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ORIGINAL ARTICLE

Pulmonary hydatid disease is difficult recognized in children

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Abstract
Purpose Pulmonary hydatid disease is a significant health problem for children in endemic areas. Pulmonary hydatid disease is more frequent than hepatic hydatid disease in children. The aim of this study was to evaluate the characteristics of pulmonary hydatid disease for children in our province which is endemic for echinococcosis.
Patients and methods This study was performed retrospectively between 2007 and 2012 by reviewing the medical records of patients (≤17 years) who were diagnosed with pulmonary hydatid disease. The medical records of these patients were evaluated with respect to the demographic characteristics of patients, management strategies, length of hospital stay, and outcomes.
Results A total of 120 (52.9 % boys and 46.3 % girls) patients with the diagnosis of pulmonary hydatid disease were enrolled in this study. The mean age was 10.15 ± 3.93 years. The significant numbers of patients were admitted with a ruptured hydatid disease and managed with lung preservation. The mean follow-up was 11.3 ± 3.8 (3–24) months. Recurrence was detected in three patients during follow-up.
Conclusion Pulmonary hydatid disease is usually symptomatic. Hydatid disease must be considered in differential diagnosis while evaluating thoracic lesions in endemic areas.

Keywords Children · Hydatid disease · Preservation · Pulmonary · Treatment

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Introduction

Hydatid disease (HD) is a significant medical, social, and economic problem in endemic areas [1]. Hydatid disease has been reported since Hippocrates, may occur at all ages and is seen in all continents [1, 2]. The incidence of HD has decreased in the recent decades [3, 4]. Human HD is a parasitic disease which may involve any organ and spreads with accidentally ingested tapeworm of echinococcus. Four typiest of Echinococcus (*E. Granulosus, E. Multilocularis, E. Vogeli,* and *E. Oligarthrus*) cause infection in humans. Echinococcus granulosus and Echinococcus multilocularis are the most common, causing cystic echinococcosis and alveolar echinococcosis, respectively. [1, 3, 5]. *Echinococcus Vogeli* and *E. oligarthrus*, cause polycystic echinococcosis and are less frequently associated with human infection [3].

HD most frequently involves the parenchyma of the liver (55–75 %); however, some of the parasites escape through the microvascular barrier and reach the lung (15–40 %). Some parasites escape the hepatic and pulmonary filtering system and cause HD in other parts of the human body [5, 6]. However, unlike in adults, lung is the most common involved site in children [1, 7]. Pulmonary HD (PHD) does not have specific symptoms. Clinical symptoms are due to size and location of the cyst, and emerging complications.

Management strategies for PHD consist of medical, surgical and combined management [4, 8].

Liver is the most extensively investigated organ for HD [5]. Most studies of PHD include both adult and children. To the best of our knowledge, there are few studies in the literature specifically dedicated to pediatric population with PHD. The aim of this study is to evaluate the children who had been treated for PHD retrospectively in an endemic region.

Materials and methods

The medical records of 120 children (age ≤17 years) who were treated for PHD from January 2007 to December 2012, were evaluated retrospectively. All medical records were evaluated for demographic characteristics of the patients, presentation symptoms, location of the cyst, management modality, radiologic findings, length of hospital stay and outcomes.

Inclusion criteria: children who were admitted with hospital related PHD. Exclusion criteria: children who had HD without pulmonary involvement.

The diagnosis of PHD was established by various combinations of physical examination findings, routine laboratory test results, and radiological imaging studies. Radiologic workup included conventional roentgenogram, ultrasonography, and computed tomography. All patients had anteroposterior chest X-ray imaging. Computed tomography (CT) was used to confirm diagnosis. Ultrasonography (US) was used to confirm intraabdominal HD involvement. Laboratory tests and serology were performed to support the diagnosis. Serologic tests were not routinely used.

Surgical intervention was performed by a posterolateral thoracotomy in most cases, and for bilateral cases, staged thoracotomy was performed. The cyst was surrounded by iodine-soaked gauze packs to prevent spillage and dissemination to surrounding structures. After injecting diluted iodine solution into the cyst and aspirating the cyst contents, the intact germinative membrane was extracted. The laminated layers were removed and then capitonnage of the cyst wall was performed.

