

Pulmonary Hydatid Disease

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Introduction

The word hydatid is Hellenic in origin and means "a cyst full of water". Hydatid disease exists all over the world and is a major health problem in the Middle East.

Epidemiology

Hydatid disease is a serious public health hazard and a major problem for the community. It exists in most parts of the world and is hyperendemic in sheep raising areas. There are foci in New Zealand, Australia, the Indian continent, the Mediterranean basin, East Africa, Eastern Europe, Iceland and Latin America, but it is most prevalent in the middle east. In Iraq, the disease is still endemic and is considered to be one of the most serious helminthic disease in the country. Domestic hygiene, the standard of living and human behavior play an important role in the epidemiology of transmission to man.

Life cycle of the parasite

Hydatid disease is caused by the adult and larval stages of the *Echinococcus Granulosus*. The adult is a cestode of 5-7 mm. in length with a scolex bearing four suckers and with a body containing two to six proglottids. The larval stage or metacystode is a cyst with acellular laminated outer layer and an inner nucleated germinal layer which gives rise to brood capsules and then scolices. The adult worm lives in dog which is the primary host. The cystic part of the cycle occurs in sheep, horse, pig, ox, camel or mule, which act as intermediate hosts.

The adult worm lives in dog's intestine for months. The terminal segment proglottids detach from the worm and release thousands of ova with the stools. These are ovoid in shape and can remain infective for long periods. Ingestion of the ova by sheep in contaminated water, grass or vegetables causes infection, and hydatid cysts subsequently develop in the viscera. The parasite gets back into the dog when the sheep is slaughtered and its viscera containing hydatid cysts are eaten by another dog. In each of these cysts thousands of scolices are found which will grow into new tapeworms in the dog's intestine and so the cycle repeats itself indefinitely.

If an ovum gets into a man instead of a sheep, the life cycle of the parasite will be broken because the entrails of men are not eaten by dogs. Contamination of food is a common source of infection in man, more frequently he contaminates his fingers by fondling a dog and then takes his food without washing his hands. The ova hatch in the stomach and the hexacanth embryos or oncospheres are released in the stomach and duodenum and pass through the wall and travel via the portal vein to the liver, where the majority settle and develop into hydatid cysts. Only those embryos which successfully pass through the capillaries in the liver to reach the lungs develop into hydatid cysts, and the few which get through the lungs are then carried to the viscera and tissues supplied by the systemic circulation. Sometimes the hatched embryo passes through the wall of the stomach or duodenum into a lymphatic channel to be

carried into the lungs via the thoracic duct, thus by-passing the liver; this explains the few cases of pulmonary hydatids without hepatic involvement.

Once implanted in the interstitial tissue of the lung, the embryo grows into a vesicle and then into a hydatid cyst.

Pathology

The hydatid cyst in the lung is surrounded by a capsule formed of atelectatic alveoli and fibrous tissue. This adventitia (pericyst) is part of the host and becomes thicker and tougher with age, thus separating the cyst from the lung parenchyma; this is of importance in the surgical excision of the cysts.

The hydatid cyst itself is composed of a nucleated thin germinal layer (endocyst) which secretes the hydatid fluid into its cavity and forms externally an acellular white laminated membrane (ectocyst). The germinal layer is the only living part of the cyst. The laminated membrane is completely separated from the adventitia. And this explains the feasibility of surgical removal of intact hydatid cyst by enucleation.

The main bulk of the cyst is made up of its contained fluid which is highly antigenic.

Brood capsules develop from the germinal layer by the process of asexual budding and vacuolation and are attached by pedicles to the innermost wall of the germinal layer. Within each brood capsule, two to three scolices (head of future worms) develop. The attachment of the brood capsule to the germinal layer becomes loose and the capsule bursts, releasing the scolices into the fluid of the cyst. These fall to the bottom by gravity and are sometimes referred to as "hydatid sands".

There is a tendency for the growing cyst to enlarge peripherally towards the pleural space (an important factor in considering the surgical approach to the cyst).

In this paper the term "simple" cyst indicates that the cyst is intact , and the term "complicated" indicates that it has been injured, ruptured, or infected.

Rupture into the bronchial tree is common whilst rupture into the pleura is very rare.

In partial rupture, the adventitia bursts, small bronchial fistulae appear and air may enter the space between the adventitia and the laminated membrane which is still intact. This appears as a crescent-shaped "perivesicular pneumocyst".

In complete rupture, the cyst itself ruptures and a small amount of fluid escapes between the laminated membrane and the adventitia. If at the same time a little air gets in through small bronchial fistulae, a fluid level will appear on either side of the shrunk but dome-shaped hydatid. This is known as "double arc sign".

