Hydatid Disease Involving Some Rare Sites in the Body

Vücüdumuzda Nadir Alanları Tutan Hidatik Hastalık

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ABSTRACT

A hydatid cyst is an endemic disease in our country. Clinical manifestation includes cyst formation, most commonly in the liver and lungs. Renal, brain, and subcuteneous localizations are rare. Here we report four cases: two cases of primary renal hydatid disease, one of intracranial hydatid cyst, and one of subcutaneous hydatid cyst. We discuss the prevalence, diagnostic workup, and management of echinococcosis.
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Keywords: *Echinococcosis*, intracranial, renal, subcutaneous, hydatid cyst

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ÖZET

Hidatik kist bizim ülkemizde endemik hastalıktır. Klinik belirtisi karaciğer ve akciğerde kist oluşumudur. Böbrek beyin, subkutanöz lokalizasyonlar nadirdir. Biz iki tane renal kist hidatik, bir tane intrakranial, bir tane subkutanöz kist hidatik olgusu raporladık. Bu olguların sunumunda ekinokokun prevelansı, tanısal özellikleri ve tedavi yönetimi tartışıldı. *(Turkiye Parazitol Derg 2015; 39: 78-82)*

Anahtar Sözcükler: Hidatik hastalık, *ekinokokkosis*, alışılmadık lokalizasyonlar

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INTRODUCTION

Hydatid disease is one of the most frequent parasitosis, caused by the larval stage of *Echinococcus granulosus*, especially in countries with a warm climate such as India, African countries, Turkey, South American countries, and Middle Eastern countries. *E. granulosus* is one of the most frequent cause of parasitosis in Turkey (1). It can reach any organ or tissue in the body, developing into small hydatid cysts. Most commonly, it occurs in the liver (80%) and lungs (10%). The precise percentage of site involvement varies and the precise incidence of unusual locations is difficult to ascertain because they are only reported as case reports. In 10% of cases, hydatid disease arises in the viscera; it mainly arises in the spleen (0.9-8%), kidney (2%), bones (3%), brain (1%), heart muscles, and peritoneal cavity (2, 3). Dogs are the definitive host and sheep are the intermediate host. Man is an accidental intermediate host. The hydatid cyst grows slowly over years and causes symptoms usually because of compression of adjacent structures. A high index of suspicion, radiological investigations, and histo-pathological examination is necessary for establishing the

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Figure 1. Gross appearance of the cyst of the kidney

diagnosis of hydatid disease at unusual sites in the body. Here we report four cases: two rare cases of primary renal hydatid disease, one of intracranial hydatid cyst, and one of subcutaneous hydatid cysts. We present a review of the occurrence of hydatid disease at unusual sites and our experience with its clinical presentation and management. We aimed to draw attention to hydatid cyst disease in the differential diagnosis of cysts although it is very rare.

CASE REPORTS

Case 1

A 66-year-old man was admitted with abdominal pain. Physical examination, complete blood count, blood urea nitrogen, creatinine, alanine, aminotransferase, aspartate aminotransferase, and electrolytes were normal. Ultrasonography revealed a hypoechoic cystic mass measuring 3.5×3.5×3 cm in size in the left kidney. The rest of the visceral organs did not show any cystic lesions and were unremarkable. The patient underwent partial nephrectomy. Examination of the specimen revealed an enlarged kidney measuring 5×5×3 cm. The cut section revealed a cystic lesion measuring 3.5 cm in the upper pole (Figure 1). Microscopically, a cellular, laminated, eosinophilic hyaline membrane was seen in the necrotic tissue (Figure 2). A section from the adjacent renal parenchyma showed atrophy and lymphocytic infiltration. The lesion showed calcification along the wall.

Case 2

A 33-year-old man was admitted to the hospital with the complaint of right flank pain. Ultrasonography revealed a renal mass measuring 8.5×6×5 cm in size in the right kidney. The rest of the visceral organs including the liver, lung, and left kidney did not show any cystic lesions and were unremarkable. Routine hematological findings revealed eosinophilic count of 230 cells/cumm and serum creatinine level of 1.3 mg/dL. Right nephrectomy was performed. On examination, the mass appeared cystic and necrotic; thus, the membrane of a hydatid cyst was observed.

