

# Hepatic Hydatid Disease: Radiographics Findings

## Karaciğer Hidatik Kist Hastalığı: Radyolojik Bulgular

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### Abstract

Hydatid disease (HD) is a unique parasitic disease that primarily affects the liver and is endemic in many parts of the world. There are four types of hydatid cysts (HCs) with various levels of organ involvement. All four HC types can be seen in the liver, with the right lobe being the most common site of involvement. There are many potential intrahepatic and extrahepatic complications that affect the liver and other organs. Intrahepatic complications include vascular and biliary complications and infection. With rare portal vein compression and decreased vascular supply, the involved lobe may show atrophic changes while the other lobe becomes hypertrophic. Intrahepatic biliary dilatation results from large cysts compressing intrahepatic ducts or due to cyst rupture into the biliary ducts. Cyst infection may also occur via a similar mechanism. Extrahepatic complications result from exophytic growth and hematogeneous dissemination. Exophytic growth causes migration of cysts into the lungs, mediastinum, heart and peritoneum. Hematogeneous dissemination involves all other organs.

**Keywords:** Hydatid disease imaging, Hydatid cyst, Liver, Imaging modalities

### Özet

Hidatik kist hastalığı, dünyanın birçok yerinde endemik olarak görülen bir hastalıktır. Öncelikle karaciğeri tutar. Farklı organ tutulumlarında saptanan 4 formu vardır. Dört formun dördü de karaciğerde görülebilir. En çok sağ lob tutulur. Hem karaciğer hem de diğer organları ilgilendiren birçok komplikasyonları vardır. Komplikasyonları intrahepatik ve ekstrahepatik olabilir. İntrahepatik komplikasyonları; vasküler ve safra yollarına ait olabileceği gibi enfeksiyon gelişimine de rastlanır. Nadiren portal ven basısına bağlı olarak akım azalabilir ve tutulan lobta atrofi karşı lobta hipertrofi oluşabilir. Safra yollarında genişleme 2 şekilde olur. Büyük boyut- taki kistler safra yollarına bası yapabilir veya kistler, küçük safra yollarına açılabilir. Aynı nedenlerle kistlerde enfeksiy- onda gelişebilir. Ekstrahepatik komplikasyonlar kistin ekzofit- tik büyümesine ve hematojen yayılımına bağlıdır. Egzofitik büyüyen kist akciğere, mediastene, kalbe ve peritona uzanır. Diğer organların tutulumu ise hematojen yayılıma bağlıdır.

**Anahtar Kelimeler:** Hidatik kist, Karaciğer, Görüntüleme yöntemleri

## Introduction

**H** ydatid disease (HD) is a common parasitic disease produced by the larval stage of the *Echinococcus* tapeworm. There are two types of echinococcus infection. *E. granulosus* is common in regions where animal husbandry is common (the Mediterranean, Africa, South America, the Middle East, Australia, and New Zealand). The eastern part of Turkey is an endemic region. *E. granulosus* is a more frequent cause of hydatid disease in humans than *E. multilocularis* [1-5].

In this paper, we discuss a variety of radiological and pathological findings in surgically- and laboratory-proven cases of hepatic hydatid disease at our hospital over the past 10 years. In addition to radiographic findings, we present the local complications of hydatid disease.

### Life Cycle of *E. granulosus*

Humans become infected by ingesting eggs from the tapeworm *E. granulosus* either by eating contaminated food or through contact with dogs. The ingested embryos invade the intestinal mucosal wall, enter the portal circulation, and develop into a cyst in the liver. Humans are intermediate hosts. In the liver, cysts grow to 1 cm during the first six months and approximately 2-3 cm annually thereafter, depending on the host tissue resistance [1,2].

### Cyst Structure and Radiographic Findings

The hydatid cyst has three layers: (1) the outer layer – the pericyst - consists of modified host cells: fibroblasts, giant cells and eosinophils, which form a fibrous and protective zone, (2) the middle, laminated, acellular membrane allows for nutrient passage, and (3) the inner germinal layer is thin. Scolices, the infectious embryogenic tapeworms, develop from an outpouching of the germinal layer.

The middle laminated membrane and the germinal layer form the true cyst wall, usually referred to as endocyst; the acellular laminated membrane is occasionally referred to as the ectocyst. The thicknesses of these layers depend on the tissue in which the cyst is located. The layers tend to be thick in the liver, less well developed in muscle, absent in bone, and sometimes visible in the brain [1-3].

