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Extra Hepatic Primary Hydatid Diseases in Different Anatomic Location - A Case Series with Review of Literature

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Abstract

Hydatid disease is caused by the parasite Echinococcus granulosus. Humans are accidental intermediate hosts, and the parasite commonly affects the liver and the lungs. Primary extra hepatic, extra pulmonary hydatid disease is rare. We are presenting cases of retroperitoneal extra hepatic hydatid cyst, extra hepatic hydatid cyst in calf muscle, extra hepatic extra pleural hydatid cyst. With this case series we want to create awareness about occurrence of that hydatid cyst at different anatomical locations and one of the differential diagnoses of tumours at such location.

Aims: To create awareness to consider hydatid cyst as differential diagnosis at different anatomical locations.

Settings and Design: Prospective cohort study at Karnataka Institute of Medical Science, Hubli.

Methods and Material: All cases admitted at Department of Surgery, Karnataka Institute of Medical Science, and Hubli.

Results: All the admitted cases were operated; intraoperative finding and histopathology came as features consistent with hydatid cyst. Postoperative period was uneventful and discharged with oral Albendazole.

Conclusion: Hydatid cyst can be present in any anatomical location. Presentation at times is misleading. FNAC of Hydatid cyst is contraindicated due to anaphylactic reactions once the cyst ruptures. Therefore diagnosis of hydatid cyst should be considered as a differential diagnosis of every cystic mass in any anatomical location before diagnostic invasive intervention especially when they occur in areas where the disease is endemic. Total and careful surgical excision is the gold standard therapy followed by postoperative albendazole.

Keywords: Hydatid Disease; Retroperitoneum; Extra Hepatic Echinococosis; Albendazole; Hydatidosis

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Introduction

Hydatid Disease (HD) is an infestation that caused by the larval stage of Echinococcus granulosus [1]. Echinococcosis represents a common condition in many parts of the world where livestock is raised in association with dogs [2]. Echinococeus granulosus that lives in the small intestine of candids are ingested by herbivores [3]. The main hosts are dogs that pass eggs into their faeces. Intermediate hosts, for example, sheep, goats, cattle, and human, ingest the eggs and develop cyst formation. Human is the accidental intermediate host in the life cycle of Echinococcus granulosus. Eggs ingested by intermediate hosts liberate an embryo in the duodenum, which passes through the intestinal mucosa enters either the lymphatic or the portal circulation and it is transported to the liver, lungs, and other organs [4]. The annual incidence of HD has been reported as 18 to 20 cases per 100.000 inhabitants. The liver is affected in approximately two-thirds of patients, the lungs in approximately 25.85% to 90% of patients with Echinococcus granulosus infection have single-organ involvement and more than 70% of patients have only one cyst. Surgical excision is the treatment of choice with postoperative combined treatment with Albendazole and Praziquantel to prevent recurrence [5].

Case 1

A 60 year old female presented with history of abdomen pain since 1 month insidious in onset,

located in right lower abdomen, dragging type of pain radiating to right lower limb. She also complaints of mass per abdomen since 1 month insidious onset, non-progressive, in right side of lower abdomen. Patient denied any change in bowel habits, vomiting, weight loss or fever. On examination firm mass measuring 35 cm × 20 cm present in right iliac fossa, right lumbar and right hypochondrium with well-defined medial and lateral border. Superior border merges with right costal margin, inferior border is not well made out, medially extends up to midline and laterally up to lateral abdominal wall. Mass is bimanually palpable, non-tender, not mobile. Per vagina examination revealed bulge in right lateral fornix. USG abdomen revealed cystic mass with multiple thick septations arising from pelvis. MRI abdomen and pelvis suggested a large predominantly cystic lesion measuring 22 cm × 16 cm seen in right adnexa extending into lumbar region and hypochondriac area with multiple internal septae suggestive of large multi-loculated cystic lesion probably arising from right ovary (Figure 1).

