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Case Report

Pathology Section

Primary Hydatid Cyst of the Lung: A Review of the Literature

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ABSTRACT

Hydatid cyst or Echinococcosis is one of the most important helminthic zoonotic diseases in the world. Hydatid disease remains a serious health problem in endemic countries, like India. Living in rural areas is an important risk factor for the disease. Cystic hydatid disease may develop in almost any part of the body. The lung (25%) is the second most commonly affected organ after the liver (75%). The two organs can be affected simultaneously in about 5%-13% of the cases. Primary hydatid disease indicates that the cyst in question had developed from an embryo which had been derived from a dog. Secondary hydatid disease means that a primary cyst which had been lodged elsewhere had ruptured and caused a new cyst to develop by embolism or by direct spread. The clinical presentation of hydatid disease is often non-specific and many patients may be asymptomatic. As long as 5 to 20 years may elapse before the cysts enlarge sufficiently to cause symptoms. The symptoms depend on the size and the site of the lesion and on the accessibility of the organ which is involved for the clinical examination. Serology and imaging modalities establish the diagnosis in most of the cases. Hydatid cyst should always be considered in the differential diagnosis of cystic and mass lesions of the lung in endemic areas like India. It is diagnosed by viewing the cystic membrane. The final diagnosis can be confirmed by a histopathological examination. We are hereby reporting a case of 55-year-old man with a primary hydatid cyst of the lung.

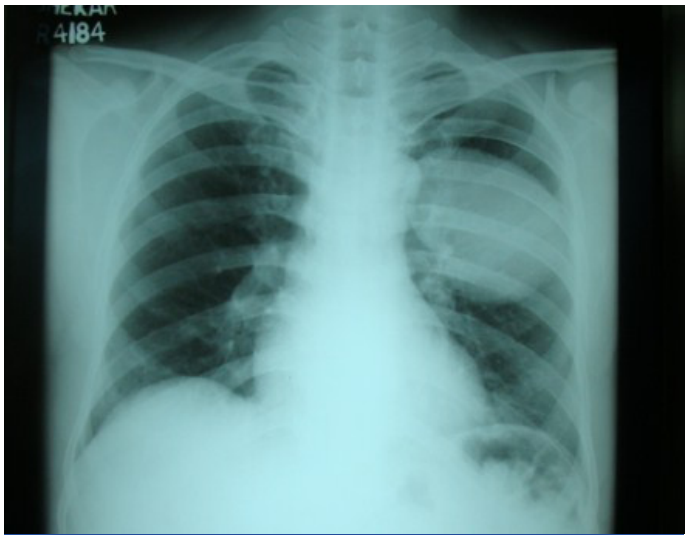
Key Words: Primary Hydatid Cyst, Lung

INTRODUCTION

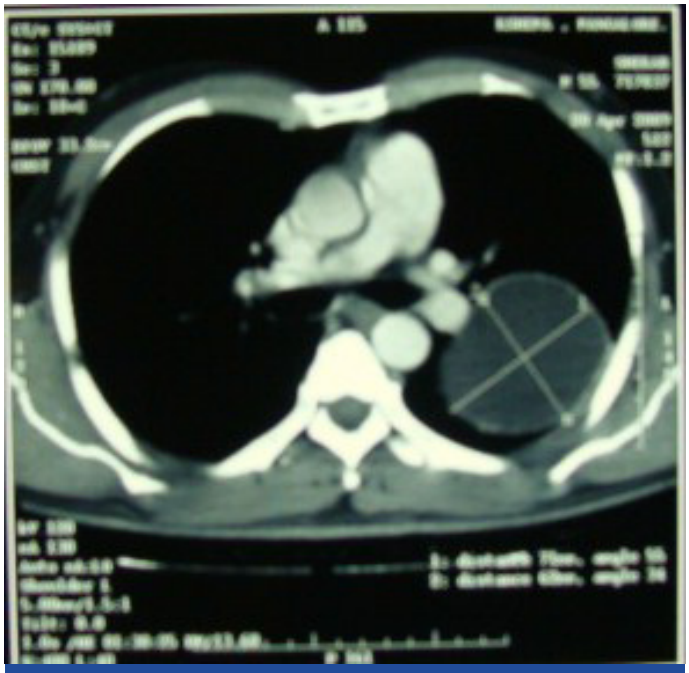
Hydatid disease is a worldwide zoonosis which is produced by the larval stage of the Echinococcus (E) tapeworm. 4 species of it have been recognized and a vast majority of the infestations in humans are caused by E.granulosus. E. granulosus causes cystic Echinococcosis, which has a worldwide distribution. Humans are exposed less frequently to E. multilocularis, which causes alveolar Echinococcosis. E.vogeli and E. oligarthus are rare species and they cause polycystic Echinococcosis. Hydatidosis is quite common in India and so this disease should always be considered in the differential diagnosis of cystic and mass lesions of the lung [1,2].

