

Multivesicular Primary Mediastinal Echinococcosis

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Abstract

A rare case of multiple, multivesicular mediastinal hydatid cyst with cardiac involvement but without associated hepatic & pleuropulmonary hydatidosis prompts this case report and brief review.

Introduction:

Echinococcosis or hydatid disease also known as "Liver full of water, watery tumor of chest" etc, first coined by Rudolphi in 1800. It is derived from Greek word 'Hydatid' meaning 'a drop of water' (1). Echinococcosis is a zoonotic disease caused by larvae of the dog tapeworm Echinococcus. Four species are recognized. E. granulosus which is the most common cause of human infestation causes cystic echinococcosis, which has a worldwide distribution. Humans are exposed less frequently to E. multilocularis, which causes alveolar echinococcosis, E. vogeli and E. oligarthrus which causes polycystic echinococcosis. Hydatid disease is mainly found in areas where cattles, goats and sheep are raised as they are the definitive host. Dog & human are intermediate or carrier hosts. Dogs pass ova in their feces which in turn contaminate food and water ingested by man. The larvae pass through the intestinal wall of man into portal venous system. As the liver is traditionally considered as first filter and lungs as the second filter, they are the most common sites for hydatidosis. It is observed that other sites like mediastinum, breast, bone, spleen, kidney, heart etc. may be affected with or without involving the liver and or lung, as in our case.(2,3) Hydatid cyst of mediastinum is extremely rare and that too multiple, multivesicular is not reported in literature.

Case Report

A 37 year female, sweeper by occupation, came to emergency department with complaints of breathlessness since 1 year, increased since 1 week, dry cough & fever since 7 days. Except tachypnea rest of the examination was normal. Routine X- ray chest showed cardiomegaly. USG Chest showed multiple variable sized cystic lesions in mediastinum. Echocardiography revealed multiple cystic lesions largest in septum measuring 4.5*4 cms, multinodular mass in Left atrium measuring 4.7*3.6 cms. with moderate MR, mild TR & PH. CT Chest & Abdomen and Cardiac MRI showed multiple multivesicular, well circumscribed cystic lesions with daughter cysts seen in visceral mediastinum above diaphragm, extending to anterior & posterior mediastinum. Intracardiac cysts were present. Compression and mass effect were seen on posterior wall of LA & LV. There was no evidence of lung or liver cyst. Patient underwent surgery but could not tolerate it. On autopsy, various sized hydatid cysts were present in pericardial space, anterior & posterior mediastinum. Anterior segment of lung showed infarcted area with corresponding pulmonary artery showing embolus of hydatid cyst (germinal epithelium)

Discussion

Infestation by hydatid disease in humans most commonly occurs in the liver (55-70%) followed by the lungs (18-35%), the two organs can be affected simultaneously in about 5-13% of cases.(4) 15% hydatids occur in extra hepatic or extra pulmonary locations. Cardiac involvement is 0.5-3%, involving left ventricular cavity in 52%, septal location in 42%, right ventricular location in 31%, pericardium in 10% and

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both intracardiac and the pericardial cavity in 10%. (5,6) Mediastinal echinococcosis is very rare, incidence being 0.1-0.5%. (7,8) The cyst is located most commonly in the posterior mediastinum in 45%, while in the anterior mediastinum in 36%, and in the middle mediastinum in 18%. (9)

The larvae pass through both barriers into the arterial circulation or the larvae could travel via intestinal lymphatics into the thoracic duct, thus bypassing the liver and get implanted in various other sites. (10) Hydatid cyst of liver and other abdominal hydatids are frequently multivesicular but 90% of pulmonary hydatids are single and univesicular. Multiple & multivesicular mediastinal hydatid cysts are not reported in literature. The larvae in the cyst form the germinal layer (endocyst) and lay down the thick white membrane (ectocyst). The pericyst is merely the host's reaction to the parasite. (11) It takes half a year from larval insertion to 1-2 cm cyst formation. Formation of daughter cyst depends not only on immunologic relationship between parasite and human but also on resistance offered by the enveloping structures. (12) If resistance offered is more, primary cyst ruptures and pieces of germinal epithelium form daughter cysts. (12) As lung tissue offers least resistance the lung cysts are usually univesicular as compared to liver cyst which usually contain daughter cyst, because of its tissue resistance. As the symptoms depend on the size & site of the lesion, most of them are incidentally found. Among symptomatic thoracic hydatidosis, chest pain-82%, cough-54%, back pain & fever-18% each was found in one study. (9)