All patients underwent prophylaxis with Albendazole for 3–6 months postoperatively (15 mg/kg/day of albendazole given twice a day orally), in cycles consisting of 45 days of treatment alternating with 14 drug-free days). Patients were regularly followed up postoperatively and visits were performed at 1 month after discharge and every 3 months thereafter for a period of 6–18 months. These visits included clinical examination, abdominal US, X-ray and CT if necessary, and laboratory investigations of blood count, liver function tests, and serologic tests were used to evaluate disease recurrence during the follow-up period.

Statistical analyses

Statistical analysis was performed using the SPSS ver. 11.5 statistical software. Preliminary analyses involved sample frequency tables. Data of the patients were collected and a descriptive statistical analysis of demographic characteristics (age and gender), clinical presentation, PHD locations and outcomes was performed. The data were expressed as mean ± SD (standard derivation) with 95 % confidence intervals. Data were analyzed using the Chi square test. Probability (p) values <0.05 were considered to indicate statistical significance.

Results

This study was performed to evaluate PHD in children. Data of 120 patients with PHD [56 (46.3 %) girls, 64 (52.9 %) boys] were reviewed and evaluated (Table 1). There was no significant difference for gender ($p > 0.05$). Age distribution of the present study was between 4 and 17 years (10.15 ± 3.93 years). The 63.7 % of all patients were ≥9 years. The distribution of age is illustrated in Table 2.

Fifty-three patients had solitary PHD while the rest of the patients had multiple PHD or had concomitant extrapulmonary HD. The most common site of extrapulmonary involvement was liver in 26 patients, while 2 patients had renal involvement and 1 patient had splenic involvement as illustrated in Fig. 1. The mean size of cysts was 3.89 ± 4.41 cm (between 2 and 8 cm). The 71 cases of the

Table 1 Characteristics of the PHD

Gender	56 girls, 64 boys
Mean age (mean ± SD, years)	10.15 ± 3.93
The mean size of cysts (cm)	3.89 ± 4.41
Number of complicated PHD	42
Number of solitary pulmonary hydatid cyst	53
Number of patients with symptom	92
Mean hospitalization time (mean ± SD, days)	7.27 ± 3.13