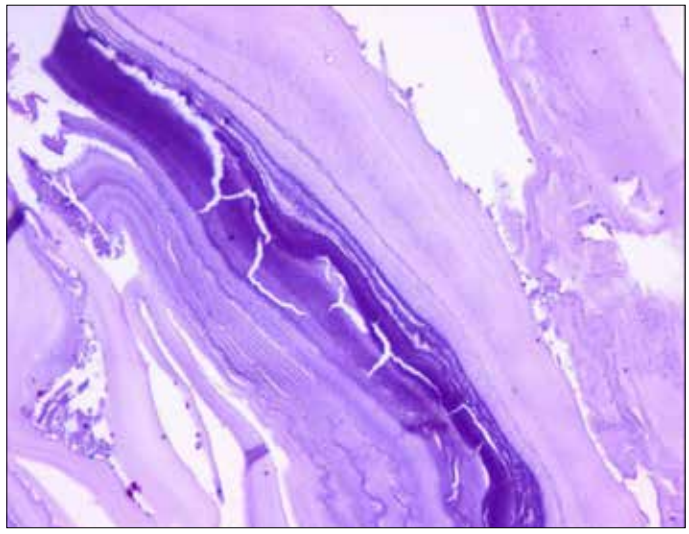

Figure 2. A cellular, laminated, eosinophilic hyaline membrane was seen in the cyst wall (Hematoxylin and Eosin staining, ×400)

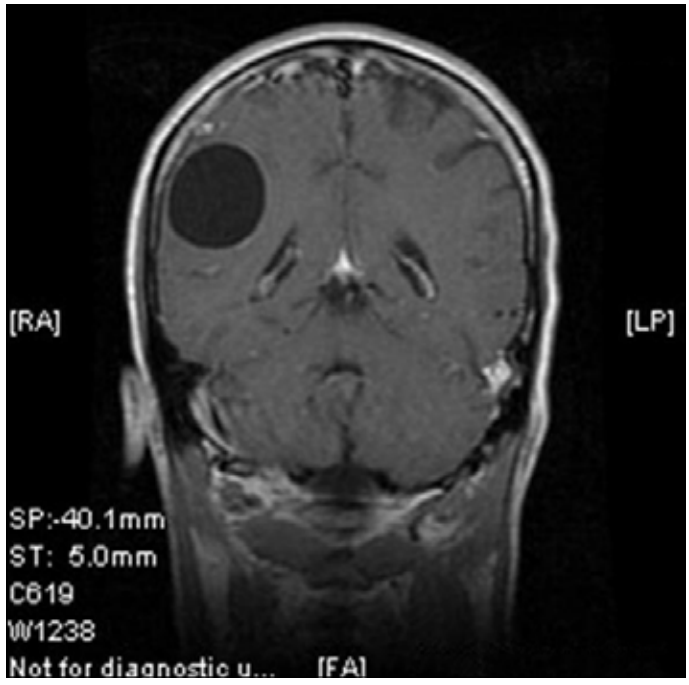

Figure 3. MRI revealed a cystic parietal lobe

Both the patients have been on postoperative oral albendazole therapy with regular follow-up since 3 months, which has been uneventful.

Case 3

A 53-year-old man presented with the complaint of diffuse episodic headache for 3 months previously that had increased in intensity over the previous week. There was no history of seizures, trauma, surgery, or hydatidosis. An intracranial hydatid cyst was diagnosed on computed tomography (CT) and magnetic resonance imaging (MRI). A single, spherical, well-defined, thin-walled homogenous, isodense cyst with a diameter of 34 mm was seen in the right parietal region (Figure 3). Thoracic and abdominal CT revealed cystic lesions. Laboratory data showed

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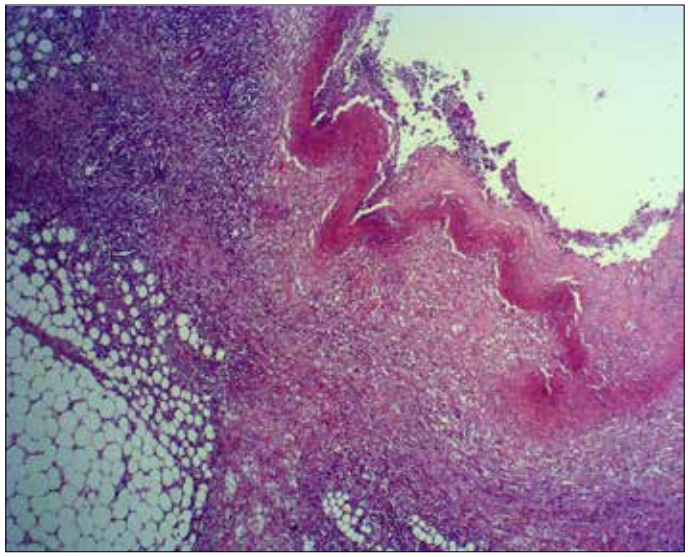

Figure 4. Subcutaneous cyst hydatid (Hematoxylin and Eosin staining; ×100)