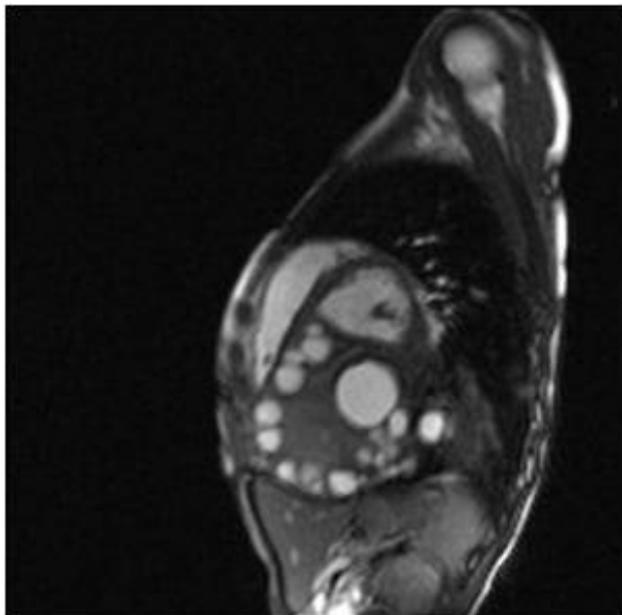
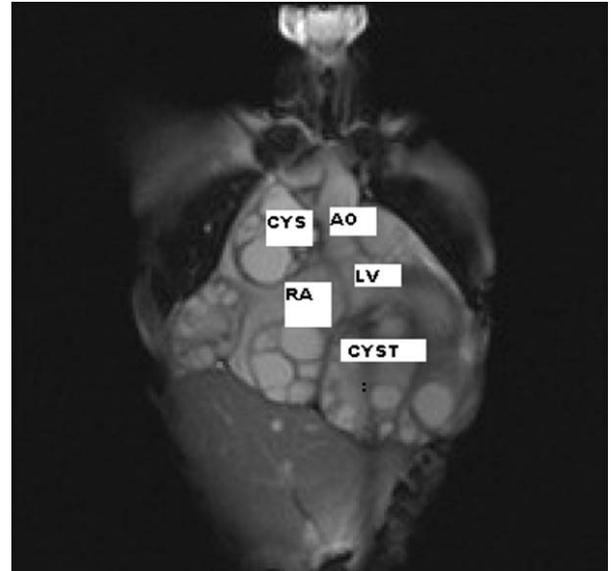
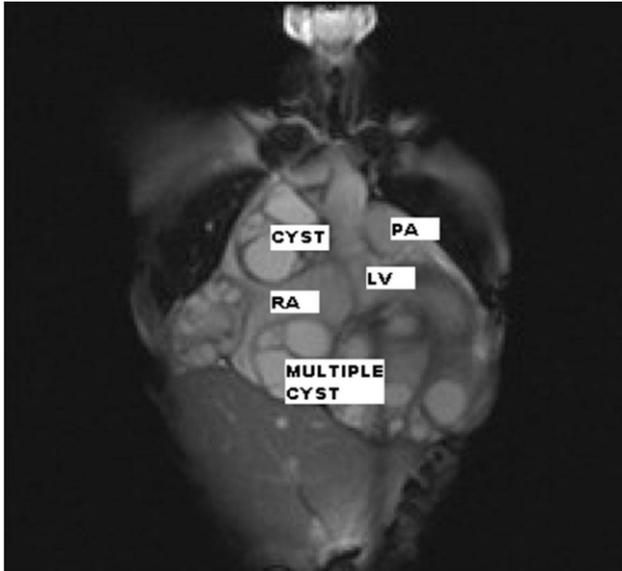
The incidence of cystic lesions of the mediastinum is 20%. Most common cystic lesions include 1) Pericardial cyst, 2) Bronchogenic cyst, 3) Enterogenous cyst, 4) Dermoid cyst. etc. (13) Incidence of mediastinal

hydatid cyst is 0.1-0.5% in various studies. (7,8) Currently, chest radiograph, ultrasonography, CT scanning and magnetic resonance imaging (MRI) facilitate diagnosis of hydatid cyst. Casoni and Weinberg tests are not routinely used because of their high rates of false positive or false negative results. Newer serological tests include, enzyme-linked immunosorbent assay (ELISA), latex agglutination and indirect haemagglutination (IHA) test and an immunoblot assay using lentil-lectin purified glycol proteins and it has >99% specificity and the test is highly sensitive. Less than 15% of cases exhibit eosinophilia. Specific diagnosis could be made histologically by demonstration of parasite in excised tissue, by fundoscopic visualization of parasite or by neuroimaging studies by demonstrating a cystic lesion with scolex.

The gold standard for therapy is radical removal of the germinative membrane and pericyst. . For uncomplicated lesion percutaneous aspiration, infusion of scolicidal agents like hypertonic saline, cetrimide, formalin, ethanol etc. and reaspiration (PAIR) is now indicated instead of surgery. As medical therapy, albendazole, given at 400 mg twice a day for 28 days and repeated from 1 to 8 times, separated by the drug free interval of 2 to 3 weeks, is most efficacious in reduction of cyst size & prevention of recurrence after surgical removal. (14)

In summary, although rare, hydatid cyst should be considered in the differential diagnosis of mediastinal cystic lesions especially in endemic regions. Imaging with CT scan of the thorax, MRI is the most efficient method of diagnosing these lesions. Surgical resection is the gold standard treatment, additional adjuvant medical therapy is essential to avoid recurrence.

MRI Images: showing multiple multivesicular, well circumscribed cystic lesions with daughter cysts seen in visceral mediastinum above diaphragm, extending to anterior & posterior mediastinum.



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