Review Article

Pulmonary hydatid cyst: Review of literature

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Abstract

Echinococcosis is a rare infectious disease in human being that occurs by the larval stages of taeniid cestodes of the genus Echinococcus. Human cystic echinococcosis is the most common presentation. The liver is the most common site of echinococcal cyst, followed by the lungs. The symptoms of lung infestation lead to sudden onset of chest pain, cough, fever, and hemoptysis after a cyst rupture. The diagnosis is confirmed by radiology supplemented with serology. Chest X-ray and computer tomography of chest is the principal investigation for pulmonary hydatid cyst (PHC). The treatment of PHCs is either pharmacotherapy and/or surgery. Surgical intervention is the most preferred treatment of choice; pharmacotherapy is useful in selected patients. Pharmacotherapy includes oral administration of benzimidazoles group of drugs like mebendazole or albendazole.

Keywords: Enucleation of cyst, hydatid cyst, pulmonary hydatidosis

Introduction

Echinococcosis is a rare infectious disease of human being that occurs by the larval stages of taeniid cestodes of the genus Echinococcus. Till now six species have been identified, of which four are pathogenic: Echinococcus granulosus [responsible for cystic echinococcosis (CE)], Echinococcus multilocularis (responsible for alveolar echinococcosis), Echinococcus shiquicus found in Tibet, and Echinococcus felidis in African lions are the other species which have no zoonotic transmission potential.[1,2] Echinococcosis continues to be a major community health burden in several countries, and in some terrain it constitutes an emerging and re-emerging disease.[3] CE is the most common human diseases of this genus and it accounts for >95% of the estimated 2--3 million cases worldwide.[4] CE is endemic in many parts of the world like Mediterranean countries, southern America, Australia, eastern and northern Africa, and in the Tibetan terrain of Asia.[5] CE may often be diagnosed accidentally because the individual may remain asymptomatic for a long time due to the silent nature of the pathogen.[6] In India, there are many literatures on the above disease in animals as well as human being.[7]

Clinical Features

The eggs of Echinococcus hatch and release embryos in the small intestine after ingestion. The invasion of mucosa leads to blood-borne distribution of the larvae to the organs like liver and lung, where the cyst develops. Most primary infections consist of a solitary cyst; however, multiple cysts or multiple organ involvement have also been reported. The liver is the most common site of the echinococcal cyst of the pastoral strains (>65%), followed by the lungs (25%); the spleen, kidneys, heart, bone, and central nervous system are rarely involved. The slow growing nature of the cyst is responsible for its late presentation in adulthood even if the disease is acquired in childhood.[3] The pressure symptoms of the enlarging cyst or its

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complication produces symptoms in extrahepatic hydatidosis and the presentation may be late due to its slow growing nature.[8] Usually, cysts greater than 5 cm in diameter leads to bronchial compression. Cyst rupture, secondary infection, suppuration, and pneumothorax are the common presenting complications of pulmonary hydatidosis. Cyst rupture may lead to sudden onset of chest pain, hemoptysis, cough, and fever or rarely a salty taste in the mouth. Urticaria and wheezing to anaphylaxis may occur due to hypersensitivity of the ruptured cyst which at times may be life threatening.[9] A generalized toxic reaction may be due to the presence of parasites.[10] A pulmonary hydatid cyst (PHC) most commonly produces symptoms of cough followed by chest pain, breathlessness, expectoration, fever, hemoptysis, and anaphylactic phenomena.[11] Hemoptysis is more common presenting complaints in adults' population.[12] Screening for the presence of hepatic hydatid cysts should be done in all patients with PHCs because of the high incidence of the coexistence and also due to the asymptomatic nature of cysts in this location. It has been reported that patients with hepato-PHCs presents earlier than the PHCs.[13]

Diagnosis

Chest X-ray and computer tomography of the chest supplemented with serology is the investigation modality for this disease. Peripheral blood smear showing leukocytosis, eosinophilia, and raised erythrocyte sedimentation rate are nonspecific parameters observed in patients with this infection.[3]

Imaging techniques

Chest X-ray is the initial investigation tool for PHC [Figure 1]. Better imaging modalities like computed tomography, and magnetic resonance imaging of the lungs are most useful to image PHC and its complications.[12]

