Case Report *Olgu Sunumu*

Isolated primary hydatid cyst of adrenal gland: a case report

Primer izole adrenal kist hidatik: Olgu sunumu

Mohammed I. Abdulmajed[1], Berkan Reşorlu[2], Cengiz Kara[2], Kadir Türkölmez[1]

[1]Ankara University School of Medicine, Department of Urology, Ankara, Turkey
[2]Ministry of Health Keçioren Training and Research Hospital, Department of Urology, Ankara, Turkey

Abstract

Although Echinococcal cysts can involve almost any part of the body, they usually occur in liver and lungs. Other organs are occasionally involved including brain, muscle, kidney and heart, making diagnosis and treatment more complex. We present a rare case of extrahepatic Echinococcosis in a 48-year-old female patient with history of unexplained hypertension. Patient was found to have an adrenal lesion on imaging, which turned to be an isolated primary adrenal hydatid cyst after surgical excision.

Key words: Adrenal gland; echinococal cyst; extrahepatic echinococcosis; treatment.

Özet

Ekinokokal kistler vücudun birçok bölgesinde meydana gelebilsede sıklıkla karaciğer ve akciğeri etkilemektedir. Nadir görülen beyin, kas, böbrek ve kalp gibi diğer organların tutulumlarının ise tanı ve tedavisi daha zordur. Bu yazıda ender bir ektrahepatik ekinokokozis olgusu olarak açıklanamayan hipertansiyon nedeni ile başvuran 48 yaşında bir kadın hasta sunulmuştur. Görüntülemede adrenal lezyonu olan hastada, cerrahi eksizyon sonrası izole primer adrenal kist hidatik olduğu anlaşılmıştır.

Anahtar sözcükler: Adrenal bez; ekinokokal kist; ekstrahepatik ekinokokozis; tedavi.

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Hydatid disease is caused by a tapeworm (*Echinococcus granulosus* and, to lesser extent, *Echinococcus multilocularis*). Eggs from adult tapeworms are passed in the feces of the definitive host, including dogs and foxes, and ingested by intermediate hosts like human, cattle, and particularly sheep. In the intermediate host, eggs hatch and form the larval cysts that pass through the intestinal mucosa into the portal circulation and are usually entrapped in the capillary bed of the liver or the lungs, undergo cystic degeneration developing into clear fluid-filled cysts-Hydatid cysts (Fig. 1).[1]

Hydatid cysts in organs other than the liver, or the lungs are usually part of generalized hydatid disease and can only become the primary site when embryos bypass primary filtration organs, liver and lungs.[2] Hydatid cysts are mainly found in liver (50-70%) and lungs (20%), whereas other body organs are rarely involved as myocardium, brain, bones, spleen or kidney.[3] In this case we present an extrahepatic Echinococcosis in a woman with history of unexplained hypertension found to have an adrenal lesion on imaging, which turned to be an isolated primary adrenal hydatid cyst after surgical excision.

Case report

A 48-year-old female with one-year history of unexplained high systolic blood pressure (170-160 mmHg) and vague right loin pain referred to Out-Patient Clinic of Ankara University Department of Internal Medicine. Combination therapy with bisoprolol and amlodipine started. History, including enquiry into contact with sheep or dogs, occupational hazard, or suboptimal food hygiene, and physical examination were unyielding and followed by abdominal ultrasonography, revealing 48x45 mm heterogeneous, thick-walled solitary right adrenal cystic mass with peripheral distribution of echoes-calcification. The mass found avascular on Color-Doppler ultrasonography. Plain chest and abdominal radiographs revealed no pathology.

Subsequently, the patient was referred to our clinic, where abdominopelvic computed tomography (CT) and adrenal magnetic resonance imaging (MRI) were advised. CT confirmed 6x4x6 cm right adrenal cystic mass with peripheral calcifications and enhancing thickened-walls (Fig. 2). Adrenal MRI showed 6x4 cm hyperintense-T1 right adrenal cystic mass (Fig. 3).

For evaluation of possible pheochromocytoma or functioning adrenal adenoma, hormonal tests were performed including baseline serum cortisol, dehydroepiandrosterone sulfate (DHEA-S), serum baseline 17-hydroxyprogesterone (17-OHP), urinary metanephrine, normetanephrine, vanilylmandelic acid (VMA) excretion, low-dose 2 mg dexamethasone suppression tests, plasma aldosterone and renin activity. Results were inconclusive (Table 1).

