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Case Report

Hydatid Disease Mimicking as Hepatobiliary Malignancy

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Abstract

Hydatid disease (HD) a parasitic disease is most frequently caused by *Echinococcusgranulosus* or *Echinococcusmultilocularis* and rarely by *Echinococcusvogeli* or *Echinococcusoligarthus*. It is endemic in many parts of the world and is frequently seen in India. Liver is involved in about 74% of the cases, followed by pulmonary involvement in about 24% of cases. Secondary involvement due to dissemination can be seen in any anatomic location and hence may make differential diagnosis difficult. In our case patient presented with features suggestive of hepatobiliary malignancy clinically, radiologically and even on operating table.

Introduction

Hydatid is an important pathogenic, zoonotic and parasitic infection (acquired from animals) of humans, following ingestion of tapeworm eggs excreted in the faeces of infected dogs. Hydatid disease predominantly affects the liver and lungs. The relatively rare anatomic locations are the central nervous system, muscles, subcutaneous tissue, kidneys, bones, and body cavities. [1]

Liver hydatidosis can cause dissemination or anaphylaxis after a cyst ruptures into the peritoneum or biliary tract. Infection of the cyst can facilitate the development of liver abscesses and mechanic local complications, such as mass effect on bile ducts and vessels that can induce cholestasis, portal hypertension, and Budd-Chiari syndrome [2]. Occasionally the cyst may progressively increase in size, mimicking gross ascites or liver tumor. Often the large size of the cyst makes the diagnosis extremely difficult [3, 4]. Treatment of hydatid liver cyst has to be considered mandatory in symptomatic cysts and recommended in viable cysts because of the risk of severe complications [5].

CASE REPORT

A 25 year old female presented with itching and yellowish discoloration of eyes of 4 months duration. On examination there was icterus, hepatomegaly and palpable gallbladder. Investigations showed raised bilirubin (27.3 mg/dl) and alkaline phosphatase (ALP=281 IU/L). Ultrasonography showed dilated intrahepatic biliary radicals, echogenic area causing extrahepatic biliary obstruction and common bile duct was not traced. Magnetic resonance cholangiopancreatography (MRCP) revealed large intrahepatic mass lesion (**Fig. 01**). MRI (**Fig. 02**) showed hypointensity on T1 and mixed intensity on T2 with no significant contrast enhancement. CECT abdomen (**Fig. 03**) showed enhancing irregular mass (7.8 x 8.4 x 6.5 cm) involving segments V and VII with infiltration into junction of right and left hepatic duct. Anterior branch of portal vein and periportal lymph nodes were enlarged. Patient was shifted for surgery. Intraoperative findings were huge intrahepatic mass involving segment IVA and VIII with infiltration into segment IVB, V and VI and common hepatic duct blocking right ductal system completely. Palliation tube drainage and liver biopsy was done. Histopathology (**Fig. 04**) showed dilated spaces lined by hyaline tissue and infiltration of

hepatic tissue with eosinophills and giant cells. Features were suggestive of parasitic pathology with no evidence of neoplastic lesion.CEA levels were 1.5ng/l. Hydatid serology was suggestive (1:160) of hydatid disease hence patient was put on antihelmenthics.

After 10 days patient was again admitted with features of cholangitis due to blocked T tube. Investigations revealed bilirubin of 4.78 mg/dl and ALP of 254 IU/L. Patient was managed with ERCP with ENBDO. The ERCP (**Fig. 05**) showed smooth stricture occupying the whole common hepatic duct and intrahepatic ducts minimally dilated. The stricture could not be negotiated with Soehendra dilator; hence 5F ERCP catheter was put in left hepatic duct as an ENBDO. Patient improved clinically as well as biochemically.

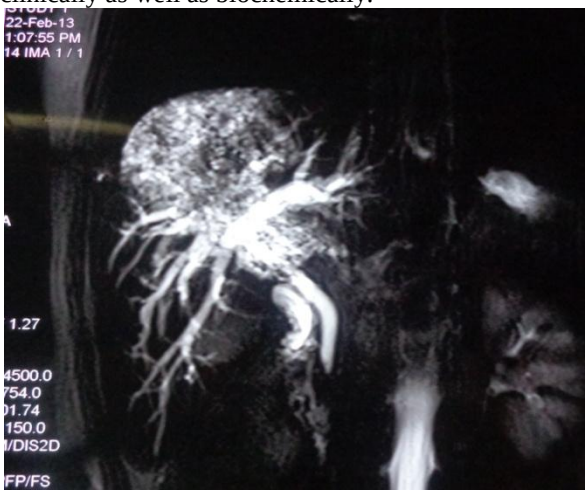

Fig. 01: MRCP Showing large Intrahepatic mass lesion with Dilated intrahepatic radicles