The radiological features of a PHC are sharply defined, round-to-oval homogenous opacity of variable size [Figure 1].[12] Peripherally located cysts are larger than those located at the major bronchovascular structures.[14] PHCs are usually solitary but multiple cysts may also be found.[15] Calcification and daughter cyst formation is rare in PHCs unlike extrapulmonary hydatid cysts.[16] The pleural hydatid cysts may calcify.[17] Computed tomography features of PHCs are smooth walls of variable thickness and homogenous internal contents of water or near-water density [Figure 2]. The adventitious (pericyst), laminated, and germinal layers of an intact cyst are clubbed together and hence seen as a single wall.[18] If there is any air into the potential space between the pericyst and ectocyst (laminated membrane of the parasite), the local detachment of parasitic membranes from the pericyst occurs which is known as "the sign of detachment." This segmental peripheral radiolucent feature is known "the crescent" or the "meniscus sign" which is a reliable sign for pulmonary hydatid disease but is not pathognomonic.[19] Radiologic features of ruptured hydatid cyst are known by different names like double-arch or cumbo sign, iceberg sign, sign of the rising sun, serpent sign, and whirl sign. If air enters into the parasitic membranes, the endocyst collapses and an air-fluid level is seen. If the parasitic membranes float on the fluid surface, this produces the "water lily sign" or "Camelot sign" resembling leaves of a water lily. If all the parasitic contents are aspirated or removed, only the pericyst produced by the host remains, which is also known as "empty cyst sign."[20] Detached or collapsed endocyst membranes, collapsed daughter cyst membranes, and intact daughter cysts are the pathognomic computer tomographic features of ruptured hydatid cysts. The presence of air pockets or air bubbles within the cyst and ring enhancement of the pericyst on contrast enhanced CT indicate either infection or cyst–bronchial tree communication.[21] The differential diagnosis of infected hydatid cyst is abscess or neoplasm due to its high density.[22] Proper evaluation of extrapulmonary intrathoracic hydatid cysts like chest wall, mediastinal, pericardial, fissural, and pleural localization necessitates magnetic resonance imaging.

Laboratory Tests

Immunoelectrophoresis or electrosyneresis, indirect immunofluorescence, enzyme-linked immunosorbent

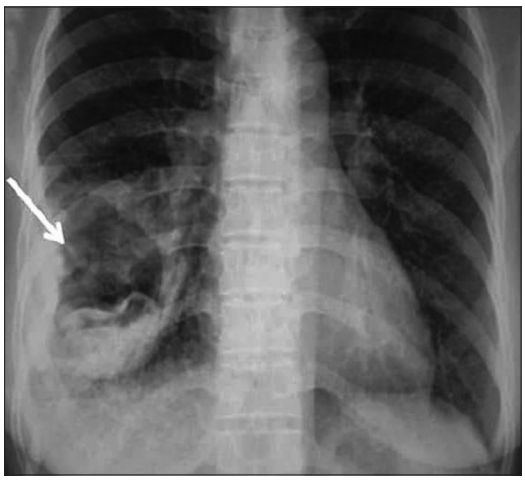

Figure 1: Chest X-ray showing hydatid cyst of right lung

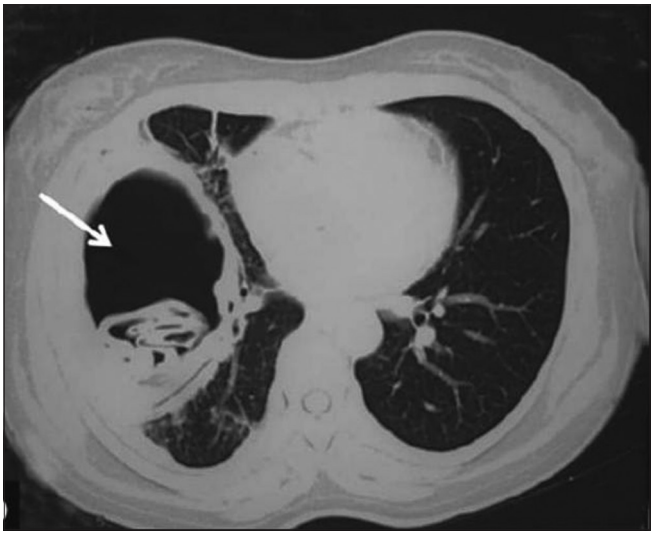

Figure 2: CECT of thorax showing hydatid cyst of right lung

assay, or hemagglutination are the serological evidence of echinococcosis.[23] The hydatid serology is not sensitive tool for isolated pulmonary hydatidosis.[18] Serologic tests may still be remain positive long time after cyst removal.[20]