Table 2 The distribution of age in patients with PHD

Age (years)	Number of PHD
0–5	18
6–8	23
9–13	47
>13	32
Total	120

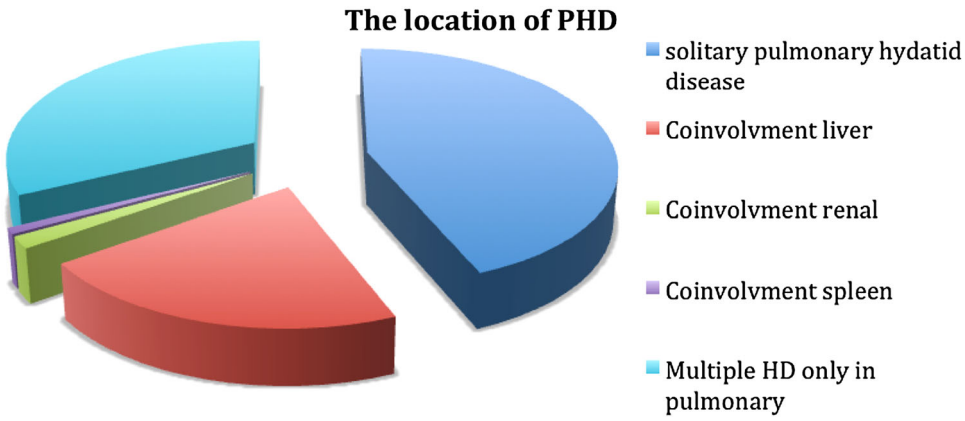

Fig. 1 The location of PHD

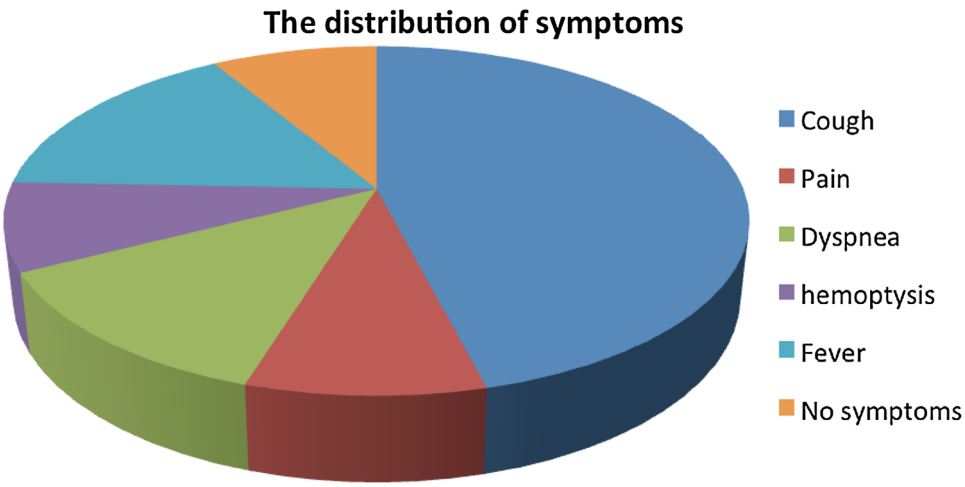

Fig. 2 The distribution of symptoms

PHD were located in the right lung with 6 patients having bilateral PHD. Fifteen patients were initially admitted with liver HD and were found to have PHD too during clinical evaluation.

The most common admittance symptoms were cough (80.2 %), chest pain (15.3 %), dyspnea (22.1 %), haemoptysis (14. %) and fever (27.6 %), while the 14.9 % percent of patients had no symptom (Fig. 2). There were 34.2 % of patients with ruptured hydatid cyst initially at admission. However, the 8.3 % of all patients had tube thoracostomy for assumed diagnosis of empyema, and diagnosed as PHD when the patients underwent surgery for prolonged recovery and seen germinative membrane on surgical exploration. The remaining 57.5 % patients had intact cyst. All patients had slight leukocytosis initially at admission. Serological tests were performed in 40.8 % of patients; 61.03 % of the results of these patients were positive.

Anaphylactic shock was observed in two patients; one of these patients was managed by medical treatment and the other underwent surgery.

Except two patients, all patients were managed surgically under preoperative and postoperative antiparasitic drug coverage. Patients who underwent surgery were managed by cystotomy and capitonnage of the cystic cavity with lung preservation. However, partial resection was performed (segmentectomy) in three patients. The surgical procedure was performed by a posterolateral thoracotomy. Bilateral thoracotomy was performed in six patients and thoraco-abdominal approach was carried out in three patients for multiple-site hydatid disease. We performed VATS for PHD just in one patient.

The mean length of hospital stay was between 3 and 27 (7.27 ± 3.13) days. Overall morbidity rate was 15.1 %. The most common postoperative morbidity was prolonged air leak, which occurred for more than 4 days in 25 patients. Three of these patients required a second operation due to bronchopleural fistula lasting more than 14 days. Surgical site infection was not encountered in any of the patients. Atelectasis occured in four patients. Recurrence was detected in three patients in other part of

body and treated with Albendazole. The mean duration of follow-up was 11.3 ± 3.8 (3–24) months.

Discussion

The 10–20 % cases of HD are diagnosed during childhood [9]. Most cases of HD are infected during childhood period, but they do not present until adulthood [9]. In children the most common organ affected by HD is controversial [10, 11]. In previous studies it was mentioned that the most common involved organ was the liver in adults and the lungs in children [12]. Because patients with PHD more easily become symptomatic than liver HD, due to the compressible nature, higher vascularity, and lower negative pressure of the lung [13, 14].