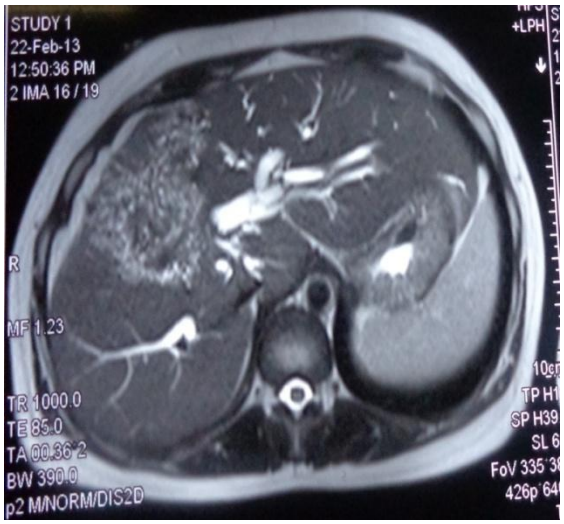

Fig. 02: MRI (T2 weighted) showing large intrahepatic mass lesion with mixed intensity

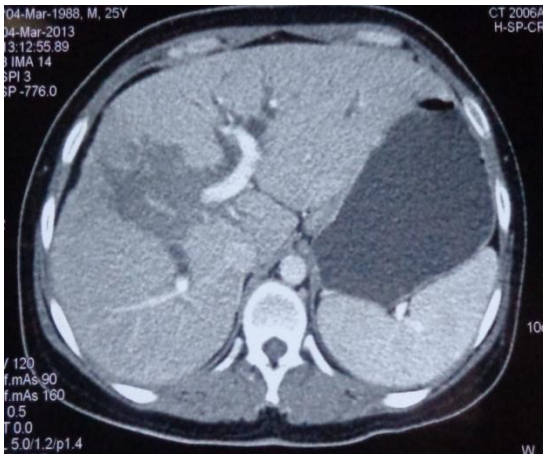

Fig. 03: CECT showing large irregular enhancing mass involving segment V & VII

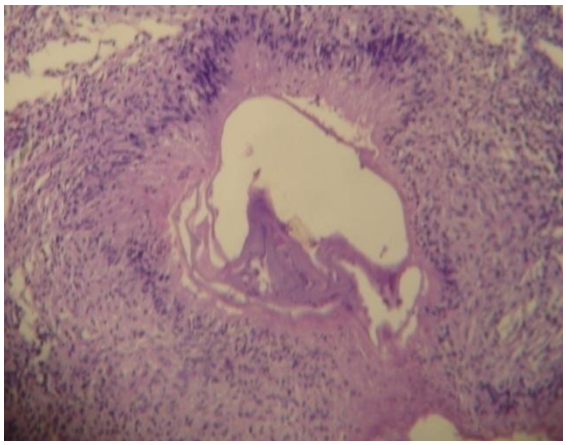

Fig. 04: Biopsy liver showing cystic lesion suggestive of hydatid disease

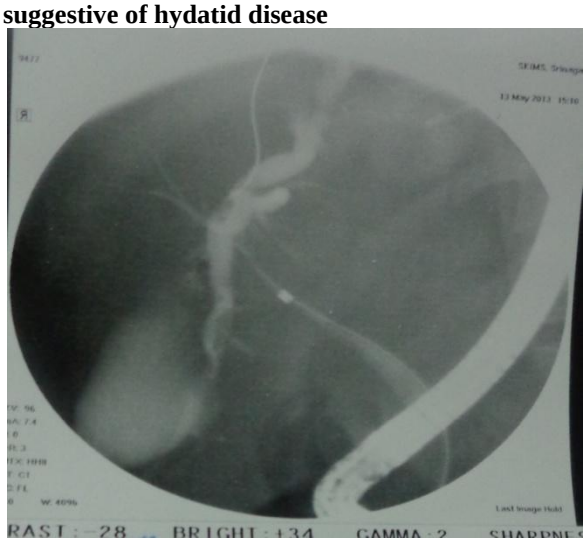

Fig. 05: ERCP showing smooth stricture of common hepatic duct

Discussion

Hydatid disease (HD) is an important health problem all over the world particularly in rural areas and / or among immigrant population from endemic countries [1]. The disease is caused by the larval stage of *Echinococcusgranulosus* (dog tapeworm), *E. multilocularis* or *E. vogeli* [6]. It occurs when humans ingest the hexacanth embryos of the dog tape worm. The definitive hosts (dogs, other canids, hyenas and cats) carry the adult tapeworms subclinically. Humans are accidental, intermediate hosts infected by coming in to contact with the definitive host or by consuming vegetables or water contaminated by ova. Intermediate hosts are initially asymptomatic; however, the growth of the larvae, which form cysts in vital organs such as the liver and lungs, can lead to illness and death. Approximately 2-3 million human cases are thought to occur worldwide. [7]

Infestation by hydatid disease in humans most commonly occurs in the liver (55–70%) followed by the lung (18–35%); both can be affected simultaneously in about 5–13% of cases [8]. Other organs like lungs, spleen, kidney, brain, bone or peritoneum may be involved.[9]A hydatic cyst consists of three layers: the outermost layer is adventitia, the intermediate is laminated membrane (endocyst), and the innermost is the germinal layer which gives rise to scolices and daughter cysts.