Surgical excision of the mass using anterior subcostal trans-peritoneal approach was performed. The large cyst was found deep in the retroperitoneum almost covered by liver (Fig. 4) and its walls were thickened and adhesive to surrounding tissues. Hypertonic (20%) sodium chloride in 0.5% silver nitrate solution injected into the sac before aspiration and no obvious spillage of cyst fluid into the abdominal cavity occurred. Following aspiration, shrinkage occurred and the cyst removed fully (Fig. 5) preserving normal adrenal tissue using clips to provide adequate hemostasis. Histopathologically, thick membranous specimen of 6.5x2.5x0.5 cm weighing 14 cc was examined to reveal necrotic tissues, dystrophic calcification and epithelial cyst wall suggestive of hydatidosis. Albendazole, 90 mg/day oral dose, prescribed for one month. The patient`s post-operative period was uneventful and he was discharged on the 4th post-operative day.

Discussion

Hydatid cysts are mainly found in liver (50-70%) and lungs (20%), whereas other body organs are rarely involved as myocardium, brain, bones, spleen or kidney.[4] The adrenal gland is affected in about 0.05% of patients with hydatid disease.[3] Seldom, Echinococcal infection is limited to adrenal gland,[2] as in our patient.

The Echinococcal cysts in the adrenals are mostly symptom-free or, occasionally, result in ill-defined symptoms like dull abdominal pain, as in our patient. More serious rare manifestations are acute abdominal

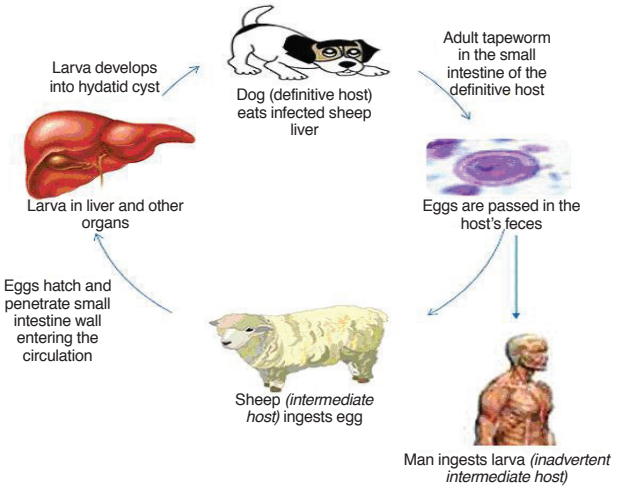

Figure 1 Life cycle of *Echinococcus granulosus*.[1]

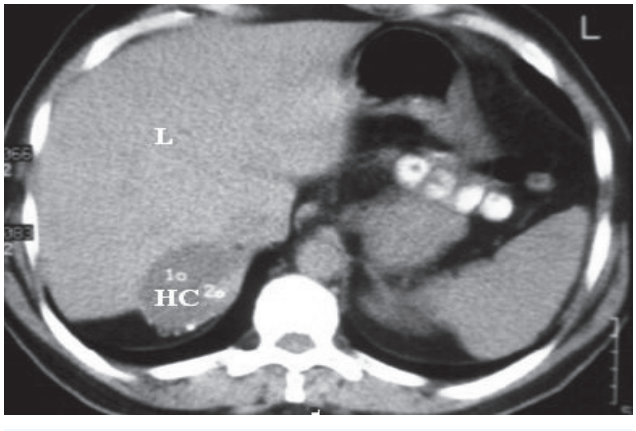

Figure 2 Upper abdominal computed tomography of the patient demonstrating 6x4x6 cm right adrenal cystic mass with peripheral calcifications and enhancing thickened-walls (L: liver, HC: hydatid cyst).

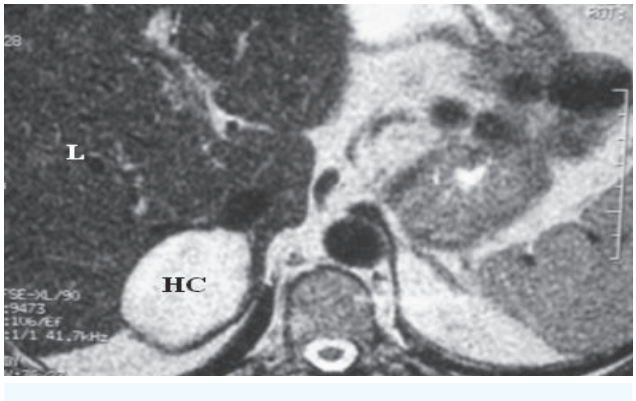

Figure 3 T1 phase dynamic adrenal magnetic resonance imaging showing hyperintenseisodense 6x4 cm thickened-wall right adrenal cystic mass at transverse view. (L: liver, HC: hydatid cyst).

