

Review Article

Diagnosis and Treatment of Pulmonary Hydatid Disease

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Abstract

Hydatid disease also known as hydatidosis is a zoonotic parasitic infestation of worldwide distribution formed by the larval forms of cestodes belonging to the genus *Echinococcus*. Four species of echinococcosis have been identified that create public health concerns. Human hydatidosis is mainly caused by the larval stage of two species of the tapeworm (*Echinococcus granulosus* and *Echinococcus multilocularis*). This disease is endemic in Eurasia, including Turkey.

Although hydatidosis most often involves in the liver and the lung, it can occur in any organ or tissue. Most patients with pulmonary hydatidosis have no remarkable symptoms in the initial period of the disease. Main symptoms may be chest pain, cough, dyspnea, fever and hemoptysis. Ruptured pulmonary cysts may lead to severe complications such as asphyxia, anaphylactic reaction, tension pneumothorax, hemoptysis, empyema thoracis, and secondary hydatid dissemination. Diagnosis of pulmonary hydatidosis is based on chest radiographs or computed tomography. While serologic response is usually good in hepatic hydatidosis, it may be slight in pulmonary hydatidosis.

Surgical intervention should be performed in thoracic hydatid disease regardless of the severity of the symptoms or the size of the cysts. Various surgical interventions such as enucleation, pulmonary resection, pericystectomy, and cystotomy-capitonnage, can be performed through thoracotomy incision. Medical therapy should be performed after surgical intervention.

INTRODUCTION

Hydatid disease also known as echinococcosis or hydatidosis is a zoonotic parasitic infestation of worldwide distribution formed by the larval forms of cestodes belonging to the genus *Echinococcus spp* (family Taeniidae) [1-3]. It has been known since the time of Galen and Hippocrates. The term hydatid cyst was first used by Rudolphy in 1808 [4].

Four species of echinococcosis have been identified that create public health concerns [3]. *Echinococcus granulosus* (cystic echinococcosis) is the most common species causing human hydatidosis. *Echinococcus alveolaris* (*Echinococcus multilocularis*) is the most virulent species and composed of numerous irregular cystic lesions of various sizes [5]. Human hydatidosis is mainly caused by the larval stage (metacestode) of these two species of the tapeworm (*Echinococcus granulosus* and *Echinococcus multilocularis*) [1,6]. *Echinococcus vogeli* and *Echinococcus oligarthrus* lead to polycystic echinococcosis and they are less frequently associated with human hydatidosis [3,5]. Recently, two other new species have been reported (*Echinococcus shiquicus* and *Echinococcus felidis*), but it is not yet known whether they infect humans [5].

The adult form of this parasite (a tapeworm) lives in the small

intestines of definitive hosts, such as dogs, jackals and foxes, and produces eggs that are passed into the feces. Herbivores such as sheep, goats, camels, cattle, cervids, swine, and horses are the natural intermediate hosts. Humans are accidental intermediate hosts [2,3,7-9]. If the eggs are ingested by the intermediate hosts, oncospheres (embryos) released from the eggs penetrate the intestinal wall until they reach a small vessel. These embryos spread by blood circulation and/or via lymphatics, and migrate to target organs which are mainly the liver and the lung where they mature into hydatid cysts. Most of them are stuck in the hepatic sinusoids. Embryos with less than 0.03 mm may pass through the hepatic sinusoids and settle in the lungs [3]. The life cycle of the parasite is completed by the ingestion of an infected organ of intermediate hosts by any definitive hosts.

Although hydatidosis most often involves the liver and lung, it can occur in any organ or tissue. Cardiac involvement is rare [10]. Hydatidosis is an endemic parasitic infestation in cattle-raising regions of the world [11]. It is endemic in Eurasia, including Turkey. Pulmonary hydatidosis can be primary or secondary. Primary hydatidosis is determined by the evaluation of the embryo; the secondary is a result of the evaluation of fertile elements originating in primary cyst [9].

