Primary Adrenal Hydatid Cyst: A Case Report

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Abstract
A 38 year old woman who presented with right flank pain of unclear aetiology. Imaging studies demonstrated a cystic mass in the right upper pole of the kidney causing the ureteropelvic junction obstruction with a with a significant proximal urinary tract dilatation. She underwent a careful excision of the cyst and the pathological examination confirmed a hydatid cyst (HC) of the right adrenal gland. We describe the radiological and perioperative findings of this rare case. We have also reviewed the recent medical literature of similar case reports.

Key words: adrenal gland, hydatid disease, echinococcosis, hydatid cyst, ureteropelvic junction.

Introduction
Hydatid disease is an endemic illness in many countries, and it poses an important public health problem. Hydatid disease is a parasitic disease caused by the larval stage of Echinococcus granulosus and E. alveolaris. Humans may become intermediate hosts through contact with a definitive host (usually a domesticated dog) or ingestion of contaminated water or vegetables (1). The adult worm lives in the small intestine of the primary host. Ova are passed in feces and ingested by the intermediate host, which may include humans. Hatched embryos migrate through the intestinal mucosa, enter venules and lymphatics, and reach the liver. If embryos bypass the liver, they can reach the lungs via the inferior vena cava and heart. Embryos may reach any other organ or tissue in the body as the adrenal glands, via the systemic circulation, (2) where it develops into small HC (1,3). Infection with Echinococcus larvae generally results in the formation of hydatid cysts in the liver and lungs (2,4), although the infection may affect almost any organ in the body including brain, muscle, kidney, heart and even endocrine glands. Unusual sites for this disease may cause diagnostic problems making correct preoperative diagnosis difficult. The localization of HC in the adrenal gland is extremely rare (5). Only 17 cases have been described in the literature (2,6). With the wider application of ultrasonography (US) and computed tomography (CT) more of these cases are now being reported. Herein, we report a case of an adrenal hydatid cyst causing uretero-pelvic junction obstruction in a patient with right flank pain.
Case Report
A 38-year-old woman was referred to us for right flank pain. She denied any hematuria or lower urinary tract symptoms. Her medical history was not significant. Physical examination was unremarkable. Laboratory data included a complete blood cell count, electrolytes, eosinophil count, serum biochemistry and urinalysis were within normal limits. A plain abdominal X-ray showed a 3-cm round opacity in the right upper region of the abdomen, with curvilinear calcifications (Fig. 1). Abdominal Ultrasound showed a calcified heterogeneous cystic mass between the superior pole of right kidney and liver in the retroperitoneal area with pelvic and caliceal dilatation. The intravenous pyelography showed the right ureteropelvic junction obstruction with a significant proximal urinary tract dilatation (Fig. 2).

Abdominal computed tomography scan confirmed the ureteropelvic junction obstruction secondary to a solitary cystic mass, located in the upper pole of the kidney. It showed peripheral calcifications, wall thickening, and central partial fluid image with no enhancement after intravenous contrast media injection (Fig. 3, 4). It was 50 x 70 x 50 mm in dimensions. Radiological findings were suggestive of a primary adrenal echinococcosis. No other

**Figure 1:** Abdominal X-ray: a small round opacity in the right upper region of the abdomen, with curvilinear calcifications (projection on the adrenal gland area) (arrow).

**Figure 2:** IVP: a wall calcified upper renal mass associated with ureteropelvic junction obstruction of the right kidney (arrow).

**Figure 3, 4:** CT of the abdominal cavity: a cyst in the right retroperitoneal space, above the upper pole of the right kidney with curvilinear calcifications of the wall (arrow).
Hydatidosis is a parasitic disease (2), endemic in many parts of the world. It is most commonly found in the Middle East, north of Africa, Australia, Iceland and South America, and also remain endemic Tunisia (4). Hydatid disease is capable of involving almost any organ (3). In a series of 275 patients (5), the sites of involvement (in decreasing order of frequency) included the liver (74.8%), lungs (48.3%), peritoneum, kidney, brain, mediastinum, heart, bone, soft tissues, spinal cord, spleen, pleura, adrenal glands, bladder, ovary, scrotum, and thyroid gland. Patients may present with disseminated disease.

