Original Article

Pulmonary hydatidosis in a tertiary care hospital

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ABSTRACT

Background: Hydatid disease is caused by the larval stage of *Echinococcus*. Liver is the most commonly involved organ followed by the lungs. Pulmonary hydatidosis can be primary or secondary. The disease may be asymptomatic for several years. Cause of concern is the fatal anaphylaxis, which may be life threatening. **Materials and Methods:** The present retrospective study is over a period of ten years (2003-2012). The demographic data including the clinical features, radiological findings, other organ involvement, surgical and medical management done and histopathological findings were compiled from the records. **Results:** During the study period a total of eight cases, five male and three female, with age ranging from eight to 43 years were diagnosed as pulmonary hydatid disease. Five patients had presented with complicated cysts. Six patients had solitary cysts involving the lung while bilateral lung involvement was seen in two cases. One patient had multiple pulmonary cysts. Three patients had associated cysts in liver and two in spleen. Surgical lobectomy was done in four cases. Histopathology showed acellular laminated ectocysts in all the cases, whereas endocyst with brood capsules was seen in five cases. **Conclusions:** Pulmonary hydatidosis is not uncommon. Anaphylaxis, although rarely seen, may be a disastrous event. High index of clinical suspicion and mass awareness for interruption of transmission of parasite can lead to proper treatment and possible eradication.

KEY WORDS: *Echinococcus*, histopathology, hydatid cyst, pulmonary, symptoms

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INTRODUCTION

Hydatid disease, a worldwide zoonosis is caused by the larval stage of *Echinococcus*.[1,2] The two main types of human hydatid disease are caused by metacestode stages of *E. granulosus* and *E. multilocularis*. Most frequently caused echinococcosis is by *E. granulosus* commonly seen in the Mediterranean region, Africa, South America, Middle East, Australasia, and New Zealand.[3] Alveolar echinococcosis caused by *E. multilocularis* is less prevalent and has been reported only from Iran, Turkey, Iraq, and Tunisia.[4] The reported annual morbidity from the disease in humans is 1.04-2.4 per 1,000,000 although the actual rates are thought to be more than twice because of underreporting.[5]

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The unilocular cystic form caused by *E. granulosus* is far more common than the rare multilocular alveolar form caused by *E. multilocularis*.[6] Liver is the most commonly involved organ followed by the lungs. Lung involvement is seen in 10-30% of the cases.[7] Secondary involvement caused by hematogenous spread may be seen in almost any location. Pulmonary hydatidosis can be primary or secondary.[8] The disease may be asymptomatic for several years. Although rare, fatal anaphylaxis can occur which may be life threatening.

Diagnostic modalities include clinical, radiological, and serological examinations, done before the surgical treatment.[9] *E. granulosus* cysts are characterized by typical radiological features and easily diagnosed. The *E. multilocularis* lesions are more difficult to diagnose. The alveolar cysts exhibit exogenous proliferation and may simulate a malignant tumor.[10] The diagnostic accuracy of chest radiography is 84% in children and 74% in adults.[11] The definitive diagnosis rests on the histopathological examination of surgically resected tissues. The aim of the present study was to gain an insight into the diverse clinicopathological spectrum of pulmonary hydatidosis.

MATERIALS AND METHODS

A total of eight cases with lung involvement by hydatid disease over a period of ten years (2003-2012) were included in the study. All but one case were ante mortem. The demographic data including the clinical features, radiological findings, and other organ involvement were compiled from the records. The clinical features like history of cough, dyspnea, expectoration, fever, or anaphylaxis were searched for in the medical records. Detailed radiological findings like solitary or multiple cysts, simple or complicated cysts, unilateral or bilateral lung involvement, and involvement of other organs like liver and spleen were recorded.

The specimens received had been earlier fixed in buffered formalin and processed for paraffin sections. The slides were retrieved from the archives and the routine hematoxylin and eosin stained sections were studied. Fresh sections were cut from the blocks wherever required. The parasite morphology, the ectocyst, endocyst along with brood capsules, and pericyst were studied. A careful search for scolices and hooklets was undertaken. A special stain, i.e. Masson's trichrome was done to highlight the hooklets.

The treatment rendered, whether medical or surgical, was also noted from the available records.