Table 1. Laboratory resilts of the patient

Hormonal Analysis	Results	Normal values
Basal serum cortisol	11.2 μg/dL	9-23
Urinary metanephrine	75 μg/day	74-298
Urinary nor-metanephrine	182 μg/day	105-354
Urinary VMA	1.4 mg/day	1.4-6.6
1 mg DST-serum cortisol	3.2 μg/dL	
Renin	1.0 ng/dL	1.9-6.0
Aldosterone	21 ng/dL	3.81-31.3
DHEA-S	117.2 μg/dL	80-560
17-OHP	0.5 ng/mL	

VMA: vanilylmandelic acid, DST: dexamethasone suppression test, DHEA-S: dehydroepiandrosterone sulfate, 17-OHP: 17-hydroxypogestrone.

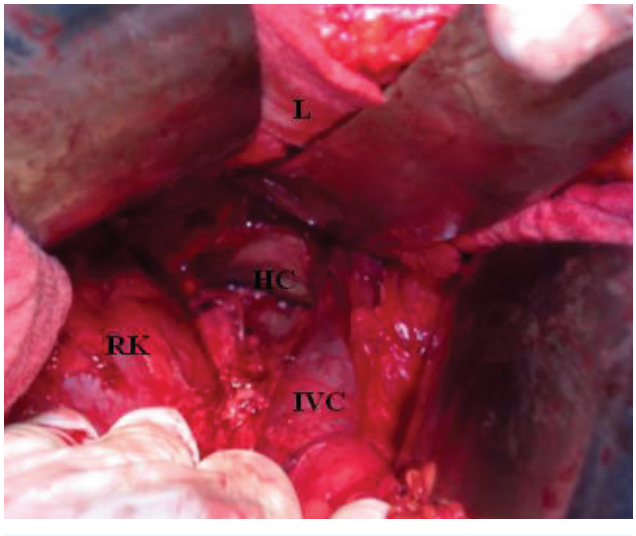

Figure 4 A scene from the adrenal cyst excision surgery showing the hydatid cyst (HC) surrounded by the liver (L) superiorly, the right kidney (RK) inferiolaterally, and the inferior vena cava (IVC) medially.

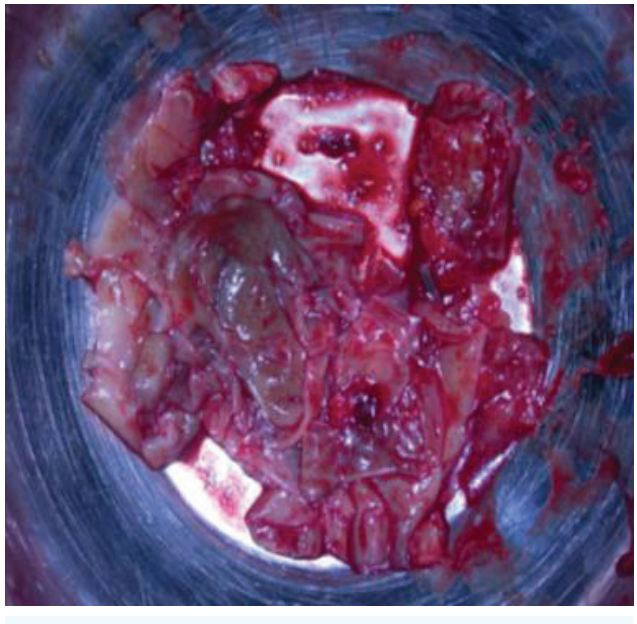

Figure 5 Specimen showing the excised wall of adrenal hydatid cyst.

pain and tender mass accompanying intracystic hemorrhage, rupture or infection; or anaphylactic shock caused by cyst rupture.[5]

Endocrine abnormalities are infrequently seen in adrenal hydatid disease.[6] However, till 2006, 3 cases of coexisting arterial hypertension, the first sign of endocrine changes, and primary adrenal hydatid cyst were reported in the literature.[7] In our case, investigation for high blood pressure in an otherwise normal patient revealed an isolated adrenal cyst, which was confirmed as a primary hydatid cyst after excision. Although no reliable link between arterial hypertension and primary adrenal hydatidosis can be established,[7] a possible explanation is the Goldblatt phenomenon-hypertension resulting from partial occlusion of a renal artery by external pressure.[8]

