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A Cystic Mass does not Always Mean Hydatid Cyst in Endemic Areas

Endemik Bölgelerde Kistik Lezyonlar Her Zaman Kist Hidatik Anlamına Gelmez

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Abstract

Within the medical community, there is a tendency to describe all cystic lesions in the liver and lungs as Hydatid disease (HD) in areas with HD endemics. This approach may sometimes cause a misdiagnosis. We have three cases with children aged between seven, seven and ten year old, all of whom had been diagnosed, via radiologic imaging, with HD cysts and started on treatment without confirmation. The true diagnoses of these cases were undifferentiated embryonal sarcoma (UES) in two and oesophageal duplication in one case, respectively. The indirect haemagglutination assays (IHA) were obtained in two of them. Although their results were negative, albendazole treatment was started in all cases. Confirmatory tests should be run in dubious cases. An IHA test can aid the diagnosis, although its effectiveness is limited. The possibility of false positive and negative results always exists, especially in lung cysts. A percutanous biopsy is strongly advised for differential diagnosis.

Key Words: Hydatic cyst, undifferentiated embryonal sarcoma, oesophageal duplication

Özet

Endemik bölgelerde gerek karaciğer ve gerekse akciğerde görülen kistik lezyonları kist hidatik olarak tanımlamaya bir eğilim vardır. Ancak bu eğilim zaman zaman yanlış tanıya yol açmaktadır. Yaşları 7 ve 10 arasında değişen herhangi bir doğrulama yapılmadan radyolojik olarak kist hidatik tanısı alarak tedavi görmüş üç hasta ile ilgili tecrübelerimizi paylaşmak istedik. Hastalarımızın ikisinde tanı andiferansiye embriyonal sarkoma (UES), diğerinde ise özofagus duplikasyonu olarak saptandı. İndirekt hemaglutination testi (IHA) iki hastada araştırılmış ve negatif olarak saptanmıştır. Tüm hastalara ise ampirik olarak albendazol tedavisine başlanmıştı. Ayırıcı tanı için tetkikler şüpheli olgularda mutlaka yapılmalıdır. IHA testi tanı için sınırlıda olsa yardımcı olabilir. Yanlış pozitif ya da negatif sonuçlar özellikle akciğer kistlerinde olmak üzere mevcuttur. Ayırıcı tanıda şüpheli olgularda perkütan biopsi önerilir.

Anahtar Kelimeler: Kist hidatik, andiferansiye embriyonel karsinoma, özofagus duplikasyonu

Introduction

Hydatid disease (HD) is a zoonosis caused by larval forms of echinococcus tapeworms. It has an endemic distribution, especially in rural areas of developing countries. This disease can affect almost all organs in the human body. When a cystic mass is identified in the liver or lungs, the presumptive diagnosis is usually of a hydatic cyst in endemic areas. This prejudice can cause misdiagnosis and delay treatment.

There are no clinical features common to all cases of HD; its symptoms depend upon the organ or organs affected [1]. Clinical symptoms will not occur until the cyst affects neighbouring structures or ruptures as a result of trauma [2]. The effectiveness of serologic tests in suspected cases was limited [3].

Case Report

Several different institutions, two separate illnesses in three children who had been presumptively diagnosed with (and treated for) hydatid cysts were reviewed with the approval of the patients.

The first two cases involved previously healthy male children. Both were seven years old. They were admitted to our institution with complaints of abdominal distension, nausea, and vomiting. A physical examination revealed abdominal tenderness localized in the right upper quadrant in both children. One of the children had a massive hepatomegaly causing respiratory distress due to the elevation of the diaphragm. Laboratory data came back normal, except that it showed high levels of lactate dehydrogenase (600 UI/L) in one case and mildly elevated aspartate aminotransferase (42 UI/L), alkaline phosphates (72 UI/L) and glutamic-pyruvic transaminase (GPT) (53 UI/L) in the other case. Liver function tests and serum assays for tumour markers, such as serum α-fetoprotein (AFP), yielded normal results in both cases.