Treatment of PHC

PHCs may be treated pharmacologically and/or surgery. Surgical intervention is the treatment of choice. PHCs are sometimes treated pharmacologically by oral administration of benzimidazoles group of drugs like mebendazole or albendazole which includes smaller cysts, patients with contraindication for surgery, disseminated disease, multiple cysts, recurrent cysts, and patients with intraoperative spillage of hydatid fluid.[11,24] Albendazole is the drug of choice because of its higher bioavailability and requirement of minimum contact period of approximately 11 days.[25] Its usual recommended dose is 10–15 mg/kg/day, taken twice daily, approximately should be given for at least 3–6 months for PHCs. Continuous dosage administration has been found to be more efficacious than the earlier belief of interrupted monthly dosage with a gap of 2 weeks to avoid hepatotoxicity.[26] Patients with hepatic hydatid cyst may have a coexistent pulmonary hydatid cyst and hence these individuals must have a screening protocol for lung involvement as patients with widespread PHC who are not amenable for surgery do benefit from albendazole.[27]

Patients who developed complications during medical management (mebendazole or albendazole) for PHCs do present around 2 months after the initiation of treatment and hence these patients should be followed for at least 2 months.[28] Cysts larger than 6 cm in diameter are at risk of rupture, inactive or calcified cysts, patients who are prone for bone marrow depression, pregnancy, specifically the first trimester of pregnancy are contraindications of medical management.[28]

Surgery

Surgery is the gold standard treatment of choice for PHCs of any size [Figure 3]. Large cysts that are superficial and vulnerable to rupture, infected cysts, cysts in close proximity to vital anatomical structures, and cysts exerting substantial mass effect are treated surgically.[29] The cysts are most commonly approached through posterolateral thoracotomy. Median sternotomy or two-stage thoracotomy is reserved for bilateral cysts. In patients with cysts in different hemithoraces with one of the cysts ruptures, operate the unruptured cyst first owing to the risk of rupture.[30] The detrimental effects of spillage of cystic contents can be contained by placing gauze soaked with 20% hypertonic saline solution or concentrated 10% povidone--iodine solution in the operative field.[31]

The following surgical techniques are done:

Enucleation (Ugon method): It consists of removal of the cyst with its intact germinative membrane [Figure 4]. It is suitable for the small PHCs with little risk of rupture. Positive pressure ventilation helps in enucleation of cysts. Larger cysts should not be treated because of the risk of rupture.[32] Postoperative complications like air leak and infection may occur in few cases due to the presence of pericyst.

Pericystectomy (Perez--Fontana method): This procedure involves removal of hydatid cyst along with the pericyst which allows complete removal of the parasite. Though pericyst is not a part of the parasite, pericystectomy may lead to increased risk of airway leak.[33]

Cystotomy with capitonnage (Barrett's method): Cystotomy involves aspirating fluid from the cyst along with removal of the germinative membrane (Barret technique). Capitonnage helps in reduction of the risk of the infection of the residual cavity, airway leak, and empyema formation, but there is a risk of disfigurement of the lung parenchyma.[34]

Cystostomy with closure of the bronchial openings and capitonnage (Posadas method): The procedure is similar to Barrett's method, but in this method the opened airways are closed prior to capitonnage. This helps in reduction of risk of the infection of left over cystic cavity, airway leak, as well as empyema formation. Hence, the outcome of this procedure is satisfactory except that it may lead to atelectasis due to lung parenchyma disfigurement.[33]