In previous studies it was reported that HD was more common in males of all age groups [12]. In the present study, PHD also appeared slightly more prevalent in boys, possibly because they contact infected materials more often than girls. The incidence of HD increases with age [12]. Typically, HD is encountered more frequently in children older than 9 years [12]. In the present study, the 63.7 % of patients were ≥9 years. Previous studies reported that the size of the cyst in children increased with age; however, it is also significantly associated with the immune system and tissue elasticity [15]. In the present study there was no correlation between size of HD and age of patients. Approximately 40–80 % of patients with HD have a solitary cyst in a visceral structure, while others have multiple cysts [16]. In the present study, multiple cysts were identified in 55.5 % of patients. The literature search for the incidence of co-involvement of liver and lung by HD yields conflicting results [16]. In the present study, co-involvement of the liver and lung by HD was found in 21.6 % of patients. The prevalence of HD was higher in the right lung and in the lower lobes of both lungs [13]. In the present study, 40.9 % of PHD was involved in the lower lobes.

The most symptomatic patients with PHD are usually caused by mass effect from the cyst volume, amount of pressure that the cyst exerts on surrounding tissues, or rupture of the cyst causing pneumothorax, pleural effusion, or empyema [13, 14, 17]. In the present study, patients were admitted with nonspecific symptoms such as cough, chest pain, and fever. Complicated PHD may have different clinical manifestations [2]. The differential diagnosis includes abscess, empyema, congenital cystic anomalies, and tumors. In the present study, most of the complicated PHD cases were presented as empyema.

The serological tests are used for the definitive diagnosis of Echinococcal infection; however, they are not positive in all HD cases [18]. In general liver HD elicits an antibody response more frequently than lung cysts. Overall, approximately 85–95 % of liver cysts and 65 % of PHD are associated with positive serology, although this varies with the specific serologic test used [19]. In the present study, the serological tests were performed in a low percentage of patients because most patients were admitted with nonspecific symptoms or urgent symptoms' cases were admitted. The serological tests have not been studied in the most of complicated and/or symptomatic patients. This is because the patients underwent radiologic study at initial admission, as well as these regions where HD was endemic in terms of patient.

The diagnosis is usually relies on radiologic imaging studies as our study [2]. Chest X-ray is the first choice and routinely used modality of imaging while CT gives better information about the structure of the cyst and helps in differential diagnosis [15]. US is useful for detection of intraabdominal HD [8]. In the present study, the commonly used diagnostic tool was chest radiography and CT. CT is successful for detecting HD and may identify the cystic nature and localization [8, 19]. However, the serologic and radiologic investigations may also be misleading [4]. Misdiagnosis is common in PHD [17]. In the present study 8.3 % of all patients underwent surgical intervention with misdiagnosis such as pleural abscess or empyema.

There is no standard treatment strategy [15]. Management of HD may be medical, operative or combined [1]. A previous study suggested that medical treatment may be safe and successful in selected children with PHD [4]. In this study, uncomplicated PHD and cysts smaller than 5 cm are suggested as eligible to be treated with medical therapy. However, pulmonary hydatid disease was at risk of rupture more often than liver hydatid disease after medical treatment. Pulmonary hydatid disease is earlier symptomatic and more complicated than liver hydatid disease. Therefore, patients who were symptomatic without regard to the diameter of the cyst decided surgery in the present study. Therefore, the mean cyst size is <5 cm in the present study. Most of our patients were symptomatic. Therefore, the number of medically treated cases was very low.

Surgical treatment of HD consists of cystectomy with closing bronchial openings with or without capitonnage, segmentectomy, wedge resection, lobectomy or pneumectomy [8]. The current surgical treatment is complete excision of the disease process with maximum preservation of the lung tissue [1]. Necessity of capitonnage is a controversial issue [7]. In the present study capitonnage was performed for most of patients because we believe that capitonnage has important role to decrease postoperative complication rates and shorten the length of hospital stay. We concluded that PHD may be treated effectively and safely with parenchymal preservation of the lung as much as possible. The most common postoperative complication was air leakage, pneumonia, recurrence and empyema [13].

In the present study, mostly air leak occured postoperatively and three of them underwent secondary operation. Recurrence occured in three patients.