### Types of hydatid disease and radiographic findings

#### Type I hydatid cysts

Type I cysts constitute the initial and active phase of hydatid disease. The three layers are intact. The external rupture of these cysts, due to either iatrogenic or traumatic causes, can cause disseminated disease. Type I cysts are important in disease spread to other anatomic sites in addition to the well-described hematogeneous and lymphatic route. The walls of ruptured type I cysts are generally imperceptible since they become infected and rupture. Ultrasound (US) reveals an anechoic, well-defined cystic lesion with small echogenic foci or “falling snowflakes” consistent with hydatid sands changing with patient position (Figure 1). Computed tomography (CT) detects a water-attenuating lesion with well-defined borders. Pseudo contrast enhancement at the cyst wall can be seen due to compressed host tissue (Figure 2).

True wall enhancement is seen in infected Type I cysts. Magnetic resonance imaging (MRI) reveals a homogeneous hypointense lesion on T1-weighted images and a homogeneous hyperintense lesion on T2-weighted images. The presence of a hypointense rim at the cyst periphery has been described as a characteristic of hydatid cysts (as opposed to non-parasitic cysts), but it is non-specific. This hypointense rim may be seen in long-standing Type I cysts due to a fibrotic response of neighboring host tissue or to slight calcifications within the cyst wall.

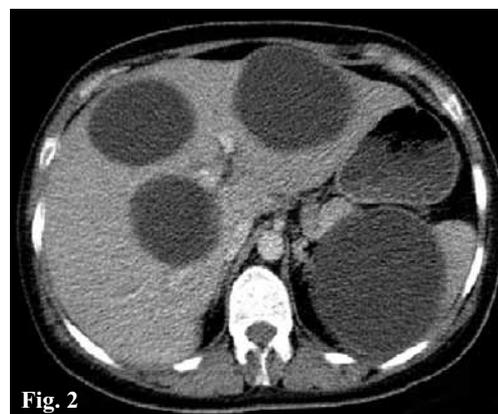
#### Type II hydatid cysts

Type II HCs represent the active phase of hydatid diseases in the parasite life cycle and in the dissemination of the

**Fig. 1—** Color Doppler imaging shows Type I HCs within the 7th segment of liver. Cyst is anechoic and has no perceptible wall.



**Fig. 2—** There are 3 type I HCs in liver and 1 Type 1 HC in spleen on axial late venous phase CT image. There is no contrast enhancement at the wall of the cysts.



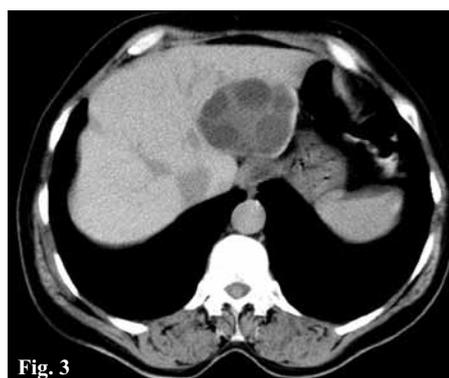


Fig. 3

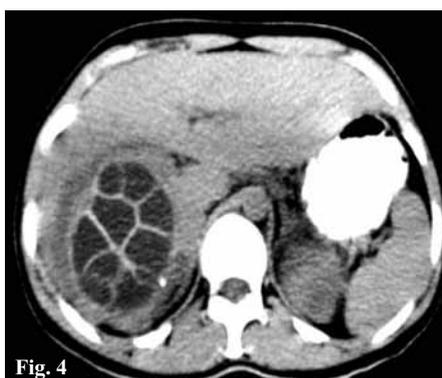


Fig. 4



Fig. 5

**Fig. 3—** Noncontrast-enhanced axial CT image reveals Type IIA HC at 4th segment of liver near the capsule. Note that daughter cyst arrange within the periphery of main cavity, mother cyst shows increased density relatively.

**Fig. 4—** There is a Type IIB HCs at 7th segment of liver with bizarre shape cysts almost occupying all cystic cavity on non-contrast-enhanced axial CT image.