Cystostomy with the closure of the bronchial openings alone: In this method, the capitonnage procedure is not done which results in less disfigurement of the lung parenchyma but carries an increased risk of air leak and infection is increased.[35]

Open aspiration by Figuera technique: This procedure is similar to percutaneous aspiration, instillation of scolicidal agents, and reaspiration (PAIR) used for hepatic hydatid cyst. In this procedure, the cyst membranes and daughter cysts are sucked out. This procedure is less invasive but carries an increased risk of infection and air leak within the cavity and subsequent empyema formation.[33]

Segmental resection: This procedure is reserved for ruptured hydatid cyst. The method follows the conventional anatomic or nonanatomic resection technique. This procedure has

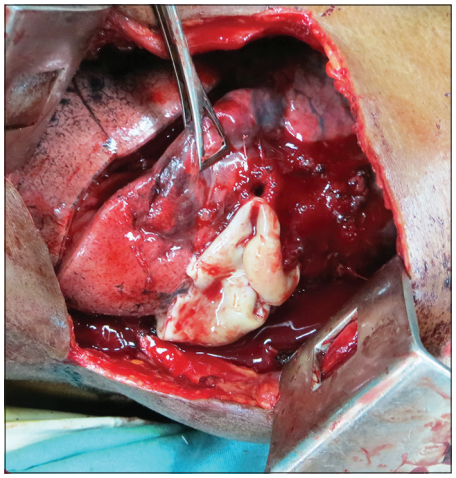

Figure 3: Operative picture of enucleation of hydatid cyst

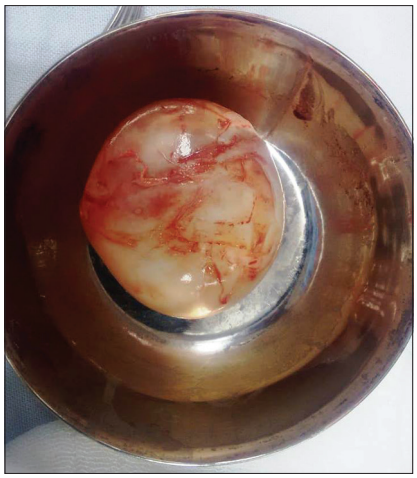

Figure 4: Intact specimen of hydatid cyst of lung

reduced infection and recurrence rate. There is reduction of lung volume and hence the compliance.

Lobectomy: This procedure involves anatomic resection of one or more of the lobes of the lung involving the cysts. If a cyst involves greater than 50% of the lobe, infected cysts not responding to treatment, multiple unilobar cysts, and the consequences of hydatid disease such as bronchiectasis, pulmonary fibrosis, or severe hemorrhage, lobectomy is better suited.[36]

In case of multiple cysts, the plan and type of surgery should be given to cyst based on their susceptibility to rupture, size, and the risk of dissemination. An unruptured cyst should be treated first in patients with both an intact and a ruptured cyst. Due consideration should be given to obliterate the dead space after management of large cysts to minimize the air leak and empyema.[12] Superficial and small to moderate hydatid cysts may be managed with video-assisted thoracoscopic surgery.[37]

Postsurgical Follow-Up

All patients with hydatid cyst surgery should receive albendazole (10 mg/kg/day) for 6 months to prevent recurrence of the disease.[33] The risk of recurrence is as high as 11% if antihelminths are not prescribed post-surgery.[38] The postoperative follow-up consists of clinical examination, liver function tests, and chest X-ray once a month for the first 3 months which is then continued every 3 months till the end of first postoperative year.[39] Surgical removal of the cyst should be taken very meticulously to avoid its spillage on the surgical field as it may lead to a life threatening anaphylactic reaction as well as incomplete extraction of the cyst leading to a risk of recurrence. Prophylactic measures like community education initiatives, proper hand hygiene after contact with animals like dogs are essential preventive measures.[40]

Conclusion

PHCs are the rare infectious disease of the pulmonary system and needs proper management. Delay in the management leads to constellation of complications which if not taken care may lead to a life threatening situation. Surgery for PHCs, irrespective of size, and intact or ruptured, can be safely performed, with low morbidity and negligible mortality rates, and is the treatment of choice.

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Conflicts of interest

There are no conflicts of interest.

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