RESULTS

During the study period a total of eight cases were diagnosed as pulmonary hydatid disease. There were five males and three females. The age ranged from eight to 43 years. Five patients had presented with complicated cysts. Six patients had solitary cysts involving the lung while bilateral lung involvement was seen in two cases. One patient had multiple pulmonary cysts [Figure 1]. Three patients had associated cysts in liver and two in spleen. The radiological diagnosis of hydatid disease was available in four cases. Two cases were suggestive of lung abscess. The clinical and radiological findings are given in Table 1. Histopathology showed an acellular laminated ectocyst in all the eight cases. The endocyst with brood capsules was seen in five cases. Hooklets were observed in six cases while a scolex was seen in a single case [Figure 2]. Granuloma formation with foreign body giant cells was noted in two cases. The pericyst composed of inflammatory granulation tissue with prominence of eosinophils was seen in four cases. Of the eight cases, a surgical lobectomy was done in four cases.

DISCUSSION

The lungs are the second most frequent site of hydatidosis in adults.[1] Most of the cysts are acquired in childhood and are asymptomatic for years before being diagnosed radiologically on X-ray chest, incidentally most often. Hydatid cysts in the lungs are multiple in 30% of cases with bilaterality seen in 20% and the preferential location being the lower lobes in 60% of the cases. Calcification is a rare phenomenon.[5] Bilateral lung involvement was seen in two cases and cysts were multiple in a single case in our study.

Sudden coughing attacks, hemoptysis, and chest pain are the most common clinical symptoms.[2,5,11] Fever and cough were the most common symptoms in our study. In the event of a cyst rupture cyst fluid, membranes, and scolices may be expectorated.[1] The cyst may even rupture into the pleural cavity. Following cyst rupture allergic reactions may develop; however, fatal anaphylaxis is uncommon. In our study one case was post mortem and the patient could have had an anaphylactic shock. A serious complication is setting of a bacterial infection.[1]

The diagnosis of pulmonary hydatid disease rests on clinical, radiological, and serological examinations.[9] The most important diagnostic tool is radiology. Sharp demarcation and homogeneous density are commonly seen

Figure 1: Gross photograph showing multiple fluid-filled cysts of variable sizes and a laminated whitish membranous layer

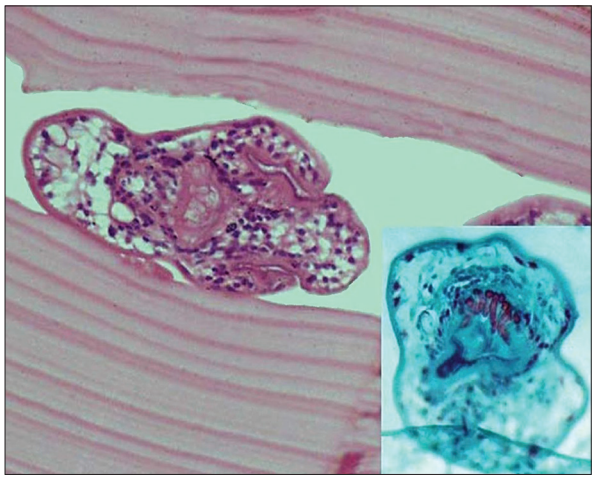

Figure 2: Photomicrograph showing scolices with the presence of scimitar-shaped hooklets (H and E, ×600). Inset shows hooklets highlighted on Masson's trichrome special stain (MT, ×600)

Table 1: Clinical and radiological features in pulmonary hydatidosis

Age/Sex	Location	Presentation	Radiologic findings
8 yrs/M	Right lung and liver	Fever, pain in abdomen	CT chest showing a single cyst with daughter cysts and septations in lower lobe, right lung, with radiologic features of a hydatid cyst in liver on USG abdomen
14 yrs/M	Right lung	Fever, cough	Well-circumscribed mass with minimal pleural effusion and obliteration of costophrenic angle on X-ray chest
15 yrs/M	Bilateral lungs	Cough with expectoration	Cysts in lungs on X-ray chest with bilateral pleural effusion
21 yrs/M	Bilateral lungs and liver	Dyspnea, chest pain	Multiple cysts in lungs and liver with findings typical of hydatid disease on radiology
22 yrs/F	Right lung and spleen	Pain in abdomen	Not available, diagnosed on autopsy
25 yrs/M	Left lung and liver	Hemoptysis	CT chest and USG abdomen suggestive of a hydatid cyst
30 yrs/F	Right lung	Fever with cough	Large cavitary lesion with air- fluid level on chest X-ray; ? abscess lung
43 yrs/F	Right lung and spleen	Pain in abdomen, fever	Right upper-zone consolidation with collapse and cystic area; ? abscess lung and a tiny cyst in spleen; ? abscess on CT scan