Complete rupture of the cyst may permit expulsion of its contents through large bronchial fistulae and allow air to enter the cyst; the laminated membrane then collapses and floats upon what remains of the fluid, producing irregular projections above the level of the fluid. The appearance is likened to a "water- lily" floating on a pond. One of two complications may develop in this cavity with the retained disintegrated membranes inside it: either some parts of the germinal layer survive in spite of complete rupture and form "daughter cysts" in the space occupied by the original cyst, or the parasite dies after complete rupture, and the retained disintegrated fragments of the laminated membrane acts as a sequestrum and with the associated bronchial fistulae lead to infection and the formation of a "lung abscess". Occasionally all the membranes are coughed out leaving a "pulmonary cavity" and a

cured patient. Rarely, "calcification" may occur in the adventitia of complicated cyst but this does not necessarily signify death of the parasite.

A "primary hydatid cyst" indicates that the cyst has developed from an ovum, whilst a "secondary hydatid cyst" means that a primary cyst lodged in the lung or in the liver has ruptured and caused new daughter cysts to develop. These daughter cysts develop either from fragmentation of the germinal layer left behind after spontaneous evacuation or inadequate surgical removal of the parasite, or from scolices in the liquid which has been split during surgery or spontaneous rupture. This latter possibility is questionable, since the elements inside a hydatid cyst should go on to grow into tapeworms, however; Deve in 1945 proved that live hydatid elements could revert to new hydatid cysts by the process of retrograde metamorphosis.

Clinical features

1. There is a high incidence of the disease in children who fondle dogs and in young men who work with dogs on farms.
2. There is no significant difference in sex distribution.
3. The lesions may be asymptomatic unless, or until, some complications develop.
4. Many patient present to the thoracic surgeon with a circumscribed opacity found incidentally on a routine CXR.
5. Patients with intact or mildly infected cysts may experience transient symptoms of pain, cough, and hemoptysis.
6. The acute onset of rigor, fever, cough with purulent expectoration and pleuritic pain is experienced by all patients with lung abscess.
7. Fragments of membranes or daughter cysts, described by patients as "grape skin", may be coughed up when cysts have ruptured into the bronchial tree.
8. Severe dyspnea may occur in the rare event of rupture into the pleural cavity causing hydro. or pyopneumothorax.
9. Rupture of a cyst may cause signs of anaphylaxis. this rare presentation is due to the high antigenicity of the hydatid fluid.

Diagnosis

I. Radiographic findings

1. a well defined circular or oblonged shaped opacity of homogenous density is diagnostic of a simple intact hydatid cyst.
2. a pathognomonic radiographic finding is a "perivesicular pneumocyst" due to the presence of air between the adventitia and the laminated membrane. this appears as a slender crescent "signet-ring" in cases of adventitial rupture.
3. the "double arc sign" appears as a cyst with a fluid level on either side of the shrunken but dome-shaped hydatid.
4. the "water-lily" or "camalote sign" appears as a cyst with the collapsed membrane floating in the fluid and is seen in cases of rupture of the cyst itself.
5. daughter cysts may produce radiographic appearances identical with the primary cysts. A large daughter cyst with a partially or completely drained pericyst produces the "rising sun sign".
6. if suppuration occurs, a huge abscess with conspicuous fluid level marking is seen.
7. often, an empty cavity with remnants of collapsed membranes are seen.

8. rarely, all the membranes are coughed up and a "pulmonary cavity" surrounded by a thin margin is seen.
9. in rare instances, the adventitia may calcify, and this gives the appearance on X-ray of a thin shell of an egg.
10. hydropneumothorax may be seen, and this indicates rupture into the pleural cavity.
11. bilateral multiple cysts is a common radiographic finding.

II. Non invasive procedures

- *Isotope liver scan
- *Ultrasonic scanning
- *CT scan

These procedures are of great value in demonstrating the exact site, size, and the number of cysts, specially the liver.

III. immunological tests

1. Casoni intradermal test: Absolute reliance can not be placed on a positive Casoni reaction because it may be positive in an apparently unaffected individual living in an endemic area , and it may remain positive long after the cyst have been removed . this test have been abandoned in most centers because of its non-specificity and difficulty in obtaining test fluid.

2. Weinberg complement fixation test

This is the best method of detecting infestation and is accurate.

3. Latex slide agglutination test

This is simple, reliable, and non specific.

4. Indirect haemagglutination test

This is relatively non specific

5. Immunoelectrophoresis

This demonstrates the specific band 5 of echinococcus granulosus.

IV. Blood test

Eosinophilia is common finding, but this is of little value since multiple parasitic infestation are common in the middle east.

V. parasitological test

These are based on the finding of scolices, brood capsules, and daughter cyst in the sputum or pleural aspirate.

