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Case Report

Primary adrenal hydatid cyst: A rare entity and literature review ☆,☆☆

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ABSTRACT

Hydatid cyst is a zoonotic disease rarely involving the adrenal glands, even in areas where the disease is endemic. The purpose of this article is to present the clinical and imaging findings of adrenal hydatid cysts, along with a comprehensive literature review of related research. Here we report a rare case of an 18-year-old male with a right adrenal cystic lesion showing characteristic features of a hydatid cyst on ultrasonography and computed tomography, which turned out to be an adrenal hydatid cyst after surgical excision. In conclusion, when a cystic lesion is detected in the suprarenal regions, adrenal hydatid cysts should be considered in differential diagnosis, especially in patients who live in or have come from endemic areas. Medical imaging can play a crucial role in diagnosing, managing, and post-surgical assessment of this condition.

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Background

Hydatid cysts are a parasitic disease caused by a tapeworm of the echinococcus species from the Taeniidae family. Humans are most frequently infected from 2 main types of this disease, cystic echinococcosis caused by the echinococcus granulosus and vesicular (alveolar) echinococcosis caused by echinococcus multilocularis. This disease has 2 other types that are not very common; multicystic echinococcosis and monocystic echinococcosis. Cystic echinococcosis is the most common type of hydatid disease and accounts for 95% of all echinococcal diseases in humans which is known as hydatid cyst disease [1,6].

The life cycle of the parasite usually takes place between dogs and other carnivores, sheep, goats, cows, camels, pigs, and other wild herbivores. The eggs of the echinococcus parasite are present in the feces of infected carnivorous animals such as dogs, wolves, jackals, hyenas, and foxes. These animals become infected with the parasite when they eat the

Abbreviations: CE, cystic echinococcosis; CECT, contrast enhanced computed tomography; CT, computed tomography; EG, echinococcus granulosus; ELISA, enzyme-linked immunosorbent assay; HU, Hounsfield unit; MRI, magnetic resonance imaging; WHO, world health organization; IgG, immunoglobulin G.

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guts and intestines of herbivores infected with hydatid disease [4,5].

Although humans have no active role in the biological cycle of the disease, they can get the disease randomly by eating water, food, and vegetables contaminated with the echinococcus eggs or rarely through contact with animals infected with this parasite [4]. After ingestion, the eggs disintegrate in the digestive tract and the larvae are released, which then pass through the hepatic circulatory system and reach the lungs [1]. Larvae that are not trapped by the lung filtration system can be transported through the bloodstream to the spleen, kidneys, adrenal gland, brain, heart, bones, muscle, and other organs of the human body [1,2,12]. Occasionally, larvae can enter the systemic circulation via the lymphatic system without entering the portal system [2].

The echinococcus parasite creates cystic lesions in the different organs of the human body. The most common site for hydatid disease is the liver and the lung [6–8]. The adrenal glands are an unusual location for hydatid cysts, but they should be considered in the differential diagnosis of adrenal cystic lesions, especially in patients living in endemic areas. Adrenal hydatid cysts are usually detected incidentally, which may develop as a primary (isolated) or secondary (associated with other organs) disease. The epidemiology of hydatid cysts is closely linked to regions where sheep and cattle farming is prevalent, as these animals serve as intermediate hosts for the echinococcus parasite.

Adrenal hydatid cysts may remain asymptomatic for years or may be detected incidentally during radiological examinations of the abdomen, undertaken for other reasons. Symptoms and signs depend on the size of the cysts. When the cysts get enlarged and complicated, a person may experience abdominal pain or the feeling of a lump in the abdomen, especially in the flank regions. Pheochromocytoma-like symptoms such as arterial hypertension, headache, and palpitations can also be seen [3,6,9,12].

The diagnosis of adrenal hydatid cysts is based on imaging findings and serology, but serological examination has only a confirmatory role. Routine laboratory tests are usually nonspecific [17].