yrs/M: Years/Male, F: Female, CT: Computed tomography, USG: Ultrasonography

in case the cysts are intact. Air-fluid levels, water-lily sign, meniscus sign, and a cavity may be seen in complicated cysts.[5] Rupture of the cyst into a bronchus makes it complicated. It does not necessarily mean an infected cyst. The complicated cysts can be accurately localized on computed tomography (CT), which facilitates the surgical management.[12] Apart from the conventional appearances of hydatid cysts on CT scanning of the lung, some unusual CT features of pulmonary hydatidosis include a crescent-like rim of air at the lower end of the cyst; inverse crescent sign and a bleb of air dissecting into the cyst wall resembling the shape of a ring; signet ring sign. Moreover, high CT density indicating fibrosis or infection of the cyst and/or thick wall should not negate a diagnosis of hydatid disease. Bronchoscopy may allow a definitive diagnosis of pulmonary hydatid disease in patients with atypical clinical and radiological features.[13] The bronchoscopic finding of a white glistening membrane is pathognomonic for cystic hydatidosis.[14] Furthermore, bronchoscopy aids in the collection of material for cytological examination.[13,14]

Hydatid disease due to hematogenous dissemination may involve varied anatomic locations.[1,3,15] Lungs, liver, and spleen are the frequent ones while the heart, pericardium, orbit, gastric wall, mediastinum, subcutaneous space, muscle, and adrenal gland are relatively infrequent. Accompanying hepatic and/or splenic cysts are well seen on ultrasonography (USG). The hepatic cysts in pulmonary hydatidosis may be seen in 6-34.8% of patients.[16] Three patients had associated hepatic cysts and two had splenic cysts along with pulmonary cysts in our study. None of the patients in our study underwent serological testing.

Percutaneous aspiration can help in reaching a confirmatory diagnosis by demonstrating scolices, hooklets, or membranes.[17] However, the possibility of cyst rupture with dissemination of contents and anaphylaxis hinders the procedure for routine diagnostic use. The histopathological findings described include the three layers: (i) The outermost pericyst, composed of dense fibrocollagenous tissue with inflammatory cells; (ii) the laminated acellular eosinophilic ectocyst; and (iii) the inner germinal layer or the endocyst from which the brood capsules containing scolices and hooklets develop.[1,18] Granulomatous inflammatory response and calcification may be seen. We observed the ectocyst in all the eight cases, endocyst with brood capsules in five cases, and pericyst in four cases. Hooklets were seen in six cases while scolex was seen in a single case. Granulomatous reaction with foreign body giant cells was noted in two cases. The cysts of *E. multilocularis* differ from those of *E. granulosus* and consist of a spongy pale tissue containing multiple scattered cysts which are lined by a laminated membrane which can be seen microscopically. Central liquefactive necrosis and cavitation are commonly seen in the affected organ.[18] The cavity lacks a cuticle and contains little or no fluid. Most of the cysts in *E. multilocularis* are sterile.

Surgical management is the dominant treatment modality in pulmonary hydatidosis.[16] The current treatment is complete excision with maximum preservation of lung tissue. Surgical intervention should consist of closing all bronchial openings after the membrane of the cyst has been removed.[19] There is an increased possibility of rupture during the separation of the pericyst from the laminated membrane during the enucleation procedure especially in larger cysts.[20] Pericystotomy procedure may not be advocated as it prolongs the surgery time and is associated with an increased risk of hemorrhage.[19] Segmental resection, wedge resection, and lobectomy are justifiable. Surgical lobectomy, done in four cases in the current study is the procedure of choice in large cysts which involve more than half of the lobe, cysts with severe suppuration not responsive to other treatments, multiple cysts and in cases with bronchiectasis, pulmonary fibrosis or severe hemorrhage subsequent to pulmonary hydatidosis.[20] Medical therapy with benzimidazoles (mebendazole and albendazole) with or without praziquantel is indicated in disseminated disease, including secondary lung or pleural hydatidosis, high-risk patients for surgery, spillage during surgery, and small pulmonary hydatid cysts.[17]

However, there is no consensus regarding the choice of surgical procedure but lung preserving surgery is preferred.[16] Medical treatment may be useful in decreasing the recurrence rate.[21] Pulmonary hydatidosis may recur in the same or a different location in the lung. Patients with a pleural lesion must be followed up on a regular basis for any signs of recurrence. Recurrence reported is between 0

and 1.45% of the patients who are on a regular long-term follow-up.[22-25]

CONCLUSION

Pulmonary hydatidosis is not uncommon. Anaphylaxis, although rarely seen, may be a disastrous event. High index of clinical suspicion and mass awareness for interruption of transmission of parasite can lead to proper treatment and possible eradication.

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