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Alveolar Hydatid disease in Liver: a Case Report

Dr. Sheema Sheikh, Dr Salma Gull, Dr. Ruhi Wan
Government Medical College, Srinagar, Jammu and Kashmir

Corresponding Author:
Dr Salma Gull
Government Medical College, Srinagar, Jammu and Kashmir

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ABSTRACT

Hydatid disease is caused by larval stages of cestodes of the genus Echinococcus. Alveolar Hydatid is a more aggressive form of Hydatid disease caused by Echinococcus multilocularis[1] and is transmitted to humans by infected feces of wild foxes whereas rodents act as intermediate hosts. The cestodes proliferate asexually, infiltrate to the peripheral parts of the liver and metastasize to other organs, hence this potentially fatal disease is also known as malignant hydatid disease.[2]

In India, a few case reports are the only literature available about these cases, and incidence in India as calculated based on the case reports is one per year.[3]

Keywords: alveolar hydatid, liver.

INTRODUCTION

A 32-year-old female patient presented to our hospital with complaints of mild right upper quadrant pain and weight loss with 1 year history. The patient was in good general condition, with no comorbidities. Physical examination revealed mild hepatomegaly. Hemogram showed moderate anemia, with no signs of peripheral eosinophilia. LFT revealed higher levels of Total proteins and Alkaline Phosphatse.

It was noted in the patient lived in rural Northern Kashmir which is endemic zone for cystic Echinococcosis according to a Sero-Eipdemiological survey.[4]

Abdominal ultrasonography (US) revealed a large heteroechoic lesion in right lobe measuring 9.6x10.4x12.5 cm, vol 660 cc with internal cystic areas within the lesion. Mild ascites was also noted. US was suggestive of Liver Abscess.

Thoracoabdominal computed tomography (CT) revealed a large heterogeneous calcified lesion, with no definitive enhancement on contrast images, in right lobe of liver measuring 16 × 10 x 13 cm, showing mass effect on portal vein, Inferior Vena cava, hepatic vein, without showing any infiltration. (Figure 1). There was also a cavitating lesion seen in right lower lobe lung with wall thickness of 10 mm with a surrounding ground glass opacification nodule in superior segment of RLL. (Figure 2). No other clinical or radiological findings were noted. Radiological images were suggestive of Hydatid disease/ a liver malignancy that had metastasized to the lungs/ Sclerosing hemangioma. Image guided tru cut biopsy was performed for histopathological analysis.

Serology was positive for Echinococcus IgG with 24.9 NTU.

Pathologic examination revealed germinative membranous structures of Echinococcus multilocularis in the liver parenchyma that were consistent with hepatic alveolar echinococcosis (Figure 2).

Discussion:

There are four species of Echinococcus that produce infection; E. granulosus, a more common form causes cystic echinococcosis and E.multilocularis, a more aggressive form causes alveolar echinococcosis. Echinococcus vogeli and Echinococcus oligarthrus, cause polycystic echinococcosis but have only rarely been associated with human infection.[5]

Virchow was the first person to describe the clinical and histopathological aspects of Alveolar Echinococcosis.[6]

Alveolar echinococcosis is a fatal disease and can involve multiple organs including liver, lung, cerebrum and rarely adrenal glands. Symptoms vary depending on the involved organ and degree of involvement.[5, 7] Patient may present with weight loss, abdominal pain, fever, jaundice, and hepatomegaly; chest pain, shortness of breath, cough in case of lung involvement and rarely with cerebaral involvement as epilepsy, hemiparesis, dysarthria, and cranial nerve palsies.[7, 8, 6]

Alveolar Hydatid disease is caused by larval forms Echinococcus multilocularis[9] and is transmitted to humans by infected feces of wild foxes whereas rodents act as intermediate hosts. The cestodes proliferate asexually, infiltrate to the peripheral parts of the liver and metastasize to other organs.

In the liver these form an alveolar structure, made up by several vesicles surrounded by large granulomas and sometimes show central necrosis. There are numerous small irregular cysts all <2 cm.[10]

The budding daughter vesicles on the outer side appear infiltrative. Over a period a large and heterogeneous parasitic mass is finally formed which consists of peripheral, actively proliferating sites, and centrally located necrotic tissue.

These cysts can be differentiated microscopically from cysts of E. granulosus, which are unilocular, have three layers, and do not exhibit a granulomatous reaction. Scolices and hooklets are easily found in CE, whereas rarely found in AE. The margins are not infiltrative. In contrast, E. multilocularis lacks the ectocyst and larval structures infiltrates into the host tissue causing destruction. This pattern mimics invasive carcinoma.[11]

Hydatid serology and antibody detection tests are diagnostic. Ultrasonography, computed tomography (CT), magnetic resonance (MR) are useful and complementary to each other and help in arriving at the right diagnosis.

On ultrasound, alveolar hydatid manifests as a large space occupying lesion with alternating areas of increased and decreased echogenicity. The margins are irregular with scattered foci of calcification.[12] CECT confirms the morphology of the lesion and can confirm or exclude presence of calcifications.

The lesion in our case was large, heteroechoic with internal cystic areas with computed tomography (CT) revealing a large non enhancing heterogeneous calcified lesion, with mass effect on surrounding structures.

The various differential diagnoses that can be considered are simple hepatic cyst, cystadenocarcinomas, cholangiocarcinoma, abscess, atypical hemangiomas and Cystic Echinococcus.

Immunodiagnostic methods are useful to narrow down the differentials of the imaging diagnosis. IgG ELISA has a sensitivity of 80–99% and specificity of 61.7% [9] and was found positive in our case.

Radical surgery is the only option in operable cases. It should be followed by chemotherapy for at least 2 years. Inoperable cases require continuous chemotherapy for many years. Patients should be monitored for recurrence.

In conclusion, AHD of liver is rare among Indian population and is a great mimicer. A high degree of suspicion complemented by the various diagnostic modalities can help in arriving at the right diagnosis.

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