CYSTIC LIVER MASS DOES NOT ALWAYS MEAN HYDATID CYST IN ENDEMIC AREAS

Case Report

ENDEMİK BÖLGELERDE KİSTİK LEZYONLAR HER ZAMAN KİST HİDATİK ANLAMINA GELMEZ

Abdullah Yıldız

Şişli Etfal Research and Training Hospital, Department of Pediatric Surgery, Istanbul-Turkey.

Melih Akın

Şişli Etfal Research and Training Hospital, Department of Pediatric Surgery, Istanbul-Turkey.

Nihat Sever

Şişli Etfal Research and Training Hospital, Department of Pediatric Surgery, Istanbul-Turkey.

Arzu Canmemis

Şişli Etfal Research and Training Hospital, Department of Pediatric Surgery, Istanbul-Turkey.

Çetin Ali Karadag

Şişli Etfal Research and Training Hospital, Department of Pediatric Surgery, Istanbul-Turkey.

Husam Barhoom

Şişli Etfal Research and Training Hospital, Department of Pediatric Surgery, Istanbul-Turkey.

Ali İhsan Dokucu

Şişli Etfal Research and Training Hospital, Department of Pediatric Surgery, Istanbul-Turkey.

Corresponding Author

Abdullah Yıldız

Pediatric Surgery Division
Şişli Etfal Research and Training Hospital, Department of Pediatric Surgery, Istanbul-Turkey.
25100 Istanbul/TURKEY

e-mail: nihon@superonline.com

ABSTRACT

Undifferentiated embryonal sarcoma (UES) of the liver is a very rare and highly malignant entity. UES has no specific clinical or laboratory features. At computed tomography images , the tumour typically appears as a large, predominantly cystic mass with well defined borders. This cystic appearance of UES can be cause a misdiagnosis with hydatid cyst very easily due to rarity of UES and more common seen hydatic cyst especially in endemic areas. We present a eight year old male case with cystic mass in liver, which was misdiagnosed and treated as hydatic cyst. Percutaneous sample was obtained .Cytological examination did not confirm the hydatid cyst. Because of progressive respiratory deterioration, urgent abdominal operation was performed. Gross haemorrhagic cystic lesion was discovered and evacuated. Pathological examination revealed UES. Because of pure cystic appearance of UEA in liver can be suggestive of benign similar pathologies , and cause delaying or misdiagnosis in such hydatic areas.

Undifferentiated embryonal sarcoma (UES) of the liver is a very rare entity that was first described by Stocker and Ishak in 1978 (1,2). This rare neoplasm accounts for approximately 9% to 13% of all hepatic tumours in children (2). UES has no specific clinical or laboratory features, and fewer than 60 cases of paediatric UES have been reported in the literature (1). This cystic appearance of UES can cause the misdiagnosis of hydatid cyst very easily due to the rarity of UES, especially in endemic areas for hydatid cyst (3,4).

Key Words: *Undifferentiated embryonal sarcoma;hydatid cyst;sarcoma.*

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 8 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 kelimer: *Kist hidatik; özofagus duplikasyonu.*

CASE REPORT

An 8-year-old, previously healthy male had been admitted to the hospital because of a 2-week history of abdominal distention and accompanying respiratory distress. Physical examination revealed a massive hepatomegaly. Computed tomography was then performed; the scans showed a cystic lesion located in the right lobe of the liver. Hydatid cyst was diagnosed, in spite of a negative result for the indirect haemagglutination (IHA) test. During treatment, the patient's respiratory distress worsened and he was transferred to our institution.

The result of laboratory investigation at our institution were normal with the exception of a high lactate dehydrogenase level. Liver function tests and serum assays for tumour markers yielded normal results. Abdominal ultrasonography revealed a large liver mass with a cystic appearance. Reassessment of older CT images showed a cystic lesion in the right medial and posterior liver sections. This lesion elevated the diaphragm and compressed the right lung.

Percutaneous liver biopsy was performed with the aim of differential diagnosis between tumour and hydatid cyst; we aspirated 50 cc of bloody fluid (different from hydatid cyst fluid) during this biopsy. While the aspirated fluid underwent cytopathologic study, the patient's respiratory distress rapidly worsened and he was transferred to the operating room.

During emergency surgery, a grossly cystic lesion with massive haemorrhagic areas was found. A tumour debulking operation (with every effort to avoid tumour spillage) was performed to reduce intraabdominal pressure. The specimens removed during the operation were evaluated with frozen section; the tissues were necrotic. In this emergency situation, no attempt was applied to remove the entire mass.

Histologic examination of the biopsy specimen after surgery demonstrated UES. CT images obtained after operation showed significant shrinkage of the tumour, and pulmonary or bone metastases were not present. The boy did not respond well to chemotherapy; consisting of vincristine, cisplatin, and actinomycin, magnetic resonance (MR) images showed no shrinkage of the tumour. Therefore, the patient underwent a complete tumour resection, consisting of a right hepatectomy without complications.

The tumour was soft, globular, and nodular in shape, with a gelatinous, glistening cut surface. It was well

demarcated from the liver, but not capsulated. There were multiple cystic areas with high degrees of necrosis and haemorrhage. Pathologic examination (during which the sarcoma cells were closely packed and arranged as sheets, or scattered loosely in a ground substance in mucopolysaccharide) revealed large areas of necrosis (90% of the tumour). One characteristic of the tumour (as with all sarcomas) was the presence of spherical globules of eosinophilic material, which are present both within the cytoplasm and in the intercellular matrix. Surgical margins were free of tumour cells.