**Fig. 5—** Non-contrast-enhanced axial CT image detects two Type IIC HCs. There is thick, circumferential calcification at the right-sided cyst. HC grows exophytically shows stippled calcification. Note daughter cysts in two HCs.

hydatid disease. Although Type I HCs cannot spread to host parenchyma other than by external rupture, type II HCs can be spread to nearby tissue by outpouching a new cyst from main cyst cavity. An hourglass appearance and additional type II HCs can be seen. This type of HCs can be classified into three stages according to the arrangement of daughter vesicles within the cyst cavity. In type IIA HCs, daughter cysts arrange at the cyst periphery. The CT density of the mother cyst is higher than the daughter cysts (Figure 3). MR imaging shows the daughter cysts as hypointense or isointense relative to the maternal matrix on T1- and T2-weighted images. Type IIB HCs contain larger, irregularly shaped daughter cysts that occupy almost the entire volume of mother cyst, creating a "rosette" appearance (Figure 4). Type IIC HCs are type IIA and type IIB cysts that contain scattered calcifications within the cyst wall and daughter cysts within the cavity. Scattered calcification at the cyst wall does not imply a dead cyst in the presence of daughter cysts, but simply degeneration at the cyst wall (Figure 5).

#### Type III: Calcified HCs

Type III HCs constitute the inactive or dead phase of HD. In this phase, HD cannot spread, and in the absence of mass effect or other complications, there is no need for surgery. These HCs are seen in three types: (1) Total and thick continuous calcification (ring-like) of the cyst wall (Figure 5), (2) total calcification within the cyst matrix and a decrease in cyst size (Figure 6), and (3) curvilinear calcification within the ruptured internal membranes (Figure 7). CT is the preferred imaging method to evaluate these types of HCs due to calcification.

#### Type IV: Complicated HCs

The complications of HCs can be seen in all types of HCs except completely calcified Type III cysts. The complications, explained below, can be due to profound cyst size and subsequent mass effect on neighboring organs. Other com-

plications include internal and external rupture of HCs, secondary site involvement due to invasion of various anatomic barriers (e.g., diaphragm) and superinfection. Ruptures can be seen in 50-90% of cases. Internal cyst rupture is detected by the detachment of the endocyst from the pericyst and is probably related to decreasing intracystic pressure, degeneration, host response, trauma or response to medical therapy and percutaneous drainage [1-3]. Internal rupture causes death of the parasite. In the acute phase of rupture, internal membranes can be seen as floating structures inside the cavity; this is called the "water lily sign" (Figure 8). With time, cystic fluid decreases, and the HC mimics a solid mass. Collapsed membranes within the cavity are detected as serpentine structures (Figure 9).

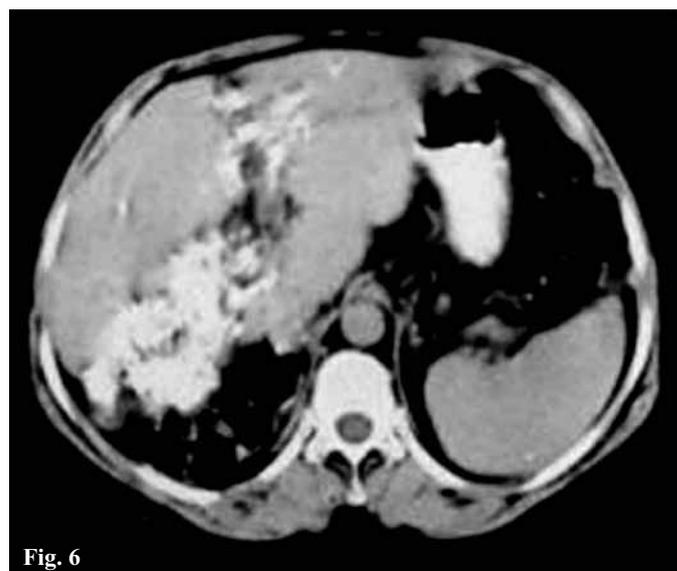


Fig. 6

**Fig. 6—** There is massive calcification and volume loss at the right hepatic lobe due to completely calcified Type III HCs.



Fig. 7



Fig. 8

**Fig. 7—** Contrast-enhanced axial CT section shows dense calcification at the wall and curvilinear calcification within the ruptured internal membranes.

**Fig. 8—** HASTE T2-weighted axial MR image reveals water-lily sign due to internal rupture of membranes. Rupture in this case is spontaneous. Cystic content is marked in newly ruptured HC.

