication of the parietal pleura was performed. No morbidity, recurrence, or mortality occurred during the follow up period of 13 ± 15.4 months (range, 2–36 months).

CONCLUSION

Extrapulmonary location of hydatid cysts within the thorax is very rare. This rarity may cause difficulties in diagnosis. The precise location of an intrathoracic extrapulmonary cyst is usually confirmed during surgical intervention. The corrective surgical procedures, should be chosen in accordance with the site of the cyst. Total extirpation and reconstruction of the affected structures are preferable to cystotomy and capitonnage.

Conflict of Interest

This study has no conflict of interest to declare by any author.

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