Case presentation

An 18-year-old man was referred to our hospital for nonspecific right upper quadrant pain with back radiation, for several weeks. Recently, the pain has been increased and localized to the right side flank area. A history of contact with sheep and dogs with work-related hazards was present. On clinical examination, his abdomen showed tenderness in the right flank area and his vital signs were stable (blood pressure 120/79 mmHg, pulse rate 71 beats per minute, respiratory rate 16 beats per minute). Hydatid serology of IgG was positive, but other laboratory findings including routine blood examinations such as complete blood count, and blood biochemistry tests (blood glucose, electrolytes, liver function tests, and renal function tests) were unremarkable.

Abdominal ultrasonography revealed a large well-defined double-walled solitary cystic lesion with no internal septation or solid component, measuring (6 × 4.8 × 4.2) cm located in the right supra renal region (Fig. 1). The mass shows no vascularity on Color Doppler ultrasound.

Subsequently, abdominopelvic contrast-enhanced computed tomography (CECT) was performed, revealing a well-circumscribed, unilocular hypodense right adrenal cystic mass, measuring (6.2 × 5.1 × 4.5) cm with a typical “double-wall sign” (Figs. 2A-C). The lesion shows no internal septation or solid component, no frank peripheral calcifications, or enhanced internal component.

The diagnosis of adrenal hydatid cyst in the present case was made based on the typical ultrasonography and CT findings with a positive hydatid serology (ELISA-enzyme-linked immunosorbent assay) test.

The patient was started on albendazole with a therapeutic dose of 400 mg twice daily 2 weeks before surgery. Given the large-sized cyst, location, and associated symptoms, he underwent a right adrenalectomy through a right anterior subcostal trans-peritoneal approach. The postoperative histopathological examination of the specimen confirmed the diagnosis of a hydatid adrenal disease. The postoperative period was uneventful without any complications and the patient was discharged on the third postoperative day. He didn’t have any evidence of recurrence during the follow-up period.

Discussion

Hydatid cysts can involve almost all the organs and tissues of the human body; however, the liver (45%-77%) and the lungs (10%-50%) are the 2 organs that are most commonly involved, followed in frequency by the kidney (3%), bone (1.4%) and brain (1.2%). Other sites such as the heart, spleen, pancreas, and muscles are very rarely affected [2,6–8,18]. The adrenal gland is an extremely rare location for hydatid disease in humans (0.5%). It occurs in all age groups, but it seems to be more common in people aged 50 to 60 years and slightly more common in women [2,9–12]. Adrenal hydatid cysts are usually asymptomatic and are often found incidentally on radiological examination, performed for other reasons. They are rarely complicated and become symptomatic due to pressure effects. The most common symptom is pain, caused by inflammation of adjacent tissues by cysts [10]. The most serious complication of this disease is cyst rupture, which may cause anaphylaxis and bleeding [9]. The large adrenal cyst can put pressure on the renal artery, which can lead to arterial hypertension (known as the Gold Blatt phenomenon) [3,6,9–11]. Adrenal hydatid cysts can also put pressure on the adrenal medulla and cause pheochromocytoma-like symptoms such as headache, palpitations, and arterial hypertension. Another serious complication of the disease is fistulation to adjacent intestinal structures [12].

Diagnosis of adrenal hydatid disease is usually made through medical imaging, serological tests, and tissue biopsy [1,4]. Echinococcal IgG assay is usually negative in un-ruptured hydatid disease [18].

Diagnostic imaging tools (Ultrasonography, CT, and, to a lesser extent, MRI) are essential to the diagnosis and clinical management of hydatid disease [14].