On exploration, cystic lesion measuring 30 cm × 35 cm present arising from retroperitoneum attached to posterior abdominal wall and iliac vessels containing daughter cysts (Figure 2 and 3). Liver, ovaries, kidney normal. Subtotal excision of cyst was done. The ectocyst layer adherent to retroperitoneal structure could not be excised. Abdomen closed with drain tube. The postoperative period was uneventful. Histopathology report showed features consistent with hydatid cyst.

Case 2

Thirty four years old female presented with chest wall swelling since 1 year. She was asymptomatic. On contrast CT chest, 3 cm × 4 cm cystic lesion inside the chest wall outside the lung on left side. Lung fields normal (Figure 5- blue arrow). On exploration extra pleural cystic lesion was excised measuring 3 cm × 4 cm. Histopathology report showed features consistent with hydatid cyst. Postoperative period was uneventful.

Case 3

Thirty years old male presented with swelling in right lower limb since 1 year. He was asymptomatic. On examination 12 cm × 5 cm intra-muscular swelling, firm in consistency was present in right calf region. USG-calf region revealed cystic swelling. Excision revealed cystic swelling containing daughter cysts (Figure 4). Histopathology report showed features consistent with hydatid cyst. Postoperative period was uneventful. All three patients were started on albendazole 400 mg twice daily for 4 weeks with gap of 2 weeks for 6 months.

Discussion

Hydatid disease is an endemic widely distributed parasitosis with potential presentation worldwide [6]. Hydatid disease has two clinical categories: cystic hydatidosis caused by Echinococcus granulosus and alveolar hydatidosis caused by Echinococcus multilocularis. Infection begins with the ingestion of tapeworm eggs, which in the human intestine hatch into embryos that penetrate the small bowel mucosa, enter venules and travel via portal circulation to the liver. Hydatid cysts most often develop in the liver. However when embryos pass through this first filter, the second most frequent location is the lung. Hydatid cysts can occur anywhere in the body [7]. In terms of frequency, renal disease follows hepatic, pulmonary and peritoneal involvement [8]. However, in clinical practice disease involving the urinary tract is uncommon [9]. From a clinical point of view, the only pathognomonic sign of hydatid disease affecting the urinary

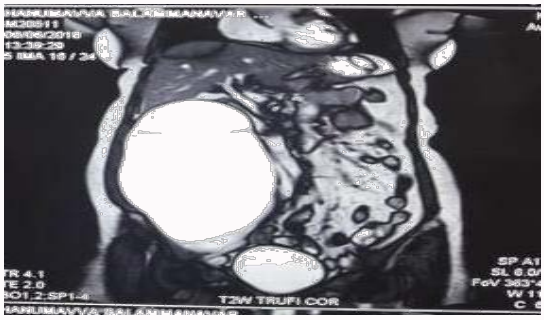

Figure 1: MRI showing large multiloculated cystic lesion arising from probably right ovary.

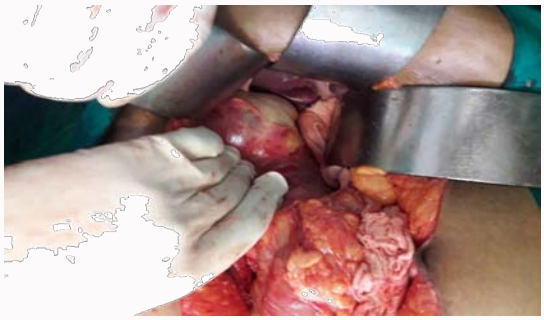

Figure 2: Cystic lesion measuring 30 cm × 35 cm present arising from retroperitoneum attached to posterior abdominal wall and iliac vessels.