Cyst structure

Hydatid cyst is composed of three layers with rock water inside. Pericyst or adventitia is the first layer. This layer results from the host tissue's reaction against the parasite. The second is the external layer of the cyst also known as the laminated membrane or exocyst. This layer is an acellular mucopolysaccharide layer. The third layer of the cyst is the inner layer also known as the endocyst or germinative membrane [3,12]. This germinal layer gives rise to the hydatid fluid and brood capsules. Fragmentation of this layer and brood capsules gives rise to daughter cysts. Protoscolices are produced within the brood capsules [5].

The rock water may contain daughter vesicles [12]. It fills the entire cyst and provides required nutrition for larval growth [13]. This fluid contains carbohydrates, proteins, lipids, electrolytes, vitamins and trace elements that may have a role in the metabolism and growth of the cyst [14]. Three of the proteins (beta-hemoglobin, albumin and transferrin) in the cyst fluid are very important for metabolism. Beta-hemoglobin and albumin provides energy to larvae. Transferrin transports the iron needed for the growth of parasite [13]. The concentration of uric acid is very high in the cyst fluid. This high level of uric acid may play an important protective role during the growth of protoscolices [13]. The quantity of uric acid in human hydatid cyst fluids was found to be significantly more than those of the other infested intermediary hosts [15]. The increased uric acid concentration in human hydatid cysts may be due to the high uric acid concentration in humans compared with animals and/or may also indicate degenerative changes in human hydatid cysts [15]. This fluid may lead to anaphylactic reaction, if the cyst ruptures.

Symptoms

Most patients with pulmonary hydatidosis have no remarkable symptoms in the initial period of the disease. Main symptoms may be chest pain, cough, dyspnea, fever and hemoptysis. The most common physical finding is decreased breathing sound at the affected hemithorax [8].

Pulmonary cysts may be complicated if they rupture into a bronchus, pleural space or biliary tree [12]. Endobronchial rupture may lead to severe complications such as asphyxia, anaphylactic reaction, hemoptysis, infection, and secondary hydatid dissemination [9]. If intrapleural rupture occurs, there may be a risk of empyema and even tension pneumothorax [8,16]. The findings of tension pneumothorax are composed of marked shortness of breath, cyanosis, tachycardia and hypotension [16]. Hydatidoptysis (expectoration of cystic material) may occur if a cyst ruptures into a bronchiole [3].

DIAGNOSIS

Diagnosis of pulmonary hydatidosis is based on chest radiographs or computed tomography (CT). Asymptomatic patients with pulmonary hydatidosis can be diagnosed incidentally on chest radiograph [8]. CT is useful in the differential diagnosis (benign and malignant tumors). Ultrasonographic examination of the abdomen should be performed to investigate the accompanying hepatic hydatidosis in all patients with pulmonary hydatidosis. Perforated pulmonary cysts adjacent to the pericardium may be detected by echocardiography [16].

Although there are different immunologic techniques for the diagnosis of hydatid disease, results of at least two tests should be combined in order to increase diagnostic accuracy [17]. IgG ELISA and IHA are the most sensitive serologic tests [17]. The decrease in cellular immunity that accompanies pregnancy might result in negative serology. Seronegativity does not rule out the disease [16]. Thus, the predictive accuracy of serological tests is limited [18]. The Weinberg complement fixation testing and Casoni's skin testing may not be done due to their low diagnostic value [7].

While serologic response is usually good in hepatic hydatidosis, whereas it may be slight in pulmonary hydatidosis [19]. Additionally, false positive test results may appear in patients infested by other helminthes' larva and adult forms due to cross-reactions [19]. The positive serologic findings are less common in pulmonary hydatid cysts compared with liver cysts [3].

Serological methods are never 100% accurate, and a marked antibody response might not be produced by some patients with hydatidosis [20]. In these circumstances, molecular methods can be required due to subgenotypes of cystic echinococcosis and subspecies of alveolar echinococcosis. Echinococcus species can be detected and discriminated by molecular methods [1].

Eosinophilia may be an important finding especially in patients with perforated cysts [8]. Eosinophilia is seen in 10%-30% of patients positive in hydatid disease. Eosinophilia increases in case of rupture and it is also high in countries where hydatidosis is endemic [21].