Laboratory tests like eosinophilia, Casoni skin test, and indirect hemagglutination (IHA) are not completely reliable.[2] For instance, Casoni test is positive in 95% of cases, however, it can also have up to 40% false positive results.[9] Similarly, IHA sensitivity rates vary from 60% to 90%, but whether the patient has detectable antibodies depends on physical location, integrity, and vitality of the larval cyst i.e. cysts in the liver are more likely to elicit antibody response than elsewhere.[10] New more sensitive and specific

serological tests are available, including complement fixation, enzyme-linked immunosorbent assay, ARC5 precipitation, and specific hydatid IgE tests,[2] however, hydatid cyst's appearance in our case interpreted as an indeterminate cystic mass and, therefore, any of the above investigations was not done.

Hydatid cyst identification in adrenal gland is based mainly on ultrasonography and CT scan[11] with diagnostic sensitivity of ultrasonography (US) ranging from 93% to 98%, while that of CT is around 97%.[12] Radiologically, hydatid cysts can be described according to World Health Organization (WHO) classification dividing cysts into active (CL, CE1 and CE2), transitional (CE3) and inactive (CE4 and CE5) cysts to facilitate treatment selection.[13] While, Gharbi classification[14] distinguishes hydatid cysts into unilocular (Gharbi type I and II), multilocular (Gharbi type III) and degenerated (Gharbi type IV) cysts. The solitary cyst in our case is ascribed into WHO class CE5 and Gharbi type IV (peripheral calcification and thickened-walls in complex and heterogeneous mass).

Bosniak's CT classification recognizes five cyst types (Category I, II, IIF, III and IV), depending on radiological features with the suspicion of malignancy, and it assists to clinical decision.[15] In our patient, the cyst found to be Bosniak Category III (peripheral calcification, thickened walls, and septations) warranting surgical exploration. Owing to this fact, and because CT and MRI demonstrate highly sensitive morphological features, histopathological confirmation is needed[16] justifying exploration and excision in our case.

Treatment of hydatid disease of adrenals is mostly surgical and the operation of choice is removal of cyst preserving ipsilateral kidney and, if possible, the remaining adrenal gland to provide adequate haemostasis.[17] Access to the adrenal gland may be through an anterior transabdominal or posterior retroperitoneal traditional open approach. Cyst injection with hypertonic saline solution before puncture can inactivate scolices and daughter cysts.[2] In our patient, anterior subcostal trans-peritoneal approach was used and cyst fluid aspiration and hypertonic saline solution injection into the sac preceded attempts to remove the cyst.

Recent minimally invasive surgical techniques like transabdominal laparoscopic and retroperitoneal endoscopic adrenalectomy have been conducted. However, their role is still debatable as some studies claim less morbidity,[18] while others do not recommend a laparoscopic approach for complex cysts[19] and warn that cysts larger than 8 cm in diameter have higher risk of spillage of daughter cysts and development of secondary Echinococcosis[20] Moreover, it is shown that open approach has the advantage of allowing the surgeon to explore the peritoneal cavity[7].

Antihelminthic agents like mebendazole and albendazole can be a treatment option pre- or post-operatively for hydatidosis to reduce operative intracystic pressure and prevent further hydatid seeding, but should not be the primary choice when hydatid cysts are disseminated, recurrent or if surgery is contraindicated.[21] The usual oral dosage of albendazole and mebendazole is 10-15 mg/kg/day and 40-50 mg/kg/day, respectively. Treatment should be started 4 days before surgery and last for 1 month for albendazole, and 3 months for mebendazole, post-operatively.[21] In our patient, though any obvious spillage has not been recorded, 1-month of oral albendazole 90 mg/day started post-operatively to avoid recurrence.

PAIR (Puncture, Aspiration, Injection and Re-aspiration) treatment of hydatidosis is considered minimally invasive, confirming diagnosis and improving efficacy of chemotherapy given before and after puncture. Nevertheless, PAIR is usually avoided in patients with adrenal hydatidosis and can become an alternative method in inoperable cases.[13]

In summary, primary adrenal hydatidosis, although rare, should be considered among the differential diagnosis of adrenal cysts, especially in high-incidence areas.

Conflict of interest

No conflict of interest was declared by the authors.

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Correspondence (Yazışma): Uzm. Dr. Berkan Reşorlu.
Kardeşler Koop. 182. Cad. 175. Sok. 14/8, 06010 Ankara, Turkey.
Phone: 0312 344 51 75 e-mail: drberkan@yahoo.com