### The involvement of various organs

Once the ingested embryos enter the portal circulation, they primarily affect the liver, but can be spread hematogenously to all organs and tissues except hair. Other organ involvement is generally secondary to liver involvement, especially for lungs, spleen and kidneys. In our patient population, isolated liver involvement was detected in 74.8%. Con-



Fig. 9

**Fig. 9—** CT reveals a solid lesion with stippled calcification at the periphery. Curvilinear isodense structure separated by slightly hypodense areas in consistence with germinal membranes can be seen in detail evaluation.

comitant liver and other organ involvement was detected in 26.9%. Another mode of spread other organs is cyst rupture into neighboring organs or peritoneum. The external rupture of the cyst is generally iatrogenic during cyst surgery or trauma. Severe abdominal trauma can decrease intracystic pressure and cause external and internal rupture of the HC.

### Hepatic Hydatid Disease

The liver is the most commonly involved organ with the right lobe being most frequently affected. All types of HCs can be seen within the liver. Type I HCs become especially important when they exert a mass effect. The diagnosis is relatively easy in endemic regions and in solitary HC. However, a simple liver cyst along with a solitary HC as well as polycystic liver disease in the presence of multiple type I HCs can cause diagnostic problems. Living in endemic regions is important clue along with laboratory findings.

#### The complications of hydatid disease in liver

##### Mass effect

One of the most important complications occurs when HCs (type I and II) reach large sizes due to an active growth phase. Large type I and II HCs can compress neighboring tissue and cause mass effect. If there is compression of vascular structures, especially portal vein, the involved lobe becomes atrophic due to decreased portal venous supply, and compensatory hypertrophy of the other lobes results. This is especially true for *E. alveolaris* due to microinvasion of portal veins and small biliary ducts. However, we also detected compensatory hypertrophy of uninvolved lobes and atrophy in the involved lobe without complete biliary obstruction (Figure 10).

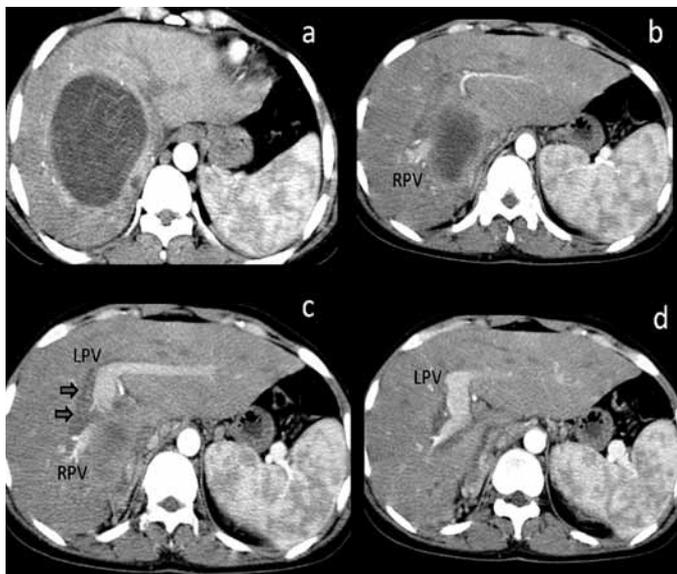
Large cysts in the hepatic parenchyma can cause biliary duct dilatation by either compression of a nearby duct by mass effect or by perforation into biliary ducts. The compression of nearby biliary ducts can cause microerosion within the bile duct wall and fistula formation. There can be wide perforation that allows cystic contents to spill within the biliary ducts. Hydatid sands in Type I HCs, daughter cysts in type II HCs and ruptured germinal membranes in type IV HCs (Figure 11) may pass through the fistula or perforation, obstruct bile flow, dilate biliary ducts and cause jaundice.

In the presence of a small fistula, it is difficult to show direct

communication between cyst and biliary tracts by imaging methods. MR cholangiography may show small biliary ducts entering in the HCs. The detection of dilated biliary ducts and echogenic material within the lumen may be suspicious in the presence of HCs by ultrasound. A ruptured internal membrane floating freely within the cystic cavity may pass through the communication between HC and biliary tract. This may be seen as tubular structures within the biliary lumen. When daughter cysts pass through a direct communication, a cystic lesion may also be seen within the biliary lumen.