Several limitations should be considered when evaluating the results of this study. The data collected were from a single center; therefore, the results may not be representative of PHD in other centers. The follow-up period was short, the recurrence rate may not reflect the truth. Therefore, 11 months' follow-up may not be long enough to pick up all recurrences. This period may be extended in further studies. Because of small patients' size with medical treatment therefore, it is difficult to conclude that medical treatment is the first line treatment for PHD. Future controlled clinical trials are needed.

In conclusion, parenchymal sparing procedures must be utilized during surgical management regardless of the complexity of PHD process. PHD may be confused with other lung pathologies especially when complications like superinfection or cyst rupture are encountered. PHD should be considered in the differential diagnosis of pulmonary masses in endemic areas.

References

1. Shehatha J, Alizzi A, Alward M et al (2008) Thoracic hydatid disease; a review of 763 cases. Heart Lung Circ 17(6):502–504
2. Kilic D, Findikcioglu A, Bilen A et al (2007) Management of complicated hydatid cyst of the thorax. ANZ J Surg 77(9): 752–757
3. Grosso G, Gruttadauria S, Biondi A et al (2012) Worldwide epidemiology of liver hydatidosis including the Mediterranean area. World J Gastroenterol 18(13):1425–1437
4. Doğru D, Kiper N, Ozçelik U et al (2005) Medical treatment of pulmonary hydatid disease: for which child? Parasitol Int 54(2): 135–138
5. Dziri C, Haouet K, Fingerhut A et al (2009) Management of cystic echinococcosis complications and dissemination: where is the evidence? World J Surg 33(6):1266–1273
6. Safioleas M, Nikiteas N, Stamatakos M et al (2008) Echinococcal cyst of the subcutaneous tissue: a rare case report. Parasitol Int 57(2):236–238
7. Sayir F, Cobanoğlu U, Sehitoğulları A et al (2012) Our eight-year surgical experience in patients with pulmonary cyst hydatid. Int J Clin Exp Med 5(1):64–71
8. Aghajanzadeh M, Safarpoor F, Amani H et al (2008) One-stage procedure for lung and liver hydatid cysts. Asian Cardiovasc Thorac Ann 16(5):392–395
9. Turgut AT, Altin L, Topçu S et al (2007) Unusual imaging characteristics of complicated hydatid disease. Eur J Radiol 63:84–93
10. Shah OJ, Robbani I, Zargar SA et al (2010) Hydatid cyst of the pancreas: an experience with six cases. J Pancreas 11:575–581
11. Sehitogullari A (2007) Our results in surgical treatment in hydatid cyst of the lungs. Eur J Gen Med 4(1):5–8
12. Mirshemirani A, Khaleghnejad A, Kouranloo J et al (2011) Liver hydatid cyst in children (a 14-year review). Iran J Pediatr 21(3):385–389
13. Usluer O, Ceylan KC, Kaya S et al (2010) Surgical management of pulmonary hydatid cysts: is size an important prognostic indicator? Tex Heart Inst J 37(4):429–434
14. Santivanez S, Garcia HH (2010) Pulmonary cystic echinococcosis. Curr Opin Pulm Med 16(3):257–261
15. Arroud M, Afifi MA, El Ghazi K et al (2009) Lung hydatid cysts in children: comparison study between giant and non-giant cysts. Pediatr Surg Int 25(1):37–40
16. Djuricic SM, Grebeldinger S, Kafka DI (2010) Cystic echinococcosis in children—the seventeen-year experience of two large medical centers in Serbia. Parasitol Int 59(2):257–261
17. Kayhan S, Akgüneş A (2011) Histopathologically diagnosed pulmonary complicated hydatid cyst cases. Turkiye Parazitol Derg 35(4):189–193
18. Bakal U, Kazez A, Akyol M et al (2012) A portable ultrasound based screening study on the prevalence and risk factors of cystic echinococcosis in primary school children in East Turkey. Acta Trop 123(2):91–95
19. Biava MF, Dao A, Fortier B (2001) Laboratory diagnosis of cystic hydatid disease. World J Surg 25(1):10–14