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Medicine

A CASE OF PRIMARY PULMONARY ECHINOCOCCOSIS PRESENTING AS LEFT SIDED GROSS PLEURAL EFFUSION

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ABSTRACT Invasion of the human lungs by the larvae of the dog tapeworm *Echinococcus granulosus* (pulmonary cystic echinococcosis, PCE) is an incapacitating disease, frequently found across a wide geographic area, CE is endemic in many parts of the world, particularly the Mediterranean countries, Central Asia including the Tibetan Plateau, Northern and Eastern Africa, Australia, and South America.[1] Global burden of the human AE is approximately 18,235 new cases per annum with the majority (91%) occurring in China.[2] We report a case of 17-year-old girl student who presented with complaints of left sided pleuritic chest pain, fever and dyspnoea. X-ray chest PA view revealed gross pleural effusion on left side that was exudative in nature and did not respond to antibiotics and anti-tuberculous therapy. HRCT chest revealed large loculated cystic lesion showing calcification in left upper lobe parenchymal region suggestive of ruptured hydatid cyst. IgG antibody for *E granulosus* was positive. Ultrasound scan of abdomen and pelvis were within normal limits. A diagnosis of Primary pulmonary echinococcosis was made. She was treated successfully with Albendazole 400 mg *bid* for 21 days; six such cycles 14 days apart.

KEYWORDS : Echinococcosis, Hydatid disease, Pulmonary echinococcosis, benzimidazoles.

INTRODUCTION

Echinococcosis in human, also known as hydatid disease, is a zoonotic disease distributed worldwide and is caused by the larval stage (metacestode) of the parasite belonging to the family *Taeniidae* and genus *Echinococcus*. Four species are recognized to cause public health concerns. *Echinococcus granulosus* (*E. granulosus*) causes cystic echinococcosis (CE) and is the most common species to cause disease in human. Although *Echinococcus multilocularis* (*E. multilocularis*), causes alveolar echinococcosis (AE) is the most virulent species and rare also.[3] Hydatid disease is a major zoonotic disease of public health and economic significance. Cystic echinococcosis (CE) is the most common presentation of echinococcosis in humans, contributing to more than 95% of the estimated 2–3 million global cases.[4] We report a case of primary pulmonary echinococcosis, who presented with large loculated exudative pleural effusion that did not respond to antibiotic and antitubercular treatment.

CASE REPORT

A 17-year-old girl without any comorbidities, presented with history of unmeasured fever without chills and malaise and for 10 days, dry cough without diurnal or postural variation and left sided pleuritic chest pain for seven days and gradually progressive breathlessness, without postural or diurnal variation of three days' duration. At the time of presentation patient was breathless even at rest.

Clinically, patient appeared severely ill, had pallor and palpable left cervical nontender lymphadenopathy. Her pulse was 110/min regular, blood pressure 110/70 mmHg, respiratory rate 26/min, temperature 102^{o}F and oxygen saturation (SaO_2) on room air was 92%. Examination of respiratory system revealed signs of volume gain on left side with stony dull percussion note and decreased breath sounds suggestive of pleural effusion right. Examination of other systems was unremarkable. X-ray (Pic 1) and ultra sound scan of chest confirmed left sided gross pleural effusion. Patient was managed with intercostal tube drainage. Pleural fluid cyto-biochemical analysis revealed exudative pleural effusion with polymorphonuclear predominance (pleural fluid leucocyte count 4500, polymorphs 85%) and a very high lactose dehydrogenase (LDH) adenosine deaminase (ADA) level of 4340 U/l and 130 U/l respectively and low pH (7.2) and pleural fluid glucose (10.2 mg/dl). Pleural fluid gram stain, ZN stain and culture were negative. She was initially managed as pyogenic pleural effusion with antibiotics Inj Ceftriaxone 1 gm IV bd and Tab Azithromycin 500mg PO OD and anti-TB drugs (EHRZ). ATT was stopped 10 days later, after negative report of pleural fluid and sputum Cartridge based nucleic acid amplification test (CBNAAT) for Mycobacterium tuberculosis (MTB was received. The patient`s condition did not improve symptomatically, clinically or radiologically even after 15 days of broad-spectrum antibiotic cover and ICD continued to drain 50-100 ml of fluid every day. Since the etiology of the pleural remained unclear even after 15 days of treatment, high resolution computerized tomogram (HRCT) chest was done. which revealed large loculated cystic lesion showing calcification within left upper lobe parenchymal region suggestive of ruptured hydatid cyst. IgG antibody for echinococcus was positive- 11 NTU (NovaTech Units), thus confirming HRCT chest finding of hydatid cyst left lung. She was managed with anthelminthic therapy (Tab Albendazole 400 mg BD). Patient started showing improvement after seven days of treatment; her fever subsided and constitutional symptoms improved. Respiratory symptoms started improving after three weeks of therapy but radiological improvement was noticed one and half months of treatment (Pic 4). The patient is now asymptomatic and on regular follow-up. The anthelminthic therapy will be continued for six months.

