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Case Report

Alveolar hydatid disease of the liver: A rare entity in India

Col Jasvinder Kaur Bhatia [a,*], Brig R. Ravikumar [b], Surg Cmde C.S. Naidu, VSM [c], T. Sethumadhavan [d]

[a] Associate Professor, Department of Pathology, Armed Forces Medical College, Pune 411040, India
[b] Professor and Head, Department of Radiology, Armed Forces Medical College, Pune 411040, India
[c] Professor and Head, Department of Surgery, Armed Forces Medical College, Pune 411040, India
[d] Resident, Department of Pathology, Armed Forces Medical College, Pune 411040, India

ARTICLE INFO

Article history:
Received 2 August 2015
Accepted 10 December 2015
Available online xxx

Keywords:
Alveolar
Hydatid
Liver
Histopathology
E. multilocularis

Introduction

Alveolar hydatid disease (AHD) is caused by proliferative larval stage of the fox tapeworm, *Echinococcus multilocularis*.

AHD is confined to the northern hemisphere, i.e. Europe, Russia, China, Japan and North America. The total number of AE cases in the world is 18,235 per year with China accounting for 91% of cases.[1] India, Nepal, Bhutan, and Pakistan border these endemic zones and may have a few cases. 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.[2]

Foxes and dogs are the definitive hosts and rodents are the intermediate hosts. Man gets infected accidentally and eggs develop into the metacestode stage in the liver, 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.[3]

A rare case of alveolar hydatid disease of liver is being reported. We describe the clinical, radiological and pathological features to highlight the diagnostic difficulty encountered, and successful management of the case.

Case report

A 37-year-old male patient with no known co-morbidities presented with abdominal discomfort of one-year duration. Abdominal examination revealed a mass measuring 15 cm × 10 cm × 10 cm occupying right hypochondrium, epigastric region, and part of left hypochondrium. The surface was nodular with a sharp inferior margin. General and systemic examinations were unremarkable. LFT, routine biochemical and hematological investigations were within normal limits.

NCCT abdomen showed a space occupying lesion (SOL) involving Seg I–IV of liver, suggestive of atypical hemangioma.

MRI revealed a large heterogeneous predominantly cystic mass lesion involving the entire left lobe and segments V and VIII of the right lobe of liver. The lesion was heterogeneously

* *Corresponding author*. Tel.: +91 8552825142.
E-mail address: drjkbhatia@gmail.com (J.K. Bhatia).
http://dx.doi.org/10.1016/j.mjafi.2015.12.005

Please cite this article in press as: Bhatia JK, et al. Alveolar hydatid disease of the liver: A rare entity in India, *Med J Armed Forces India*. (2016), http://dx.doi.org/10.1016/j.mjafi.2015.12.005

hypointense on T1WI and heterogeneously hyper intense on T2WI. The larger component of the lesion in the left lobe exhibited a thick (ranging from 1 to 2.5 cm) hypointense rim all around. Solid nature of the lesion in the right lobe showed multiple variable-sized cystic components interspersed within. The lesions did not contain fat, and they showed no restriction on diffusion or contrast enhancement on any of the phases on the dynamic contrast sequences (Figs. 4 and 5). Based on these findings, a differential diagnosis of Atypical hemangioma/Chronic Abscess/Atypical hydatid cyst was offered.

Our patient belonged to Maharashtra and had no history of visit to the endemic zone.

Serology was positive for *Echinococcus granulosus*.

Exploratory laparotomy revealed Seg II, III, IV and part of Seg V replaced completely by a cyst, measuring 17 cm × 13 cm × 10 cm with a thick peripheral tissue. Extended left hepatectomy was done. Left hepatectomy specimen measured 24 cm × 20 cm × 4 cm. External surface appeared nodular. Cut surface showed a large cystic cavity with central necrotic area, measuring 16 cm × 12.5 cm with a peripheral rim of compressed liver parenchyma. The cyst wall was thick and contained 50 ml of thick purulent fluid. Surrounding area showed multiple small cysts with jelly like material within them (Fig. 1).

Microscopic examination revealed a central necrotic area. Multiple sections from the peripheral area showed multiple vesicles of varying sizes and shapes, infiltrating the liver parenchyma. The vesicles showed branching, both within and outwards, giving rise to an alveolar or multilocular appearance. Each cyst was lined by an inner illdefined germinal layer and an outer laminated acellular layer. Occasional hooklets were noted; however, scolices were not found. There was no pericyst. The cysts were seen infiltrating into the surrounding liver parenchyma, with foreign body granulomas, giant cells, and moderate amount of chronic inflammatory infiltrate comprising predominantly of lymphocytes. Special stains PAS and ZN stain highlighted the characteristic laminated layer of the cyst. These morphological features were consistent with Alveolar Hydatid Disease (Figs. 2 and 3).

Postoperatively, the patient was started on Albendazole at the dose 10–15 mg/kg per day in two divided doses. His postoperative period was uneventful. He is presently asymptomatic and is being followed up regularly.

Discussion

Alveolar hydatid of the liver was first described by Virchow. He described the clinical features, detailed histopathology, and the specific infiltrative aspect of AE.[4]

Four species of *Echinococcus* produce infection; *E. granulosus* and *E. multilocularis* are the most common, causing cystic echinococcosis (CE) and alveolar echinococcosis (AE), respectively. *Echinococcus vogeli* and *Echinococcus oligarthrus*, cause polycystic echinococcosis but have only rarely been associated with human infection.[1]

AHD results from infection by the larval forms of *E. multilocularis*. Humans are accidental, intermediate hosts, infected either by direct contact with the definitive host or indirectly through contamination of food or water with parasite eggs.

After ingestion, the parasites reach the liver through lymphatics and portal system. The echinococcal metacestodes develop in the liver and form an alveolar structure, made up by several vesicles surrounded by large granulomas. Diameter of vesicles varies from less than 1 mm up to 15–20 cm. Brood capsules or protoscolices are rarely seen.[4] We did not see any protoscolices or brood capsules in our case. Lesions may be complicated by central necrosis producing a cavity or pseudocyst[4] as was seen in our case.

The budding daughter vesicles on the outer side form a progressive, infiltrating tumor-like growth. 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.

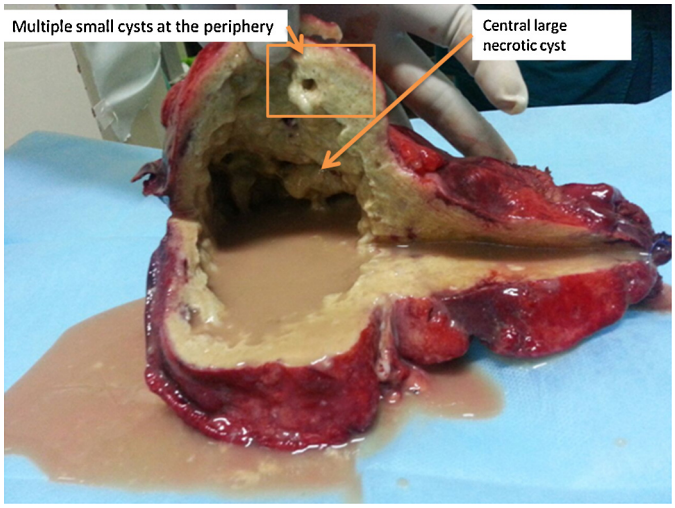

Fig. 1 – Gross specimen showing liver with a large central necrotic area with fluid and peripheral multiple cysts.

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