In the first case, an abdominal ultrasonography (US) showed a 6x5cm cystic lesion with septation and echogenic material in the right lobe of the liver. Therefore, suspicion

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of HD was aroused. An abdominal computed tomography (CT) was performed, and cystic solid content with septations compatible with HD were documented (Figure 1). In the institution where the second case was administrated, a was performed, which showed a cystic lesion located in the right lobe of the liver (Figure 2). After the boy was transferred to our institution, an abdominal ultrasonography was performed. It revealed a large, cystic-appearing liver mass. This lesion elevated the diaphragm and compressed the right lung.

The first case was diagnosed as HD and treated with albendazole 10 mg/kg per day in two divided doses one day before surgery. The albendazole treatment for the second case had been started at the other institution with the same dosage as mentioned above.

Treatment for HD was started depending solely upon radiological findings in both cases. No serological investigations were applied in the first case. In the second case, albendazole treatment was begun despite a negative indirect haemagglutination (IHA) result (<1:160).

The first patient underwent surgery the day after treatment started, and it was discovered that his cystic hepatic tumour was different from HD. In the second case, we perform a percutaneous liver biopsy. As in the first case, the result pointed to liver malignancy. The patients underwent a complete tumour resection, consisting of a right hepatectomy with no complications. Post-operative histological examination of the biopsy specimens demonstrated undifferentiated embryonal sarcoma (UES). No complications were observed during the postoperative course of the past year.

The third case involved a seven-year-old female with a large intrathoracic cyst on her left side. She was first diagnosed with HD and treated medically with albendazole. The indirect haemagglutination test was negative. After nine months of medical treatment in different institutions, she was transferred to our institution, because no regression or maturation had occurred in the cyst, surgical treatment was pursued. During the operation, the aspiration of bloody and brown cystic fluid was not considered compatible with HD. The cyst was totally extracted, and the pathological evaluation was of cystic oesophageal duplication.

Discussion

Hydatid disease (HD) is a zoonosis caused by larval forms of echinococcus tapeworms. Although there are four kind of echinococcosis (echinococcus granulosus, echinococcus multilocularis, echinococcus vogeli, echinococcus oligarthrus), mostly only two of them cause illness in humans (echinococcus granulosus and echinococcus multilocularis) [1]. The real worldwide incidence of echinococcus tapeworms in children is obscure. In Turkey, the known incidence in children is 150 cases per 100,000 children [4].

Treatments for the diagnosis of HD were started in three different institutions. In all three cases, albendazole treatment had been started without any serological or pathological confirmation of the HD diagnosis. All diagnoses depended solely on radiological imaging, although no IHA positivity was found in any of the cases. The patients were easily diagnosed with HD due to the prevalence of HD in the area and the cystic appearances of the masses. This misdiagnosis can delay correct diagnosis and can sometimes incur fatal consequences.

Although the diagnosis of HD is usually based on patient history and clinical findings, neither method is specific enough to be diagnostic [1, 5]. In spite of the diagnosis of HD being primarily based on imagery and serological tests, the differential diagnosis between HD and cystic lesions may not be easy [1]. Using the WHO IWGE guidelines can be very helpful in preventing misdiagnoses [1].

Without clear symptoms associated with echinococcosis, physicians must use serological investigations. Indirect hae-