The presentation of human echinococcosis is protean. Depending on the localization and the size it can produce some nonspecific symptoms [10].Cyst rupture can lead to anaphylaxis which can be predicted by positivity of Casoni reaction and formation of localized or generalized secondary echinococcosis.

Differential diagnosis of a hydatid cyst, from other abdominal cystic lesions, is sometimes difficult. Cysts can often simulate the appearance of cystic tumors [11] and can be easily misdiagnosed, however, rarely appear in large dimension without previous symptoms. One should bear in mind the possibility that carcinoma may rarely have clinical, radiological, and serological features, similar to those of a HD. Singh N, *et al*. reported a case of a large cell type lung carcinoma mimicking pulmonary HD, with a positive serological test for *Echinococcusgranulosus* [12].

In 2003, the World Health Organization Informal Working Group on Echinococcosis (WHO-IWGE) proposed a standardized classification based on sonographic analysis of the morphology and structure of the hydatid cysts. Five categories are recognized [13]. Types CE2 and CE3 represent the typical hydatid cysts; types CE1 and CE5 are suggestive of hydatid cysts; type CE4, simulates a cystic tumor.

Diagnosis is most commonly made through USG or CT scan of the abdomen in which lesions appear as well-defined and circumscribed with or without internal septations. A double contrast CT scan is 90-100% accurate for diagnosing secondary echinococcosis and is superior to USG in identifying additional extra hepatic intra abdominal cysts [14]. Ultrasonography is useful in longitudinal studies, such as monitoring the response of cysts to treatment and recording cyst growth rate [15].On T1- and T2-weighted MRI images, daughter cysts manifest hypointense or isointense areas compared to the maternal matrix [16,17].MRI may have some advantages over CT scanning in the evaluation of postsurgical residual lesions, recurrences, and selected extrahepatic infections, such as cardiac infections [18].

Cystic echinococcosis is one of the few parasitic infections in which the basis for laboratory diagnosis is primarily serology [19].Immunoelectrophoresis, enzyme-linked immune sorbent assay (ELISA) has a sensitivity varying from 64% to 100% depending on the antigen used [20,21]. ELISA, latex agglutination and indirect haemagglutination (IHA) test are being carried out for the diagnosis, screening and post-operative follow up for recurrence [22].

Diagnostic cytomorphological features of HD include laminated cyst wall fragments (also called cuticula), scolices, and hooklets. Identification of hooklets is regarded pathogonomonic, and observation of laminated membranes by itself is a presumptive finding in the cytodiagnosis of HD [23].

Plain radiographic features of hydatid cyst of the lungs have been described in detail by Vessal et al. [24]. Hydatid cyst of the liver can be suspected if there is evidence of a space occupying lesion (SOL) in the liver. Generally it is believed that calcification indicates death of the cyst and scolices. Floating daughter cysts and undulated detached endocyst on the surface of the fluid level can create the "water-lilly sign". "White-lines sign" (rounded, thin walled images describing almost the full circumference of the cyst, probably due to a highly compressed pericyst) has been described in 15% of the patients. [25]

First choice of treatment HD is surgery. Medical treatment is an alternative to surgery where a surgical approach is not recommended in high risk patients, and in cases with small and multiple lesions [26].The treatment options for hydatid cyst of the liver depend on stage, localization, size, and complications of the cysts and include nonoperative and operative methods. However, pre- and postoperative 1-month courses of albendazole or 2 weeks of praziquantel should be considered in order to sterilize the cyst, decrease the chance of anaphylaxis, decrease the

tension in the cyst wall and to reduce the recurrence rate postoperatively [27]. Intra-operatively, the use of hypertonic saline or 0.5% silver nitrate solutions before opening the cavities tends to kill the daughter cysts and therefore prevent further spread or anaphylactic reaction. Percutaneous drainage of liver hydatid cysts in combination with drug therapy has been found to be efficient for CE1 and CE2 types of hydatid cysts and occasionally for CE3 type but with lower success rates [28].

Conclusion

The HD can present in varied manifestations from benign cystic lesions to malignant ones and we should always keep it as a differential diagnosis for neoplastic lesions especially of lungs and liver. It is treatable disease. Surgery is the treatment of choice in most of the cases. Drugs should be used as the sole mode of therapy only when there is disseminated hydatidosis with no chance of cure by surgery, the patient is not a good candidate for surgery, the cysts me surgically unapproachable or no adequate surgical facilities me available.

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