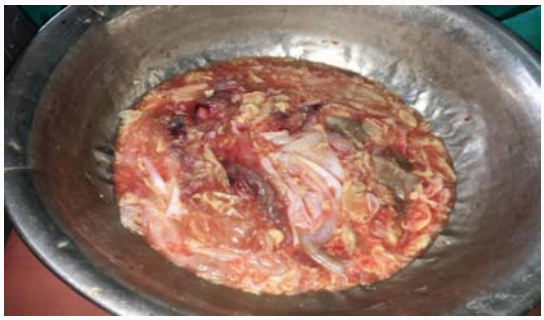

Figure 3: Excised hydatid cyst of 30 cm × 35 cm, ruptured containing daughter cysts.

tract is hydatiduria or the passage of grape-like material in the urine, sometimes accompanied by renal colic [10]. The adult worm lives in the intestines of animals like dogs. Sheep, cattle and humans are intermediate host. The overall frequency of peritoneal echinococcosis is approximately 13% of all cases. Cysts in the peritoneal cavity are mainly the result of the spontaneous or traumatic rupture of concomitant hepatic cysts or surgical inoculation of a hepatic cyst [11]. Most cases of hydatid cysts occur in the liver followed by the lungs. The muscle is a rare location for hydatid cysts (0.7% to 0.9%), even in endemic countries. Cerebral HC is extremely rare, accounting for only 2% of all intracranial masses, even in countries where this disease is endemic [12]. Extra hepatic abdominal hydatidosis accounts for about 1.6% of abdominal hydatidosis [13]. The isolated retroperitoneal location of hydatid cysts has been reported to be a very seldom occurrence as many as 1.1% of the newly diagnosed cases could manifest as retroperitoneal isolated or retrovesical lesions, which corresponds to a rate of 0.1/100,000 per year [6]. Primary isolated extra hepatic hydatid disease is mostly seen within

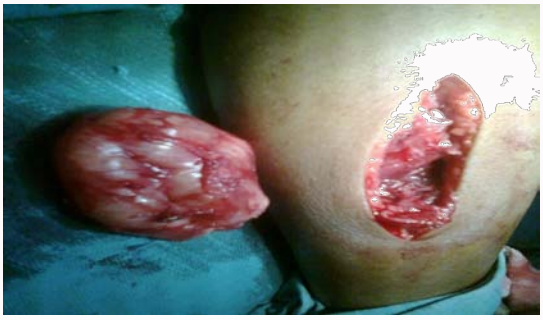

Figure 4: Excised hydatid cyst from calf.

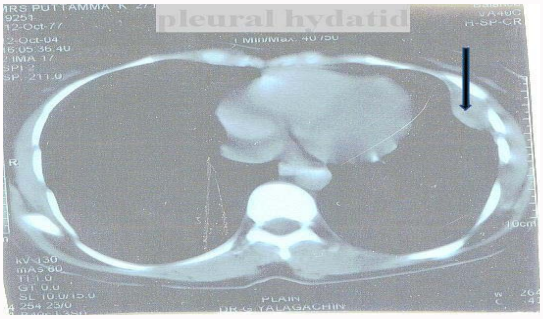

Figure 5: Plain CT showing extra pleural Hydatid Cyst.