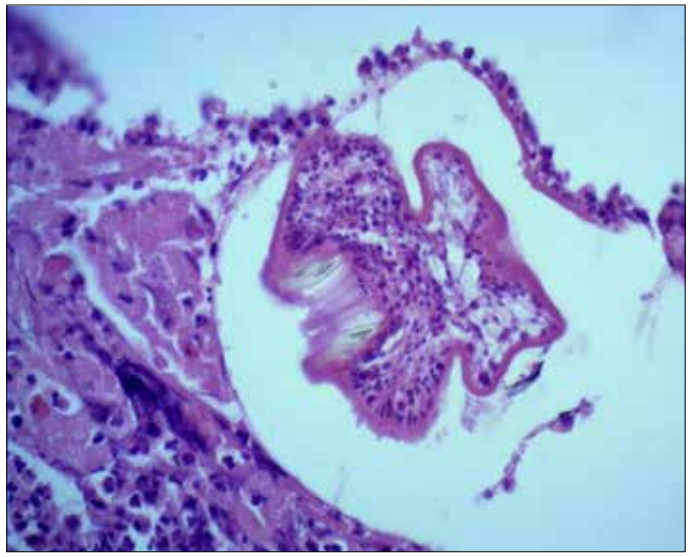

Figure 5. There is a scolex near the germinal layer (Hematoxylin and Eosin staining; ×400)

mild leucocytosis without significant eosinophilia. Parietal craniotomy and cyst removal were performed under general anesthesia. The histopathological examination confirmed hydatid disease. Albendazol therapy three times a day for 4 months was initiated as medical treatment. The patient has been followed up for 4 years without any complaints.

Case 4

A 64-year-old woman who lived and worked in a farm in Turkey presented with a 4-year-growing mass measuring 4 cm in the infraumbilical region. No ultrasonography and MRI were performed. The cyst was excised and a pathological examination confirmed it to be a subcutaneous hydatid cyst measuring 1.7 cm (Figure 4, 5). The rest of the visceral organs did not show any cystic lesions and were unremarkable.

DISCUSSION

Hydatid disease is an endemic parasitic zoonosis caused by the larval stage of *Echinococcus*. Two variants of this disease occur: classic hydatid disease caused by *E. granulosus* and the rarer variant caused by *E. multilocularis*, which is more aggressive because it infiltrates the organ involved and cannot be easily removed. *Echinococcus* belongs to the order Cestoda and family Taenia. It is about 5 mm long (4). The definitive host is dogs and the intermediate host is sheep. Human is accidental or incidental intermediate host. The intestine of the definitive host is the tissue, where adult worm resides. Eggs passed in the feces are ingested by grazing sheep, goats, and cattle. Eggs hatch, penetrate the host's intestinal wall, and reach the liver through the portal vein. They are distributed to the lungs and other organ systems by bloodstream. Eggs transformed to the larval stage, the scolex, which can multiply asexually indefinitely within the hydatid cyst. When a hydatid cyst is devoured by a canine host, the natural cycle is completed. The multiplication of the larval scolices results in a slow but steady physical enlargement of the cystic colony (5). The cyst consists of three layers. The outer most, or the pericyst, is an adventitial layer of host origin. The middle layer is the outer chitinous covering of the parasite or the laminated membrane. The innermost germinal layer gives rise to the scolices (6).

The hydatid cyst grows slowly over years and causes symptoms usually because of compression of adjacent structures. The asymptomatic period is extremely long and the disease might be diagnosed even after 20-25 years post-infection. Because the enlargement is very gradual, the patient's symptoms are rarely acute. As the cyst grows, the symptoms become more specific depending on the specific structures involved. The rupture, infection, and compression of the cysts may cause symptoms. Although the disease is sometimes asymptomatic, symptoms such as abdominal and chest pain, mass, fever, weight loss, anaphylaxis, jaundice, and neurological signs are seen. Patients with renal hydatid cysts usually present with vague pain in the lumbar region and hematuria. A preoperative definitive diagnosis of renal hydatid disease is difficult even when all examinations are performed (7).

There is no specific laboratory finding of renal hydatid disease. Moderate eosinophilia is nonspecific, although it presents in 20-50% of cases (8). Eosinophilia was seen in our patient with renal hydatidosis. The diagnostic dilemma of a hydatid cyst at unusual sites such as the kidney can lead to complications, because sometimes the cyst may present as acute surgical emergency or a chronic illness, leading to morbidity. Therefore, imaging examinations are important for early diagnosis. Ultrasonography is the most useful radiological examination for diagnosis. Noncontrast CT and MRI are highly sensitive for lesions. Hypointense rim and multicystic appearance is distinctive on MRI, which also delineates the anatomy well. CT has an accuracy of 98% and sensitivity sufficient to reveal the daughter cyst (9, 10). An indirect hemagglutination test performed can also suggest hydatid disease. The sensitivity of various serological tests used for hydatid disease varies from 64 to 87% (11).

