Hydatid Cyst of Liver Presented as Obstructive Jaundice in Pregnancy; Managed by PAIR

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Incidence of Hydatid disease in pregnancy ranges from 1 in 20,000 to 1 in 30,000. The most common site of hydatid cysts is the liver. The diagnosis of liver hydatid cysts is not difficult but the management during pregnancy is problematic. Both medical and surgical treatments are available but there is no consensus and each case has to be individualized. We present a case of liver hydatid cyst presented with obstructive jaundice during pregnancy which was managed by Puncture of the cyst under USG guidance; Aspiration of the cystic fluid, Injection of hypertonic saline, and Re-aspiration of solution without drainage (PAIR) and albendazole therapy. Very few cases of liver hydatosis were reported previously which had been managed by PAIR. (J CLIN EXP HEPATOL 2014;4:366–369)

Echinococcosis or hydatid disease is an infection of sheep, cattle, pigs, horses or rodents caused by larval stage of canine tapeworm *Echinococcus granulosus* or *Echinococcus multilocularis*. Hydatid cysts due to the former are found more commonly in sheep-rearing areas of the world namely Australia, New Zealand, Argentina, Chile, India and Mediterranean region.¹ The latter has a more restricted geographical distribution being found in Arctic and sub-arctic regions namely USA, Canada, Europe and Asia.¹ Man serves as intermediate host, being infected by ingestion of food contaminated by eggs excreted by the definitive host (canine). Hydatid cysts may remain asymptomatic for many years and may be found incidentally on imaging. These cysts can be found in any organ but liver and lung hydatids are most common.¹ Hydatid cysts during pregnancy are rare and there is no standardized recommended treatment. Both surgical and medical modalities have been tried. Hydatid disease of the liver is common in India; though it has not been reported during pregnancy from this part of India. We present the following patient as a reminder that echinococcal disease of the liver must be included in the differential diagnosis of abdominal pain, jaundice and/or fever occurring in pregnancy and to illustrate that percutaneous aspiration, re-aspiration and instil-

lation of scolicidal agent (PAIR) is safe and effective treatment for these patients.

CASE REPORT

A 33 years old multiparous 32 weeks pregnant woman referred to us from her obstetrician with complaints of severe pruritis and on and off right upper quadrant pain for last 10 days. The pain was sharp, non-radiating but exacerbated by eating and it was associated with nausea, vomiting, and low grade fever (38.0–38.5 °C.). She denied recent changes in the color of her skin, urine, or stool. There was history of viral hepatitis in the adolescent age which resolved after 4–6 weeks of conservative therapy.

There was significant history of exposure as a child to sheep and cattle, and also to dogs during childhood. She stayed in the Middle-East for 5 years. On physical exam, she had a low grade fever, mild icterus and scratch marks over extremities. Her abdominal examination revealed a palpable uterus, right upper quadrant and epigastric tenderness without peritoneal irritation. Her liver span was 12 cm. There was fullness in the epigastrium. Digital rectal exam was normal. Her lungs were clear to auscultation.

Initial laboratory results included: hemoglobin = 11 g/dl, white blood cell count of 12,600 with 76% neutrophils, 14% band cells, and 4% eosinophils; serum alanine aminotransferase (ALT) of 148 U/l; serum aspartate aminotransferase (AST) of 96 U/l; total serum bilirubin of 4.4 mg/dl with a direct serum bilirubin of 3.2 mg/dl; and serum alkaline phosphatase of 560 U/l. All viral markers were negative. Ultrasonography (USG) abdomen showed 7 × 5 × 5 cm³ cystic mass in liver segments V and VI of the right lobe which was multiseptated rosette like honeycomb cyst, suggestive of a moderately large hydatid cyst (Gharbi type II) (Figure 1) (Table 1). She had bilobar intrahepatic biliary duct dilatation, but her hepatic and

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Abbreviations: ALT: serum alanine aminotransferase; AST: serum aspartate aminotransferase; CT scan: computed tomography scan; ELISA: enzyme-linked immunosorbent assay; PAIR: Puncture of the cyst under USG guidance, Aspiration of the cystic fluid, Injection of hypertonic saline, and Re-aspiration of solution without drainage; USG: ultrasonography

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Figure 1 USG showing liver hydatid cyst and fetus.

common bile ducts were not dilated. So, she had features of obstructive jaundice. The patient was hospitalized for further evaluation.