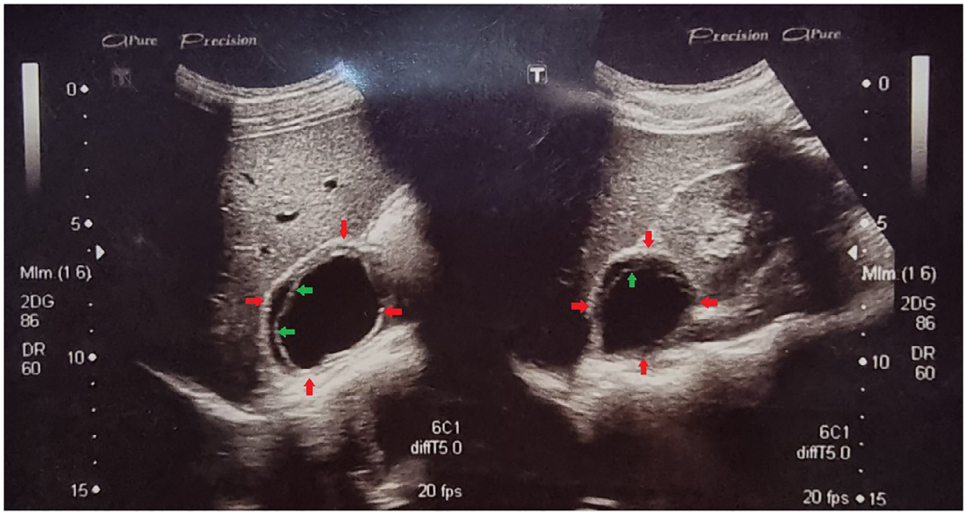

Fig. 1 – Ultrasound images revealed a large solitary anechoic cystic lesion located in the right suprarenal region (red arrows), showing a double-wall sign (green arrows).

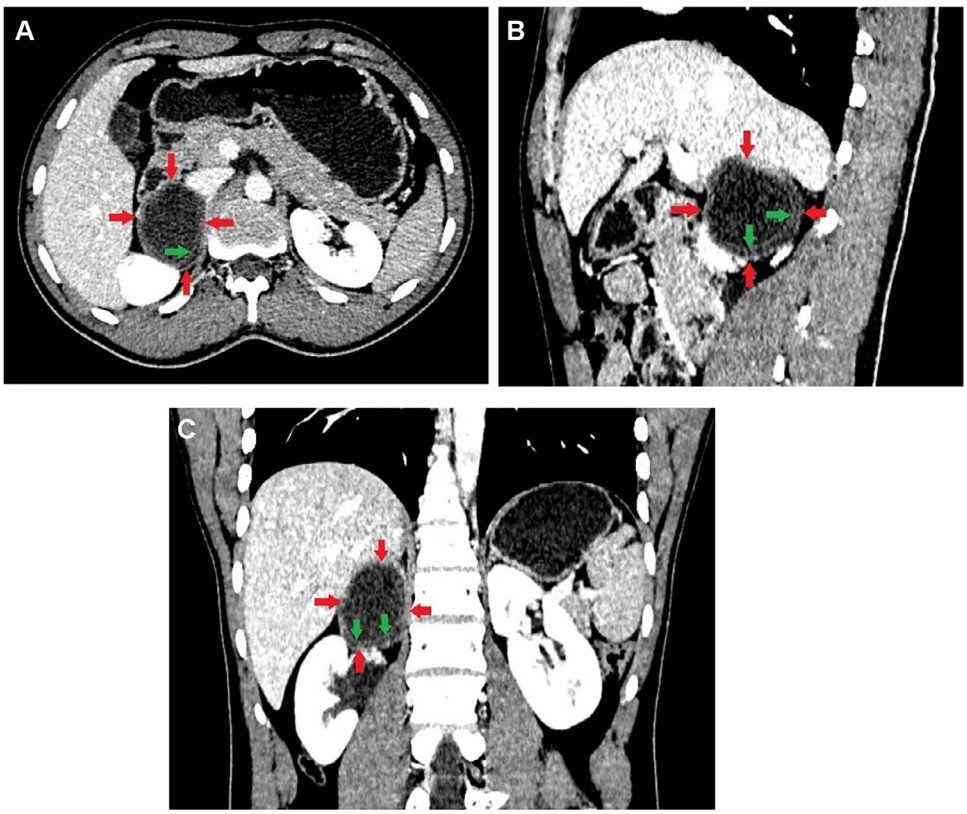

Fig. 2 – Axial (A), sagittal (B), and coronal (C) contrast-enhanced computed tomography showing a well-circumscribed, unilocular, nonenhanced cystic lesion in the right adrenal gland (red arrows) with typical "double-wall sign" of hydatid cyst (green arrows).

