Hydatid cyst - *An unusual presentation*

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Hydatid disease, or echinococcosis, is common worldwide, especially in the south coast of the Mediterranean Sea. It commonly involves the liver (73%), lung (14%), peritoneum (12%), kidney (6%), spleen (4%), and less commonly the spinal cord, brain, bladder, thyroid, prostate, heart, eye and bone. Hydatid cyst of the adrenal gland is a rare entity (0.5%), with only 24 patients being described in literature. We encountered an unusual presentation, where an adrenal hydatid cyst coexisted with liver hydatid disease.

An 11-year-old girl presented to the surgical outpatient department with pain in the right hypochondrium and recurrent vomiting of 6 months duration. No significant past medical history was forthcoming. Physical examination revealed a thin-built patient with mild anemia. Systemic examination showed tenderness in the right hypochondrium. Laboratory studies revealed normochromic normocytic anemia with elevated eosinophil count. No serological tests were carried out for hydatid disease due to its endemicity in Libya. Chest x-ray was within normal limits. The anemia was attributed to malnutrition. Ultrasound examination showed a well-defined, rounded, thin-walled subcapsular cyst in segment VI of the liver. 'Floating membranes' were seen within the lesion. In addition, there was a similar cyst in the region of the right adrenal gland with medium-level internal echoes. This led to a suspicion of hydatid disease of the liver and right adrenal (**Figure 1**). On CT scan, a unilocular thin-walled round cyst was seen in segment VI of liver. The lesion had partial rim calcification. A similar lesion was also seen in the right adrenal gland, distinct from the right kidney. Endocystectomy was carried out for both liver and right adrenal cysts, and histopathological examination confirmed them to be hydatid cysts.

Hydatid disease is caused by the parasite *Echinococcus granulosus (E. granulosus)*, and less frequently, by *Echinococcus multilocularis*. The *E.granulosus* has a worldwide distribution. Infection by *E. granulosus* is common in sheep- and cattle-rearing countries including Australasia and countries around the Mediterranean Sea. The liver and lungs are the most common site of involvement.[1] The *E. granulosus* is a tapeworm, 3-6 mm in length, which lives in the intestine of the definitive host, usually the dog. Its eggs are excreted in the dog's feces and swallowed by the intermediate hosts – sheep, cattle, goats, or accidentally by humans. The eggs hatch in the duodenum, producing larvae that penetrate through the mucosa into the mesenteric venules to reach the liver through the portal venous system. Most of the larvae are filtered in the capillaries of the liver and the lungs while the kidneys, spleen, central nervous system, and bone may become secondarily involved. The surviving larvae form slow-growing cysts. The cyst wall consists of 3 layers: 1. Endocyst (parasitic component of capsule), which is the inner germinative layer giving rise to brood capsules that may remain attached to the cyst wall harboring up to 400,000 scolices, or may detach and form sediment in cyst fluid (hydatid sand). 2. The host forms a dense connective tissue capsule around the cyst called pericyst. It is a highly vascularized adventitial layer of 0.5-4 mm. 3. Ectocyst, the external cyst membrane that is approximately 1 mm thick, which may calcify. It is secreted by the parasite. When hydatid cysts within the organs of an herbivore are eaten, the scolices attach to the intestine and grow to adult tapeworm, thus completing the life cycle. Unusual sites of infection have been reported in the literature, such as the uterine cervix, subcutaneous tissues, orbit, fallopian tubes, ovaries, breast and pancreas. The patients usually are asymptomatic or may present with pain. Presentation with recurrent jaundice and biliary colic is seen due to transient obstruction by membrane fragments, and daughter cysts expelled into the biliary tree or by 'pressure effect' on the biliary ducts. Blood eosinophilia (20-50%) is the most common laboratory finding. Urticaria and anaphylaxis may be seen following cyst rupture. Serological tests have high sensitivity and specificity with good predictive value. The Casoni intradermal test has 60% sensitivity and may be falsely positive. Complement fixation double diffusion has 65% sensitivity only.

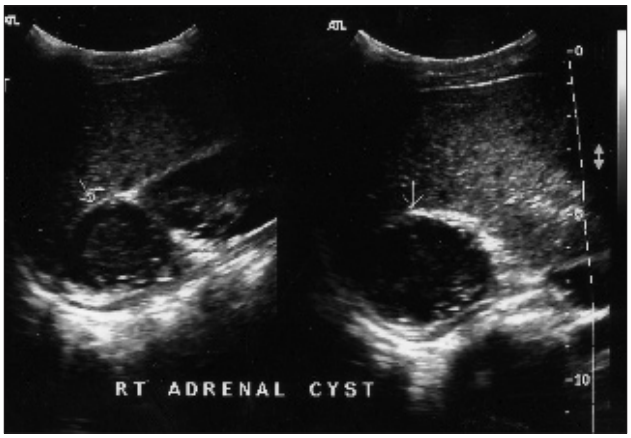

Figure 1 - Baseline ultrasonogram shows thin-walled round cyst in the right adrenal gland, with 'floating membranes' within.

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Immunoelectrophoresis is the most specific serological test today. Indirect hemagglutination also has high (85%) sensitivity. The adrenal hydatid cyst may present as a solitary renal tumor.[2] The plain films may show crescentic/curvilinear calcification. Ultrasound is the most important diagnostic tool especially for abdomino-pelvic and orbital cysts. It may demonstrate 'racemose' appearance of the cyst or the 'water-lily' sign or 'double-wall' sign. The CT scan shows well-demarcated low-density cystic masses of fluid attenuation ± internal septations with a calcific wall.[3] There may be mild enhancement of the cyst wall and septations after contrast administration. Percutaneous aspiration of the cyst contents has been reported in the literature, but the practice is discouraged due to greater risk of anaphylaxis as a result of cyst fluid spillage, although rare. Surgery is the conventional treatment in echinococcal disease.[4] Simple resection of the cyst is the best treatment, especially in cases of adrenal hydatid, as it preserves the glandular tissue.[5] Ultrasound has been used to monitor the course of medical therapy in patients with abdominal hydatid disease.

In conclusion, hydatid cyst is a common disease affecting the liver and lungs. It is very rarely seen in the adrenal gland, where the diagnosis is difficult. However, with the advancements in ultrasound and CT scan technology, more adrenal cysts are detected incidentally. The interest of this case lies in the extreme rarity of the disease in the adrenal gland. Surgery remains the treatment of choice.

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