Echinococcal serology (IgG) by the ELISA Method was performed which was positive. *Entameba histolytica* IgG serology by ELISA was negative. PAIR was done successfully and without any complications to relieve intrahepatic bile duct obstruction. Her abdominal tenderness and pruritus resolved within 48–72 h and liver function tests also return to near normal within 7–14 days. She was also put on albendazole 400 mg twice daily. She delivered a healthy female baby at 37 weeks by an uncomplicated normal vaginal delivery, with a birth weight of 2600 g. Placenta did not reveal any abnormality on gross as well as on histopathology examination. Anti-helmenthic medication was continued during the post partum period till 6 months. A CT Scan done 2 weeks post partum showed only a small ($2 \times 3 \times 1.5 \text{ cm}^3$) liver hydatid cyst. After 3 months USG was done which showed no liver hydatid

cyst. Patient was again followed up at 6 months and was doing well.

DISCUSSION

Hydatid cyst is a common health problem in Turkey and other Mediterranean countries, the Middle-East, South America, Australia, New Zealand and India. *Echinococcus granulosus* (dog-tapeworm) causes cystic Echinococcosis and *Echinococcus multilocularis* (fox-tapeworm) causes alveolar Echinococcosis are the two species of Echinococcus. The adult *Echinococcus granulosus* is 5 mm long and consists of a scolex (head) and the proglotids (2–3), which contain the eggs. These proglotids are transmitted to humans from ingestion of food contaminated with canine feces. Man is an intermediate host for *Echinococcus* while dogs, foxes and wolves are definitive hosts. These cysts are most commonly found in liver (52–77%), lungs (8.5–44%), abdominal cavity (8%), kidneys (7%), central nervous system (0.2–2.4%), and bone (1–2.5%).¹ The incidence of hydatid disease in pregnancy ranges from 1 in 20,000 to 1 in 30,000.^{2,3} The authors collected 4 cases between 1981 and 1990 with an incidence is of 1/30,000 deliveries. The diagnosis was suggested in 3 cases by the patient's country of origin and previous history of extra-pelvic hydatid disease. It is confirmed by ultrasonography.⁴ Hydatid disease during pregnancy has been reported as early as 1971. The complications of the disease are torsion, pelvic inflammation, anaphylaxis, premature delivery, dystocic dynamics, dystocic obstruction, and uterine rupture, and malignancy can occur in less than 5% of the cases No clear-cut guidelines are available on management on

Table 1 Sonographic Classification of Hydatid Cysts.

Gharbi type	WHO type	Cyst morphology
I	CE 1	Unilocular anechoic lesion with double line sign
III	CE 2	Multiseptated rosette like honeycomb cyst
II	CE 3A	Cyst with detached membranes (water-lily sign)
III	CE 3B	Cyst with daughter cysts in solid matrix
IV	CE 4	Cyst with heterogeneous hypoechoic/hyperechoic contents. No daughter cysts
V	CE 5	Solid plus calcified wall

account of paucity of reported cases.⁵ Till now there is no consensus of management of hydatid disease during pregnancy.⁶

Medical treatment consists of oral albendazole. Response to Albendazole depends on thickness of the cyst wall and the absence of calcifications. However, it cannot be used in the first trimester due to the risk of teratogenicity. Commonly reported anomalies if given in first trimester include limb defects and facial abnormalities.⁶ One case of liver hydatosis during pregnancy diagnosed at 25 weeks of gestation which was successfully managed by medical treatment only with oral albendazole was reported earlier from India.⁶ Van Vliet et al managed a 20-year-old Turkish woman with three huge echinococcus cysts of the liver who was admitted in the third trimester of pregnancy. During pregnancy she received albendazole and during vaginal delivery she received both albendazole and medication aimed at preventing anaphylactic reaction (corticosteroids).⁷