The occurrence of *E. granulosus* in some locations of the body is very rare. Such anatomic locations may cause difficulties in diag-

nosis. Hydatid disease is a differential diagnosis of cystic lesions, especially of cystic lesions encountered in patients who live in or have come from an endemic region. The diagnosis of hydatid disease is based on the patient's history, clinical findings, serum biochemical profiles, serologic tests, and pathological diagnosis. The most important factor in the diagnosis of hydatid disease is the high index of suspicion about its possibility.

Hydatid disease of the kidney is extremely rare and constitutes only 2-4% of all cases of hydatid disease with renal hydatid cysts commonly present with loin pain and hematuria (12). Kidney involvement has insidious onset and usually remains asymptomatic for many years. The differential diagnosis of renal cystic mass includes multilocular cysts, cystic renal carcinoma, and hydatid cysts according to the age of the patients and findings of radiological tests. Primary hydatidosis of the kidney should always be considered in the differential diagnosis of any cystic renal mass, even in the absence of accompanying involvement of the liver or other visceral organs, especially in endemic areas.

Intracranial hydatid disease is rare, with a reported incidence of 1-2% of all cases, and it is more common in children. Although in the other organs they are multiple, multiple intracranial cysts are rare (13). It is most frequently located in both the hemispheres. A typical intracranial hydatid cyst presents as a well-defined solitary cystic lesion in the middle cerebral artery territory in the parietal lobes, although it can be seen in any location including the skull vault, extradural, intraventricular, meningeal, posterior fossa, and brainstem (14). The parietal region is the commonest site and was seen in the present case. Intracranial hydatid cysts may also be classified as primary or secondary. Primary cysts occur as a result of direct infestation of the larvae in the brain without demonstrable involvement of other organs; secondary multiple cysts result from spontaneous, traumatic, or surgical rupture of the primary intracranial hydatid cyst. They lack a brood capsule and scolices. Patients with intracranial hydatid cysts usually present with focal neurological deficit and features of raised intracranial pressure; the latter may be due to the large size or the interference with the pathway of the cerebrospinal fluid. In view of the pathological and clinical findings, the cyst in the present case could be classified as a secondary brain hydatid cyst. Surgically, intact cyst excision is the ideal treatment. Medical treatment with albendazole seems to be beneficial both pre- and postoperatively.

Primary subcutaneous hydatid cysts are very rare and the incidence is unknown. Subcutaneous hydatid cyst may be secondary or primary. The mechanism of the primary subcutaneous localization is unclear; direct spread from adjacent sites is likely the mechanism of infection (15). Most patients complained of slow growing, painless, mobile masses with normal overlying skin. All patients were from endemic areas and most patients were from rural areas, as in our case. The location around the trunk and roots of the limbs could be explained because of an increase in vascularization and less muscular activity in these areas. A primary subcutaneous hydatid cyst should be kept in mind for differential diagnosis of soft tissue masses, particularly for patients who have lived in regions where a hydatid cyst is endemic. The differential diagnosis of soft tissue masses includes abscess, a sebaceous cyst, lipoma, tuberculous abscess, an aneurysm, hernia, sarcoma, chronic hematoma, a synovial cyst, and a necrotic soft tissue tumor. Complete excision is the best treatment option. A final diagnosis is confirmed by histopathology. After the diagnosis of a subcutaneous hydatid cyst, radiological screening of the body should be performed to look for another focus (16).

Management of hydatid cyst disease includes a combination of chemotherapy and surgical intervention. Puncture, aspiration, and injection are surgical procedures. In general, surgery is the best treatment option for hydatid cysts. Detailed examination of the thorax and abdomen must be performed by imaging studies before surgery. Kidney sparing surgery is possible in most renal hydatid cyst cases. Nephrectomy must be reserved for nonfunctioning kidneys. Medical treatment with benzimidazole, such as albendazole and mebendazole, is recommended during pre- and postoperative periods in order to sterilize the cyst, to decrease the chance of anaphylaxis, to decrease the tension in the cyst wall, and to reduce the recurrence postoperatively.

CONCLUSION

A hydatid cyst should be considered in the differential diagnosis of a cystic mass, especially in endemic areas. The combination of history, imaging tests, and serological tests aid in diagnosis. The possibility of a striking clinical resemblance between a hydatid cyst and malignant disease of the kidney has been emphasized in the English literature. We also indicated this important point in our patients. Surgery is the best treatment for renal hydatid cysts and if possible, kidney sparing protocol is the logical option; however, nephrectomy must be reserved for nonfunctioning kidneys. A primary subcutaneous hydatid cyst should be in mind in the differential diagnosis of soft tissue masses, particularly for patients who have lived in regions where a hydatid cyst is endemic. Brain hydatidosis should be considered in the differential diagnosis of brain cysts.

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