DISCUSSION

When a cystic mass is identified in the liver, the presumptive diagnosis in most hospitals in endemic areas is usually hydatid cyst. Other potential diagnoses for the presence of cystic lesion in the liver in childhood (other than hydatid cyst) may include hepatoblastoma, hepatocellular carcinoma, metastatic tumour, mesenchymal hamartoma and resolving haematoma, and UES (1).

UES is a rare, highly malignant neoplasm of the liver. It is the third most commonly seen malignant tumour in the liver and occurs predominantly in childhood, usually between the ages of 6 to 10 years, although several cases have been reported in adults (2,4). The most common presenting symptoms in UES are abdominal mass with or without pain, fever, and weight loss (5,6).

Tumour markers are typically not found in the serum of these patients and therefore have no specific or diagnostic relevance; this fact may be helpful in ruling out other tumours (2,7).Although cystic liver tumours with normal α-fetoprotein levels should raise the possibility of a UES instead of hepatoblastoma, the negativity of α-fetoprotein in 10% of cases of hepatoblastoma and positivity around the same level in UES cases have also been reported (7). Indirect haemagglutination testing should be added to the protocol especially in areas endemic for hydatid cyst.

Ultrasonography is strongly suggested to confirm the cystic nature of the lesion during differential diagnosis to establish the presence of UES. The characteristics of UES on CT and MR images present as a "pure cyst"; this is related to the high water content of the myxoid stroma of the tumours (3,7). The pure cystic presentation of this lesion may lead to the misdiagnosis of hydatid cyst. Whenever possible, MR imaging is preferable to CT due to better visualisation of liquid areas within the matrix (8).

Treatment was based on complete surgical excision, which was possible in only 65% of cases reported in the literature (7). Long-term survival is possible after complete surgical resection with tumour-free surgical margins (7). No standard chemotherapy has been developed due to rarity of this lesion (1). Most patients were treated before surgery, with aggressive chemotherapy similar to regimens used for rhabdomyosarcoma; this type of treatment more often enables complete tumour resection of initially nonresectable tumours (2).

A delay of the diagnosis of UES has been reported in 12 cases (23.5%) in the literature because the pure cystic appearance suggested a benign lesion (7). In our case, the presumptive diagnosis was a hydatid cyst.

The diagnosis of hydatid cyst is arrived at through clinical findings, imaging techniques, and serology. Proof of the presence of protoscoleces should be obtained through microscopic examination of the fluid and histology (9). Indirect haemagglutination (IHA) can be used as a screening test (10). The sensitivity of serum antibody detection using indirect haemagglutination, which is more practical in the field, ranges between 85% and 98% for liver cysts, although false negative results are not rare (11). Eosinophil counts are also more greatly

elevated after rupture/leakage of cysts, but also can be seen in UES (12). Puncture, aspiration, injection, and reaspiration (or PAIR), which is minimally invasive, less risky, and usually less expensive than surgery, also confirms the diagnosis and removes parasitic material (13).

UES should be kept in the differential diagnosis due to the aggressive nature of this rare tumour when evaluating a cystic liver mass. Percutaneous biopsy can confirm diagnosis, and PAIR (which is also curative) can also confirm UES or hydatid cyst.

CONCLUSION

Because of its rarity, UES cannot be differentiated from hydatid cyst merely through laboratory tests and radiologic imaging techniques. In cases where UES is a possibility, or it seems that hydatid cyst may not be the correct diagnosis, we strongly advise percutaneous biopsy or PAIR in order to achieve accurate diagnosis.

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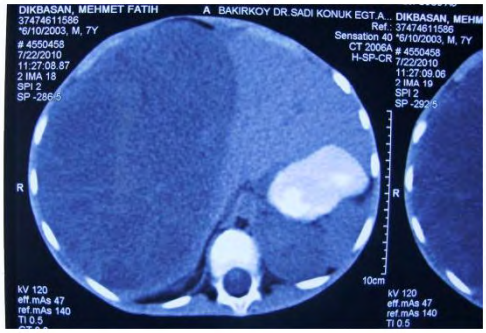

Figure.1 *Preoperative CT images*

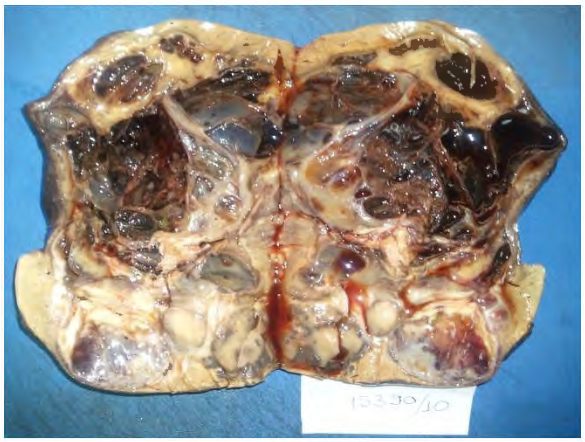

Figure.2 *Macroscopic view of the tumour.*