DISCUSSION

Echinococcosis or hydatid disease is caused by larvae of the tapeworm Echinococcus. Four species are recognized. Most of the infestations in humans are caused by *E. granulosus*. *E. granulosus* causes cystic echinococcosis, and has worldwide distribution. Humans are an accidental host in cystic echinococcosis, and are generally infected by handling infected animals generally a dog.[5,6] Following ingestion of E. granulosus eggs, the metacestode cyst can be found in any organ. The liver is the most common site (60-70% of total cases), followed by the lung in 10–30% of cases. Other sites, usually spleen, kidney, orbit, heart, brain and bone, may be involved in ~10% of cases.[7-10] The right lung is more commonly involved.[11,12] Cysts in the lungs are mostly unilateral and usually solitary.[13][4] Involvement of the Lower lobe of the lungs is most common site.[14] In 20% of cases bilateral involvement can be seen, and in 30% of cases multiple cyst can be seen.[15] Unlike an adult, lung involvement is more common than liver among children, with frequencies of 64% and 28%, respectively.[16,17] Concomitant hepatic involvement is more common in adults; 79% as compared to

33% seen in children.[18]

Most patients are asymptomatic. A cough is the most common presenting symptoms in the majority of cases.[19]The cough develops in pulmonary hydatidosis either because of bronchial irritation by the growing cyst or due to rupture of the cyst directly into the bronchus. Other symptoms of pulmonary hydatid cysts include chest pain, breathlessness, expectoration, fever and haemoptysis.[20] Occasionally some patients may present with the symptoms related to compression of the surrounding structures or even more infrequently, may expectorate the contents of the cyst. Other symptoms may result from the release of antigenic material and secondary immunological reactions that develop due to cyst rupture. The cysts are markedly seen as solitary or multiple circumscribed or oval masses on imaging.[5,6]

Diagnosis:

Imaging and serology when combined usually clinch the diagnosis of CE in the most cases. A patient with lung cysts should always be investigated for associated liver cysts. The plain chest radiograph is the most valuable diagnostic tool in pulmonary hydatid disease, however, the lung cyst may be missed if the pleural effusion is gross as in our case.[21-25] If the cyst is intact Contrast Enhanced Computerized tomography (CECT) scan may demonstrate a thin enhancing rim.[26] Immunodiagnostic testing for serum antibodies or circulating antigen provide supportive evidence of pulmonary echinococcosis. An enzyme-linked immunosorbent assay (ELISA), specific IgG ELISA is the most sensitive method and the least sensitive are immunoelectrophoresis (IEP) and specific IgE ELISA. [27] Immunoblot test (IB) tests using specific echinococcal antigen may be useful to confirm the seroreactivity, but this test is not available widely.[28]

Management:

The surgical is the definitive and preferred treatment of CE. PAIR (puncture, aspiration, injection and reaspiration) can be tried in selected patients since it entails reduced hospital stay, post-operative morbidity and reduced cost of treatment. PAIR may be performed in anechoic lesions >5-cm diameter; cysts of types I and II, as classified by Gharbi et al.[29]; cysts with a regular double laminated layer; cysts of >5-cm diameter with multiple septal division (Gharbi type III) except honeycomb-like cysts; multiple cysts 5-cm diameter in different liver segments (Gharbi types I, II and III) [29]. PAIR is indicated for inoperable patients and those who refuse surgery. It has been used for cysts in the liver, abdominal cavity, spleen, kidney and bones, but PAIR is contraindicated for brain and lung cysts. [30] The various surgical options for lung cysts include lobectomy, wedge resection, pericystectomy, intact endocystectomy and capitonnage.[31]

Medical treatment is essentially an adjunct to the surgical treatment and PAIR. In inoperable cases, patients with poor surgical risk, with intraoperative spillage of hydatid fluid or secondary lung or pleural hydatidosis(32)[32] and in cases with disseminated disease, medical therapy with benzimidazoles is valuable. Albendazole (10-15 mg/kg/d) is administered in several 1-month oral doses with 14-day intervals. New data for continuous treatment are emerging from China. The optimal period of treatment ranges from 3-6 months, with no further increase in the incidence of adverse effects if this period is prolonged. Mebendazole is also administered for 3-6 months orally in dosages of 40-50 mg/kg/d. Limited data are available on the weekly use of praziquantel, an isoquinoline derivative, at a dose of 40 mg/kg/wk, especially in cases in which intraoperative spillage has occurred. Albendazole has been found ineffective in the treatment of primary liver cysts in patients who are surgical candidates.(33)