the abdomen with an incidence of 6% to 11% [14].The psoas muscle hydatid accounts for only 1% to 3% of all cases. Retroperitoneal involvement has always been considered to be secondary to rupture or spillage from liver hydatid cysts. The growth rate of the cysts themselves is about 1 cm per year. Isolated retroperitoneal hydatid cysts are formed either through the bypassing of the protoscoleces through the liver and lungs haematogenous, or by a lymphatic passage through the gastrointestinal system [15]. Dew and Waddel had favoured airborne transmission and direct implantation of the embryo in the bronchial mucosa as another possible mode of entry. This raises the possibility of an embryo of the parasite entering a venule after penetrating the bronchial mucosa and reaching the left side of the heart to involve other sites and thus bypassing the lung. But this remains largely theoretical and needs to be proved [16]. According to the classical theory of Deve, fissuring or rupture of a primary hepatic, splenic or mesenteric cyst would seed its contents in the abdominal cavity [17]. The primary cyst might then heal and even disappear, leaving behind a scar that could be overlooked by any diagnostic modality. The pouch of Douglas would then be the preferred site for development of a secondary cyst in the pelvis, initially intraperitoneal and later sub peritoneal. From an anatomical point of view it has been proposed that cysts centrally located in the pelvis that is close to the boundaries of the bladder and rectum, are strictly peritoneal, while those with a further posterolateral location (retrovesical) are considered retroperitoneal [18]. Hydatid disease in the muscles progresses slowly and is hardly ever life threatening. When hydatid cysts are located in the muscle, diagnosis may be difficult and, as in this scenario, most cases are symptomatic and diagnosed either incidentally or when the cyst enlarges and results in compression on adjacent organs [15]. FNAC needs to be avoided whenever Hydatid is suspected. Theoretically, it can occur at any site except teeth, hair, and nails [16]. Retroperitoneal hydatid cysts are usually asymptomatic and cause symptoms from pressure or because of complications, like infection or rupture. Correct pre-operative diagnosis is difficult to make unless circumlinear calcification is seen in the plain X-ray of the abdomen [2]. Serology should be used to confirm a tentative diagnosis of hydatid disease [14]. The fluids in the cyst are crystal and clear. It is transudate of serum contains protein and may be high antigenicity [19]. As far for diagnosis of (HC) according serology tests have little role, due to its high false positive or false negative rate up to 15% to 20% [2]. Both ultrasounds and CT scans are sensitive for detecting hydatid cysts. Ultrasounds are a non-invasive, inexpensive and repeatable imaging modality widely used and accepted in the diagnosis of this disease. CT scan sensitivity ranges from 90% to 97%. Surgery is the mainstay of treatment of hydatid cysts. Excision of a cyst in the lumbar retroperitoneum adhered to the lateral abdominal wall may require reconstruction with a prolene mesh. When a cyst is adherent to neighboring structures that need to be preserved (rectum and major vessels) partial cyst excision or pericystectomy, excision of the cyst leaving areas in contact with the organs, is an appropriate option as well [20,21]. The aim of surgical therapy is complete removal of the cyst without contamination of the field. Dissection between the cyst areas and laminated external membrane implies a high risk of rupture and spilling of cyst content. Removal of germinal epithelium and fluid with scoleces may cause hydatid dissemination and allergic manifestations, even anaphylactic shock. Ideally then, total cyst excision or pericystectomy should be performed [21,22]. Total cystectomy is considered to be the gold standard in terms of surgical approach. However, the cysts in the retroperitoneal space can be associated with dense adhesions and hence a partial cystectomy may be the treatment of choice to avoid injuring neighbouring structures. Controlled evacuation of the cyst content may be necessary when the hydatid fluid is under high tension. Prophylactic measures, such as irrigation with a scolicidal solution and albendazole systemic chemotherapy in cases of cyst rupture, are strongly recommended. We prefer to use hypertonic 30% saline solution for local irrigation since it seems to be effective and does not harm surrounding tissues. Aspiration of the cyst has been considered an option to standard surgical therapy for elderly patients and an alternative to partial cyst excision or pericystectomy in patients with unrespectable disease in the liver [23]. Postoperative anthelminthic therapy has been shown to prevent recurrence of the cysts. The treatment consists of administration of albendazole for 3 to 6 months [15].

Conclusion

Hydatid cyst can be present in any anatomical location. Presentation at times is misleading. FNAC of Hydatid cyst is contraindicated due to anaphylactic reactions once the cyst ruptures. Therefore diagnosis of hydatid cyst should be considered as a differential diagnosis of every cystic mass in any anatomical location before diagnostic invasive intervention especially when they occur in areas where the disease is endemic. Total and careful surgical excision is the gold standard therapy combined with albendazole postoperatively for 3 to 6 months with 2 weeks of drug free interval in between.

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