Surgery may be conservative or extensive. Surgery is still the first choice for type III cysts, type IV cysts, and cysts opening into the bile ducts or peritoneal cavity. There is often no need for therapy for type V hydatid cysts.⁸ There are several classification schemes for liver hydatid cysts based on their ultrasound appearance; the initial classification by Gharbi et al and the WHO Informal Working Group on Echinococcosis (IWGE) classification are the most commonly preferred.⁹ Hassen Gharbi classified hepatic hydatid into five types based on sonographic appearance.¹⁰ WHO classification is almost the same as Gharbi's, with Gharbi type II corresponding to CE 3A of the WHO classification, and vice versa. Surgery may be technically difficult if not impossible during pregnancy. Whether surgery should be done during pregnancy is debatable as it may precipitate preterm labor.⁶ However, the decision on the type of surgery should be individualized. In a retrospective study from Libya over 9 years hydatid disease was identified in 4 pregnant females.² In 3 of these the hydatid cyst was removed at time of caesarean section while the fourth patient had cyst was removed after delivery. One case of pelvic hydatid cyst with intra-uterine pregnancy diagnosed at the time of delivery, resulted in obstructed labor has been reported from India.¹¹ The cyst was aspirated and excised completely. Hemi-hepatectomy during pregnancy for hydatid cyst has also been reported in literature.¹² Puncture of the cyst under USG guidance; Aspiration of the cystic fluid, Injection of hypertonic saline, and Re-aspiration of solution without drainage (PAIR) is another option. A meta-analysis compared the clinical outcomes for 769 patients with hepatic hydatid cyst treated with PAIR plus albendazole or mebendazole with 952 controls undergoing surgical intervention, had shown that compared with surgery, PAIR plus chemotherapy is associated with greater clinical and parasitologic efficacy; lower rates of morbidity, mortality, and disease recurrence; and shorter

hospital stays.¹³ However, in the literature only one case of pregnancy with liver hydatid cyst was reported in which PAIR was successfully done without any complications at 15 weeks and the patient gave birth to a healthy baby at term.³ Long-term follow-up (mean follow-up time = 57.5 months) of 6 pregnant women with liver hydatid cyst also showed PAIR to be safe and effective method of treatment.¹⁴

Recurrence of hydatid disease is always a fear. Recurrent hydatid disease has also been reported during pregnancy probably due to decreased cell-mediated immunity in pregnancy.^{5,15} Albendazole is recommended for 1–3 months to prevent recurrence.¹⁶ Albendazole was added in our case as the recent. Cochrane review states that PAIR seems promising, but there is insufficient evidence to support or refute PAIR with or without benzimidazole coverage for treating patients with uncomplicated hepatic hydatid cyst.¹⁷ In the comparison made between PAIR with or without oral albendazole and oral albendazole alone, serial ultrasonography showed heterogeneous echo pattern of the cysts in 18, uniform echogenicity in 11, and disappearance in three patients. There were no deaths. Symptoms were relieved in all PAIR-treated patients (100%; $n = 20$) versus two (20%; $n = 10$) of the albendazole-treated patients ($P < 0.001$). All the cysts treated with percutaneous drainage ($n = 22$) and only two (18.2%) of those treated with oral albendazole alone showed reduction in size and changes in echopattern compatible with loss of viability ($P < 0.01$). Maximum size reduction was observed in cysts treated with a combination of percutaneous drainage and albendazole ($P < 0.05$). Complications observed with PAIR were cyst infection in two patients (10%), fever in three (15%), cyst biliary rupture in one (5%), and urticaria in two (10%). There was no mortality. Three patients (15%) who received albendazole developed reversible elevation of liver enzymes. Neither of the studies evaluated cost-effectiveness of their interventions.¹⁷

In conclusion, hydatid disease of the liver should be included in the differential diagnosis of abdominal pain, jaundice and/or fever occurring in pregnancy in a patient with very high risk of exposure to echinococcal infection, and PAIR followed by long-term oral albendazole could be a viable option in hydatid cyst of liver diagnosed during pregnancy. This is probably the first case report from India of liver hydatid cyst diagnosed during pregnancy which was managed successfully with PAIR.

CONFLICTS OF INTEREST

All authors have none to declare.

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