Complications

The complicated hydatid cyst is the one that has ruptured into the bronchus or pleural and infrequently the pericardial cavity. [34] Most frequent complication of pulmonary hydatid disease is rupture of cyst into the bronchus.[35]The pleural rupture is more common in young age, male sex, and basal locations of the cyst. [36] Aribas *et al*. reported pleural and pericardial complications in 29.7% (pleural 27.6% and pericardial 2.1%) of 145 patients with hydatid disease.[37] Cyst may or may not be infected.[34] Rupture of the cyst may lead to allergic pneumonitis which may involve the surrounding lung parenchyma. Allergic pneumonitis usually resolves within ten days. Secondary bacterial pneumonia needs to considered if allergic pneumonitis fails to resolve in 10-14 days.[38] Secondary pleural hydatidosis may develop in 0.9-7.4% cases as a complication of surgey or intrathoracic tube insertion. [39, 40] Pressure necrosis of the pleura and subsequent risk of pleural complications may occur due to the subpleural cysts.[41] Hydatid cysts very rarely involve pulmonary artery. The liver cyst may rupture into the inferior vena cava (IVC) and and enter into the pulmonary artery through right heart.[42] The pulmonary cysts can enter also pulmonary circulation by breaching the wall of the pulmonary vessels. The vesicles or the daughter cysts mechanically obstruct the blood flow, and there are no thrombi.[43]

Prevention:

Careful washing of fresh produce and avoiding close contact with dogs can reduce chances of infection. Prohibition of home-slaughter of sheep and proper disposal of infected viscera or offal prevents dogs from consuming infected viscera, thus disrupting the life cycle of the parasite. Vaccination is also a prospect for prevention of echinococcosis, since protective immunity develops in intermediate hosts.[44-46]

CONCLUSION

The diagnosis of primary pulmonary echinococcosis and hydatid disease of lung in patients with gross pleural effusion is likely to be missed because of its infrequent occurrence. The pleural effusion may easily be attributed to tuberculosis, empyema or malignancy since the latter are commonly encountered maladies in Indian context. However, the clinician must always keep all the possibilities open, especially in difficult to treat or non-resolving pleural effusion. Pulmonary echinococcosis can lead to life threatening anaphylactic shock if the cyst rupture.

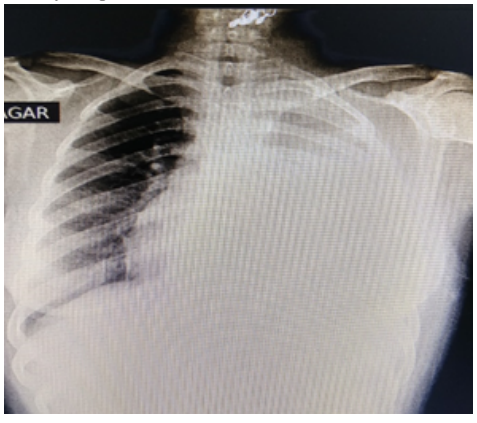

Figure 1: Chest X-Ray PA view showing left sided gross pleural effusion

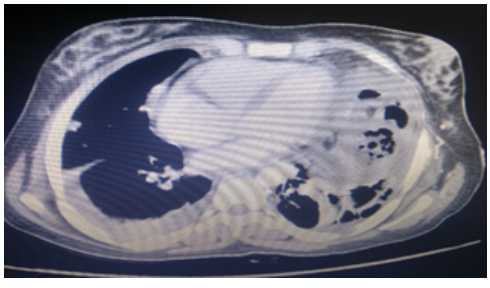

Figure 2: HRCT Thorax showing Air bubble Sign

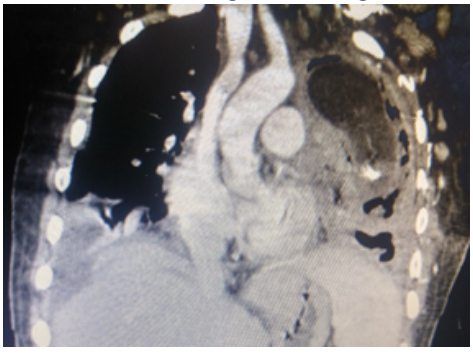

Figure 3: HRCT Thorax Cystic lesion in left upper lobe of the lung with calcification

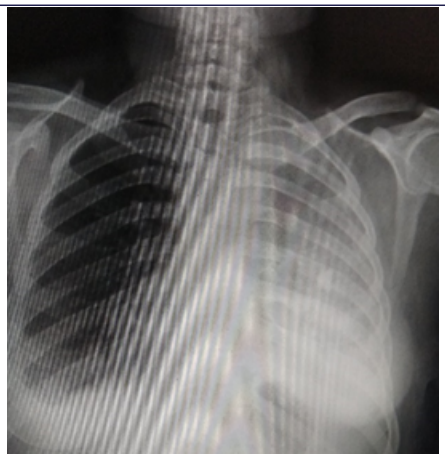

Figure 4: Chest X-Ray PA view after two cycles of Albendazole therapy

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