Ultrasonography is the first-line imaging method for the evaluation of both symptomatic as well as asymptomatic patients. The diagnostic sensitivity of ultrasonography is high, ranging from 93% to 98%. Ultrasonography is also useful for longitudinal studies, such as monitoring the response of hydatid cysts to treatment and for assessing cyst growth rate. Sonographic findings of echinococcus granulosus (hydatid cyst disease) on grayscale ultrasound include; a predominately anechoic cyst with hydatid "sand"/double wall, multiseptate cyst with "daughter" cysts and echogenic material

between cysts, water lily sign (cyst with floating, undulating membrane and detached endocyst), and densely calcified echogenic mass with posterior acoustic shadowing [16]. In 2003, the World Health Organization (WHO) proposed a standardized ultrasound classification, which defines 6 cyst stages (CE1, CE2, CE3a, CE3b, CE4, and CE5) that are assigned to 3 clinical groups (active, transitional, and inactive). The active group comprises developing cysts, which may be unilocular (CE1) or multivesicular with daughter cysts (CE2) and are usually found to be viable. The transitional group (CE3) contains cysts that are usually starting to degenerate. There are 2 subtypes of the CE3 group: the "water lily signs" for floating membranes (CE3a); and predominantly solid cysts with daughter cysts (CE3b). This subdivision is based on their different response to percutaneous treatment and albendazole, which is generally good for CE3a and poor for CE3b. The inactive group (CE4 and CE5) exhibits involution and signs of solidification of cyst content with increasing degrees of calcification and is nearly always found to be nonviable [3,13,14].

CT scan is the preferred imaging modality, especially for the evaluation of calcified hydatid cysts, which show strong posterior acoustic shadows on ultrasound [15]. CT scan has a sensitivity of around 97-98% with the advantage of inspecting any organ, detecting smaller cysts, locating cysts precisely, and sometimes differentiating parasitic from nonparasitic cysts [3,13,14]. CT Findings in echinococcus granulosus include; uni- or multilocular, well-defined cysts containing multiple peripheral "daughter" cysts of lower density than "mother" cyst. Curvilinear ring-like calcification of the pericyst (wall) can be seen, usually indicating no active infection if completely circumferential. Enhancement of cyst wall and septa may be seen on CECT. The CT findings in echinococcus multilocularis (alveolaris) which is an aggressive form of the disease, include; an infiltrative cystic and solid masses of low density (14-40 HU) with irregular and ill-defined margins and amorphous type of calcification, simulating primary or secondary malignant tumor. Minimal enhancement of noncalcified portions can be seen on CECT [16].

MRI, with its inherent tissue-characterizing ability, offers utility in the assessment of adrenal disorders including hydatid cysts. MRI may have some advantages over CT scanning in the evaluation of postsurgical residual lesions, recurrences, and selected extrahepatic infections, such as cardiac infections [14].

Conventional radiographs are not useful for the detection of adrenal hydatid cysts but can be used for cysts in the lungs, bone, and muscle [14].

Routine laboratory examination is usually nonspecific. Eosinophilia may occur in one-fourth of cases. The sensitivity of serologic echinococcus IgG ELISA tests is about 90% [19].

In general, the decision for the treatment of hydatid cysts should take into account the size of the cyst, the reported stage of the disease, and whether it is complicated or not. Both laparoscopic resection and laparotomy can be performed. Surgical treatment, including pericystectomy and resection of the entire adrenal gland, is the preferred approach for managing adrenal hydatid cysts. It is crucial to achieving complete cystectomy while preserving the adrenal gland, and in case of complications or failure to perform cystectomy, ablation of the entire adrenal gland including the cyst may be necessary [1,2,11]. Additionally, injecting the cyst with a hypertonic saline solution before puncture can inactivate scolices and daughter cysts, aiding in the management of the condition. In some cases, after administering the drug, the cyst can be pulled out through the skin. Control and prevention of this disease in the community is done by treating dogs with praziquantel and vaccinating sheep with the EG95 vaccine [4,14].

Conclusion

Adrenal glands are an extremely rare location for hydatid disease. When a cystic lesion is seen in the adrenal glands, a hydatid cyst should be considered in differential diagnosis, especially in endemic regions. The clinical and routine laboratory features of this disease are nonspecific. Ultrasonography and CT scans play an important role in the early detection, classification, management, and follow-up of the disease.

Patient consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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