CASE HISTORY

A 55-year-old man who hailed from a nearby village presented with a history of a non-productive cough of 6 months duration, who had no other significant contributory history. There was no history which was suggestive of bronchial asthma, drug intake, allergy, or chest trauma. There was no history of fever, chest pain or breathlesssness. On examination, he was found to be febrile with a respiratory rate of 28 breath per minute and a pulse rate of 90 beats per minute. His chest examination revealed diminished movements over the left hemi thorax, with decreased breath sounds in the left infrascapular area. His cardiovascular examination was unremarkable. His per abdominal examination revealed no organomegaly. His haemat-ologiacl and biochemical investigations were within normal limits. His chest radiograph showed a rounded, well defined, cystic lesion with homogenous opacity in the upper lobe of the left lung [Table/Fig-1]. Computed Tomography (CT) confirmed the same [Table/Fig-2]. The rest of the lung field and the mediastinum were unremarkable. A radiological diagnosis of hydatid cyst of the lung was rendered. Ultrasonography and CT abdomen was performed to rule out a possibility of hydatid cyst of the liver. Subsequently, the patient underwent surgery. Per operatively, the lesion was accessed by posterolateral thoracotomy. A large, white, cystic mass was seen in the posterior segment of the left upper lobe and the lesion was firmly adhered to chest wall and the apical portion of the superior segment of the lower lobe. The lung parenchyma over the mass was stretched and it was firmly adhered to the surface. Left upper lobectomy was performed due to the adhesion and the involvement of the adjacent lung. Grossly, we received a cystic



[Table/Fig-1]: Chest radiograph showing a round well defined cystic lesion with Homogenous Opacity in the upper lobe of the left lung



[Table/Fig-2]: Computed tomography confirms the cystic nature of the lesion in the upper Pole of Left lung

[Table/Fig-3]: Gross specimen showing white membrane like tissue

white mass [Table/Fig-3]. A final histopathological diagnosis of hydatid cyst was made by the gross and the microscopic features. The patient is clinically asymptomatic and a follow-up X-ray which was done after 2 years is unremarkable.

DISCUSSION

Hydatid disease has been known since the time of Galen and Hippocrates, and it was described by Thebesius in the 17th century. Rudolphi (1808) first used the term hydatid cyst to describe Echinococcosis in humans. Echinococcosis is endemic to the Mediterranean region, South America, Australia, New Zealand, Alaska, Canada, and the Middle East [3].

Hydatid disease, also known as Echinococcosis or Hydatidosis, is caused by infection with the larva of the tapeworm of the genus, Echinococcosus. In its adult stage, the parasite lives in the intestinal tract of carnivores such as dogs and cats, as well as in herbivores such as sheep. After being eliminated with faeces, the eggs contaminate fields, irrigated lands and wells. The E. granulosus cysts, following a primary infection, may inhabit any anatomic site. Humans contract the disease from water or food or by direct contact with dogs. Once the eggs reach the stomach, the hexacanth embryo is released. These pass through the intestinal wall and reach the tributary veins of the liver, where they undergo a vesicular transformation and develop into hydatids. If they overcome the hepatic obstacle, they become lodged in the lung, where they also transform into hydatids. If they advance beyond the lung, they may remain in any organ to which they are carried by the blood stream. It has been shown that the embryos can reach the lung via the lymphatic vessels, bypassing the liver, and there is also evidence that the disease can be contracted through the bronchi. The two most common organs which are involved are the liver (75%) and the lung (25%). The other less common sites which are affected by these cysts include the muscles (5%), bones (3%), kidneys (2%), spleen (1%), heart (1%), pancreas (1%) and the central nervous system (1%) [1,4,5].