Hydatidosis may recur even in surgically treated patients [22]. There may be no marked clinical or radiological findings initially in case of recurrences. In recurrent cases, seropositivity can be 70% [23]. Therefore, serological tests may be suitable for postoperative follow-up [8]. Although early recurrence was attributed to incipient microcysts, late recurrence was attributed to reinfestation. Hydatidosis may be recurred not only in the same area, but elsewhere in the pulmonary parenchyma [23]. The human basophile degranulation test (performed with parasite antigens) may be useful in estimating the possibility of recurrence after surgical intervention [24].

The diagnosis of intrathoracic hydatidosis is based on the imaging characteristics and serologic tests in most cases. Sometimes, infection and degeneration of the cyst may hamper the correct diagnosis. In these circumstances, histology confirms the correct diagnosis [12]. All cystic contents (germinative membrane, sandy rock water) should be sent for histopathological examination. All precautions must be taken not to spill of the cystic contents while taking and transporting the samples.

TREATMENT

Surgical intervention should be performed in thoracic and cardiac hydatidosis regardless of the severity of symptoms or the size of the cystic lesions [7]. Standard thoracotomy incision is often performed to remove pulmonary hydatid cysts (Figure 1). Median sternotomy incision may be suitable for removal of both pulmonary and cardiac cysts. The purpose of surgical treatment is to remove the entire cysts/entire cyst while preserving the lung



Figure 1 Operative view showing giant hydatid cyst removed through thoracotomy incision.

parenchyma as much as possible. Various surgical interventions such as enucleation, pulmonary resection, pericystectomy, and cystotomy-capitonnage, can be performed through thoracotomy incision. In countries where the hydatidosis is endemic, parenchyma-preserving surgical procedures should be preferred, because the hydatid disease may recur even in surgically treated patients. Thus, cystotomy-capitonnage should be the preferred surgical technique in endemic countries, including Turkey [22]. However, pulmonary resection may be required due to irreversible bronchiectatic changes in some cases [7]. Cystic fluid spillage may lead to anaphylaxis and secondary seeding of hydatidosis. To avoid dissemination into the surgical field, the operative field should be surrounded with towels soaked with protoscolicidal agents (20% hypertonic saline solution or 1% povidone iodine solution). If broncho-biliary communication is present, biliary leaks should be repaired before applying these agents to avoid risk of sclerosing cholangitis or pancreatitis [25]. Some hydatid cysts may be operated with video-assisted thoracoscopic surgery (VATS). During VATS, care must be taken to avoid the spillage of cyst contents, and the surgical site should be protected with protoscolicidal agents as performed in open surgery. The eventual recurrences may be avoided using these precautions. VATS should not be used in patients with giant hydatid cyst associated with tight adhesions or air trapping lung [7].

Benzimidazoles (mebendazole, albendazole) and praziquantel are used in the medical treatment of hydatid disease [12,25]. Albendazole and mebendazole act by inhibiting microtubular assembly within the parasite, leading to impaired glucose absorption through the wall of the germinative cell layer [3]. The success of medical treatment is not 100% [7,26]. The reported cure rates range from 25% to 30% [26]. Thus, medical therapy should be performed in inoperable patients or after surgical intervention to avoid a relapse as an adjunctive therapy [22]. Albendazole should be preferred to mebendazole due to its fewer side effects and efficiency in lower doses [22]. But, medical therapy should not be used in patients with significant hepatic insufficiency or bone marrow suppression [25]. Praziquantel has protoscolicidal activity. It may be used alone and/or in combination with albendazole [25]. Medical therapy also should not be used during pregnancy, because these drugs have teratogenic effects. Albendazole therapy is administered at a dose

of 400 mg twice a day for four weeks in two or three cycles with a two-week break between the cycles to avoid the possibility of liver damage [27]. The optimal duration of albendazole treatment in lung hydatidosis is unknown, but it should be administered at least 3-6 months [3]. During drug therapy, laboratory tests (biochemical and hematological) should be repeated before every course of therapy in each patient [8].

CONCLUSION

In conclusion, pulmonary hydatidosis should be treated surgically and conservative surgical procedures should be preferred, if possible. During postoperative follow up, patients should be followed up with serological tests and chest radiographs, and abdominal ultrasonography, if required.

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