Differential diagnosis

Radiological diagnosis of pulmonary hydatid disease is a problem when the cyst is simple, small and single, or when multiple and bilateral. When distinction must be made from other causes of solitary round peripheral lesions or from metastases.

In Iraq, the finding of a solitary opacity of homogenous density and clear-cut borders on a CXR will be a hydatid cyst in 90% of cases.

Hydatid cyst is a major cause of lung abscess in Iraq, but it must be differentiated from other causes of lung abscess.

Surgical treatment

The treatment for hydatid cyst is surgical removal. The various surgical techniques currently in use are:

A) removal of the cyst

the principles of removal of a hydatid cyst are evacuation of the contents of the cyst, avoidance of spillage during the procedure, closure of the bronchial fistulae, and prompt re-expansion of the lung.

1. aspiration\evacuation technique

The technique of aspirating the fluid with a fine needle followed by the extraction of the cyst through a small adventitial incision was described by Barrett in 1947. this procedure can be used for simple and complicated cysts. Aspiration may result in some leakage, but this will not be dangerous if adequate precautionary packing of the operative field has been carried out. The author has used this technique in most patients for the last 20 years with good results.

2. enucleation

Enucleation in the plane between the adventitia and the laminated membrane was first described by Ugon in 1947, suggested by Barrett in 1949, and advocated by Barrett and Thomas in 1952. Technically this is a beautiful procedure , though applicable only to simple cysts . although a plane of cleavage can readily be found between the adventitia and the laminated membrane, there is appreciable risk of accidental rupture during the dissection, if this does occur, the risk of contamination is high, much greater than in aspiration\ evacuation technique such a high risk is not acceptable to most thoracic surgeons.

3. excision

Excision of the cyst with its capsule was first described by Fontana in 1948, suggested by Barrett and Thomas in 1952 and Xanthakis in 1972. This procedure is applicable to all cases irrespective of whether the cyst is simple or complicated and it offers the best chance of avoiding soiling. It appears sound, simple and attractive, but the continuous oozing of blood and the persistent leakage of air from the raw surface of the lung may render it unsuitable.

After removal of the cyst by whether method, the edges of the sac must be trimmed and sutured, and a meticulous search must be made for all bronchial openings which should be sutured individually. The residual sac is left open, and there is no need to obliterate it.

Zafirokopoulas in 1969 reported the results of microscopic examination of cysts from more than 300 patients , in 4%, live scolices and daughter cysts were found in the layers of adventitia and the surrounding lung parenchyma, pulmonary resection was suggested for these cases. A similar results was obtained by Galloway and Rob in 1976. A follow up of the authors patients and of others shows that recurrence following removal of the cyst is minimal.

B) Resection

Pulmonary resection is reserved **for**:

- 1) giant simple cyst that causes permanent and irreversible changes in the affected part of the lung.
- 2) complicated cyst where infection has caused a lung abscess or bronchiectasis.
- 3) multiple daughter cysts involving one lobe.
- 4) calcified cysts.
- 5) patients presenting a diagnostic problem.

6) patients with massive hemoptysis, or recurrent suppuration following the removal of a cyst, resection of the involved part of the lung is advisable.

Methods of resection are:

1. segmentectomy

This is the usual procedure. The aim being to preserve as much lung tissue as possible.

2. lobectomy

This is the procedure of choice when resection is indicated.

3. pneumonectomy

This is rarely needed.

Notes

When the disease is bilateral, the side with a large simple cyst or with a complicated cyst is operated on first, and after a few weeks the other side is tackled.

In patients with lung and liver involvement, the priority is for the pulmonary lesion.

Recent rupture into the bronchial tree should be treated conservatively by treating the anaphylactic shock, postural drainage, administration of antibiotic, and regular observation for signs of recurrence.

Rupture into the pleural cavity with hydropneumothorax requires thoracotomy. The main objective is removal of the cyst, which is usually found floating in the pleural cavity, and re-expansion of the lung, even in patients with pyopneumothorax and daughter cyst formation in the pleura, the same treatment is indicated.

Medical treatment

The medical treatment of hydatidosis is far from being established, no drugs have yet been found that can effectively reach and destroy the parasite even if drugs are capable of destroying the live elements of the cyst do become available, the retained membranes will ultimately become infected and produce lung abscess.

Mebendazole (vermox) and flouromebendazole are being benzimidazole derivatives and may have a limited role in the treatment. They are safe drugs with few side effects and may be used **in** disseminated cases when the cysts are multiple and inaccessible and **when** surgery is contraindicated for reasons of extreme ill health. Mebendazole is prescribed in high dose 50 mg/kg/day in divided doses for three months.