Thameur [3] et al., reported the frequency of the intrapulmonary hydatid cyst of E.granulosus to be 94.6% and an extra pulmonary rate of 5.62% in 1,619 patients with thoracic hydatidosis. The hydatid cysts are typical; one lobe is involved in 72% of the cases, usually at the lung base. The right lung is more commonly involved than the left lung. In the case which is under discussion, a circular cyst was noticed in the upper lobe of the left lung. A hydatid cyst which is not open to the pleura appears as a circular or an oval image with well defined limits that can change according to its evolution. A ruptured cyst radiologically appears as a pneumopericyst and if the content of the cyst is completely evacuated into the bronchial tree, a cavity which is similar to tuberculosis or an abscess, appears [3, 4].

The symptoms depend on the size and the site of the lesion. Slowly growing echinococcal cysts generally remain asymptomatic until their expanding size and their space occupying effect in an involved organ elicits symptoms, as in the case which is under discussion. The common presenting symptoms are cough, chest pain, and breathlessness. The symptoms of the mediastinal hydatid cysts which are reported in the literature range from cough, dyspnoea, chest pain and dysphagia, to Horner's syndrome, Pancoast syndrome and paraparesis, though many cases are asymptomatic. A rupture or an episodic leakage from a hydatid cyst may produce fever, pruritis, urticaria, eosinophilia, or fatal anaphylaxis. The rupture of a hydatid cyst into an adjacent bronchus may occur due to vigorous coughing and due to the expectoration of a large amount of salty sputum which consists of mucus and hydatid fluid and occasionally, fragments of the cystic membrane are reported. Every patient who has hydatid cysts of the lung should be investigated for associated cysts in the liver or in other abdominal organs by chest CT and abdominal ultrasonography, as were performed for the case which is under discussion. The cysts which are accessible through the diaphragm should be resected at the same setting [1, 2, 4].

Routine haematological and biochemical tests do not help in the diagnosis of hydatid disease. Serology and imaging modalities establish the diagnosis in most of the cases. The Casoni test, the Weinberg complement fixation test and total eosinophil counts are not reliable. Radiological studies play a very important role in detecting and evaluating the echinococcal cysts. Plain films will define the pulmonary cysts, usually as rounded, irregular masses of uniform density, but they may miss the cysts in other organs, unless there is cyst wall calcification, just as it occurs in the liver. Ultrasonography, Computed Tomography (CT), Echocardiography and Magnetic Resonance Imaging (MRI) are of great value in diagnosing and

determining the anatomic extent and the relationship of the cysts. Enzyme Linked Immunosorbent Assay (ELISA) is the most widely used assay. The 5 arc immunoelectrophoresis test is confirmatory, as it detects the antibodies against the immunodominant and the specific antigen (antigen 5) of the cestode. Polymerized Chain Reaction (PCR) which uses the recombinant Deoxyribonucleic Acid (DNA) antigen is valuable in defining the particular species of the Echinococcosus. However, no serological tests like ELISA or PCR were performed in this index case due to the cost factor and the diagnosis was confirmed by viewing the cystic membrane and by a histopathological examination [2,4,5,6,7].

Preventive measures are important and they include washing of the hands with soap and warm water, washing of fruits and vegetables before consumption, and deworming of pet dogs. The World Health Organization has published an excellent overview on the treatment guidelines for echinococcal diseases in 1996. The conventional treatment for hydatid cysts in all the organs is surgery. The surgical methods which are related to the pulmonary cysts include cystotomy and enucleation of the intact cyst, with or without capitonnage, for the complicated or the intact cysts. The current treatment for hydatid disease of the lung is complete excision of the cyst, which includes the germinative membrane, with the maximum preservation of the lung tissue. Limited thoracotomy and conservative surgical procedures should be preferred to get a cosmetic skin appearance, good postoperative recovery and less post operative pain. Radical procedures such as pnemonectmy, lobectomy and segmentectomy should be avoided as far as possible. Intra-operatively, the use of hypertonic saline or 0.5% silver nitrate solution before opening the cavities tends to kill the daughter cysts and it thus prevents further spread or an anaphylactic reaction. Mebendazole and, recently albendazole and tinidazole have been used as a primary drug therapy and as an adjunct to the surgery, to diminish the recurrence and the potential spread of the organism. Furthermore, the literature indicates that the recurrence of this disease is extremely rare. Chemotherapy is the preferred treatment, if surgery is not available, when the complete removal of the cyst is impossible, when the cyst contents threaten to disseminate due to the rupture of the daughter cysts or when the cysts are too numerous [1,2,3,8,9].

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