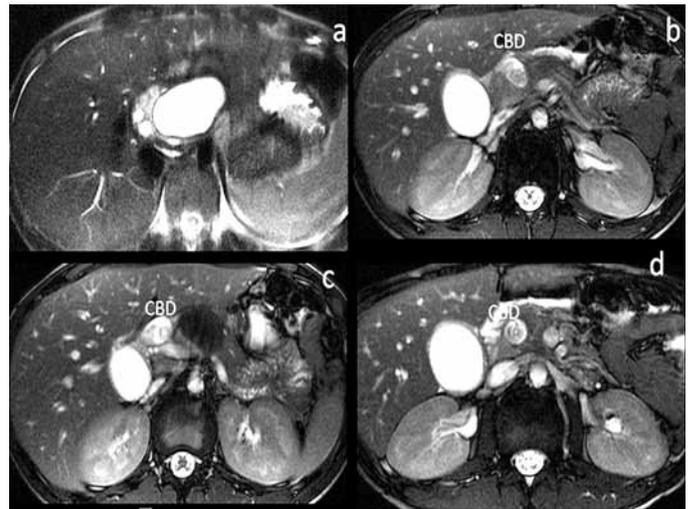
#### Rupture of HCs

The rupture of HCs can be contained (internal), communicating, and direct. Contained or internal rupture occurs when the detachment of the endocyst from the pericyst takes place, but the pericyst remains intact. Internal rupture may be



**Fig. 10—** Axial portal phase continuous CT images reveal HCs with internal, communicating rupture compress right portal vein (RPV). Note compensatory hypertrophy of left portal vein (LPV) and left lobe of the liver extending to the left site of the abdomen. There is also minimal dilatation in intrahepatic biliary tract (arrows). It was surgically proven that there were communicating biliary canalicula with cyst.

related to degeneration, trauma or response to therapy, and it represents the inactive phase of the HCs. Communicating rupture implies passage of cystic contents into biliary tracts. Communicating rupture may be seen in type I, II and IV HCs that internally rupture. When both the pericyst and endocyst rupture, direct rupture occurs. Internal rupture of cyst implies the death of the parasite, but external rupture causes dissemination to other organs. Type II HCs can only rupture externally. Trauma, degeneration and iatrogenic rupture due to surgical and percutaneous treatment cause external rupture of type I and II HCs (Figure 12). Sudden death, anaphylactic



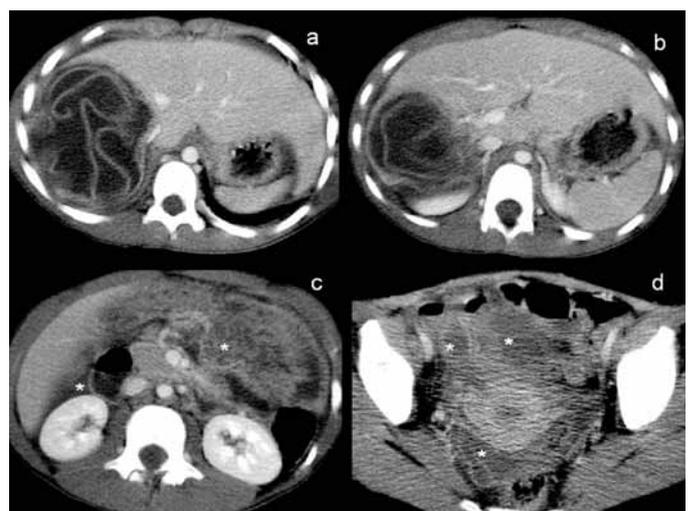
**Fig. 11—** HASTE T2-weighted axial continuous MR images of a patient applied with the complaints of jaundice shows a Type IIa HC at caudate lobe. There are twisted internal membranes within the lumen of common bile duct.

shock and dissemination of disease can be seen with cystic content spillage into the peritoneal cavity.