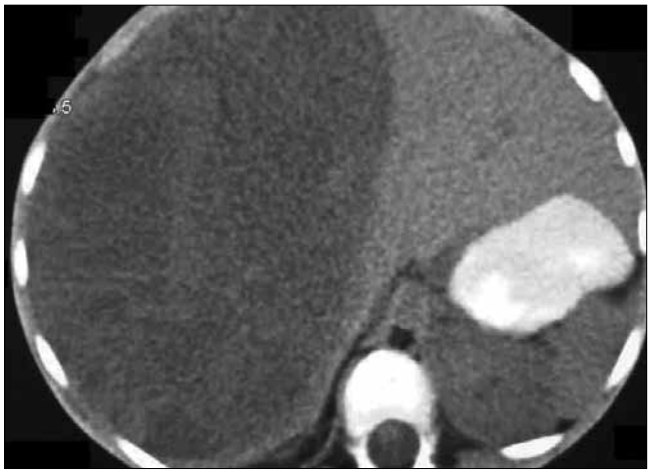

Figure 1. *The CT scan view of undifferentiated embryonal sarcoma.*

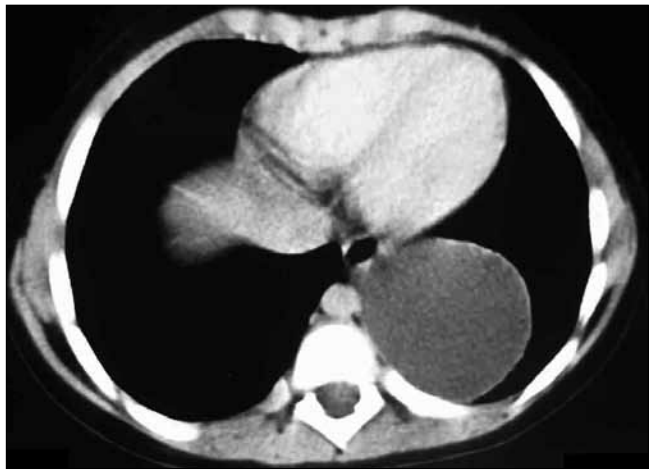

Figure 2. *The CT scan view of oesophageal duplication.*

magglutination (IHA) can be used to detect asymptomatic transporters and follow up treatment and recurrences [3]. Indirect haemagglutination (IHA) can be used as a first step screening test in confirming an HD diagnosis [1, 3]. The sensitivity of serum antibody detection using indirect haemagglutination, which is more practical in the field, ranges between 85% and 98% for liver cysts, 50% and 60% for lung cysts and 90% and 100% for multiple organ cysts, although false negative results are not rare [6]. Eosinophil counts are also more greatly elevated after the rupture/leakage of cysts, although this can also be seen in UES [7]. False-positives in IHA due to cross-reactions with other parasitic infections are very prevalent in countries with common sestod parasitosis [3]. The application of two different serological tests such as the İHA and the Enzyme-Linked ImmunoSorbent Assay (ELİSA) may increase sensitivity; however, false negative results still may not be eliminated completely [3].

Ultrasonography or CT can be selected as the primary test in field studies, and serologic tests should be performed in all cases of suspected lesions. Whenever there is any doubt, one should not hesitate to employ percutaneous approaches.

Some clinical criterion exist in cystic diagnoses, such as diagnosing a slowly growing or static cystic mass with imaging techniques; measuring anaphylactic reactions due to cyst leakage; or the incidental discovery of a cyst by imaging techniques in asymptomatic carriers. These are just some of the findings that cause suspicion of HD. According to WHO guidelines, a possible case of HD can apply to any patient with a clinical or epidemiological history and imaging findings as described above, or to any patient with a positive serology test for HD [1].

Requirements for the confirmation of a hydatid cyst include the demonstration of protoscoleces or their components in the aspirated contents of the cyst or changes in appearance on ultrasonographic images, such as cystic degeneration or maturation, either spontaneously or after the administration of albendazole for at least three months [1].

In our study, the third case was treated with albendazole for nine months. Although no maturation or degenerative change was observed during treatment, treatment was continued. In spite of albendazole treatment, a cyst may not degenerate, because echinococcosis can be resistant to the treatment [8-10]. In this condition, a percutaneous biopsy after three to six months of treatment can be helpful to rule out other possibilities [10]. If the diagnosis of HD is confirmed, the albendazole approach can be diverted to puncture, aspiration, injection, and respiration (or PAIR), which removes parasitic material in a way that is minimally invasive, less risky, and usually less expensive than surgery [1, 8]. PAIR is also suggested to rule out other possibilities. Besides differential diagnosis, microscopic examination of protoscoleces after the aspiration of the cyst's fluid also provides evidence about the parasitic nature and viability of a cyst [8].

In conclusion in endemic areas, cystic lesions in the liver or lungs can easily be misdiagnosed as hydatid cysts. Maturation of the cyst after administration of albendazole for at least three months, or spontaneous changes in the cyst's US appearance can be helpful in preventing misdiagnosis.

In developing countries, sestod infections can be as common as HD, which decreases the usefulness of IHA. In the presence of any doubt about the possibility of malignancy, a percutaneous biopsy should be performed.

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Informed Consent: Written informed consent was obtained from patients who participated in this case.

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