Primary hydatid disease of the adrenal gland is a rare event (4); as this entity has been reported in only 7% of all adrenal cysts and constitutes less than 1% of all cases (5). Adrenal hydatid cysts usually form in association with generalized echinococcosis. Adrenal glands get seeded with infection due to a secondary spread resulting from spontaneous or intraoperative rupture of a primary cyst. On the other hand primary hydatid cysts of the adrenal gland are extremely rare. Out of the 9 adrenal cysts reported by Akçay MN et al, only five of them were primary (5). As the growth of HC is slow, patients generally remain asymptomatic and the diagnosis is made incidentally (5). When symptoms are present, most are related to local visceral compression. The most important clinical features are: flank pain (4,5) and gastrointestinal symptoms (bloating, fullness, nausea, vomiting, constipation, and anorexia) (3). Physical examination is usually normal (4) but occasionally a mass may be found (5,7). The complications of an adrenal gland HC include rupture in the peritoneum or retroperitoneum, local infection, fistula, hemorrhage, or compression of adjacent tissues (5,7). Rarely, an adrenal HC may cause arterial hypertension which has been described as the Goldblatt phenomenon (4,7). Serological studies like ELISA or serum IgE levels may assist in diagnosis (2), but lack both sensitivity and specificity (8). Radiological imaging techniques provide an important aid in the diagnosis and follow up of echinococcosis (5,7). Imaging findings depend on the stage of cyst growth and associated complications (9). In 2003, the World Health Organization proposed the following classification based on ultrasound(US) features of the cysts: type 1 is a well-defined, anechoic lesion; type 2 demonstrates separation of the membrane (the “water lily” sign formed by the undulating membrane); type 3 is characterized by the presence of septa and intraluminal daughter cysts. Type 4 is a nonspecific solid mass. Type 5 is a solid mass with a calcified capsule (9). The case described in this report is a type 5 cyst. A CAT scan is superior to US in demonstrating cyst wall calcification, infection, and peritoneal seeding. It also better delineates the relationship of the cyst with adjacent organs (2). Magnetic resonance (MR) imaging shows the characteristic low signal intensity rim of the hydatid cyst on T2-weighted images (10). Both MR and CT images are able to show the exact anatomic extent, size, volume and position of the mass, the number of cysts, the relationship to other organs and possible complications. Few reports suggest MRI to be more specific than CT (10). Calcification is seen on radiography in 20%-30% of hydatid cysts as a peripheral curvilinear or ring-like pattern. Complete calcification of the cysts is suggestive of the parasite death(6). The presence of calcifications in the

Figure 5: Intraoperative view of the cyst (circle) during the dissection of the adherences with the liver and the adrenal gland (arrow).
adrenal mass very much supports a diagnosis of HC (8). The preoperative diagnosis of HC is highly suggested by US and CT scan findings (4) but the hydatid nature of the cyst can only be confirmed pathologically. The differential diagnosis of an adrenal cyst include an endothelial cyst, a pseudocyst due to infarction or hemorrhage, a cystic neoplasm like lymphangioma, a post-traumatic cyst (5,8), a cystic phaeochromocytoma (4), an abscess and other congenital or acquired cysts. Just as in hepatic and pulmonary hydatidosis, surgery remains the mainstay for the treatment of HC (5,7). Surgery with either partial or total excision of the cyst, with or without preservation of the adrenal gland, is the treatment of choice (4).

The rapid development of laparoscopic techniques has encouraged surgeons to replicate principles of conventional hydatid surgery using a minimally invasive approach. Among the several suggested surgical methods, the simple resection of the cyst is considered the best option as it allows preservation of the gland. As reported by Horchani and Dionigi (2, 4) open or laparoscopic surgery may be indicated. Secondary infestation can be prevented by intraoperative injection of concentrated saline into the cyst, which causes death of the parasites.

**Conclusion**

Primary hydatid cysts in the adrenal gland are a rare occurrence. Imaging modalities such as US, CT and/or MRI may be helpful in suggesting a diagnosis but confirmation requires a biopsy. Radiologists and surgeons should consider HD in the differential diagnosis of cystic lesions especially in patients from endemic regions.

**References**