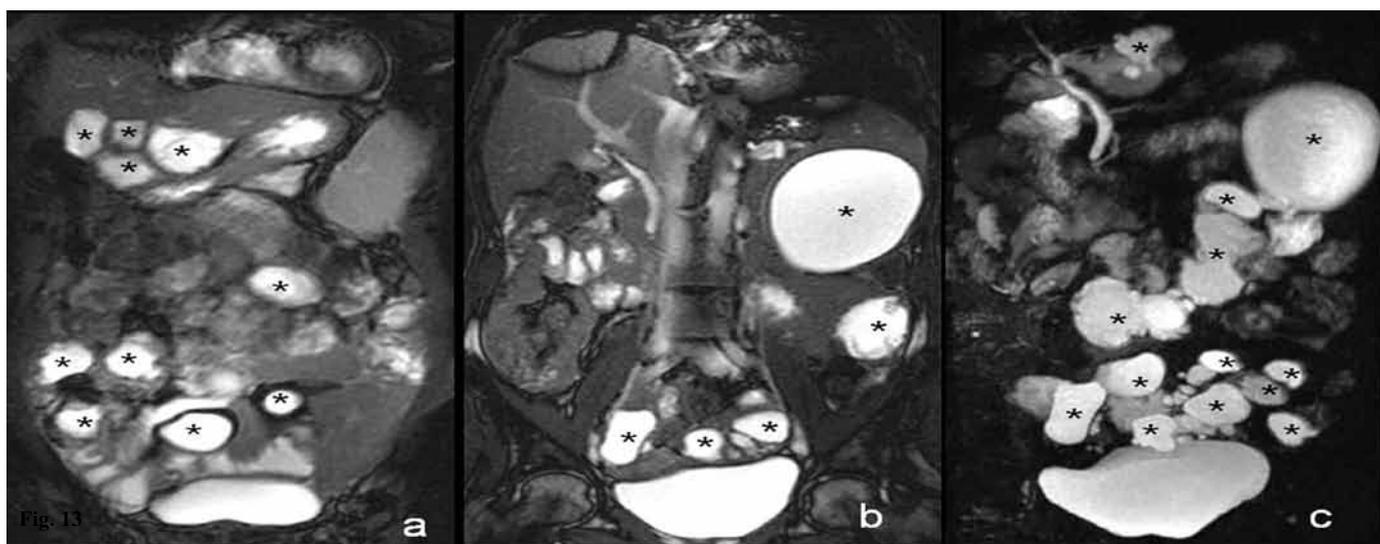
When type I and II HCs are found near the hepatic capsule, extension to other neighboring organs is easier. Rupture can also be seen in these locations due to a lack of protective tissue surrounding the cyst and deficient pericyst. Spillage of cystic content within peritoneal cavity causes disseminated disease (Figure 13).

#### Infection of the HCs

The presence of air within the cystic cavity is also an important clue. Cyst infection is generally seen in HCs that rupture. Rupture may be internal (communicating) or direct.



**Fig. 12—** Type I HC located at the 7th segment of liver shows internal and external rupture on continuous CT images. There is free interperitoneal fluid in perihepatic, omental and pelvic areas (asterisk). Rupture in that case was due to trauma.



**Fig. 13—** HASTE T2-weighted MR cholangiography (a-b) and MIP (c) images show multiple HCs grow exophytically at the visceral surface of left lobe (asterix). There is disseminated echinococcus disease in the peritoneal cavity.

Rupture of HCs permits bacteria to pass easily into cyst. The wall becomes thicker and exhibits contrast enhancement on CT and MR imaging. Patchy contrast enhancement of neighboring liver parenchyma represents inflammatory changes. CT is superior in detecting gas or air-fluid levels within the cyst (Figure 14).

#### Exophytic growth

The two most common routes of exophytic growth are via the bare area of the liver and gastrohepatic ligament. HCs extend to lung and mediastinum when located at the bare area of liver. The cyst in located near the gastrohepatic ligament can extend into peritoneal cavity (Figure 13).

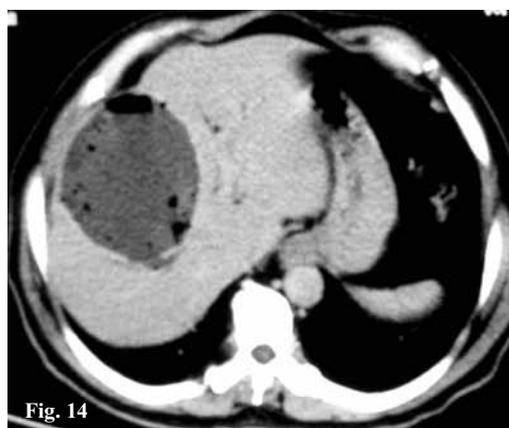
#### Transdiaphragmatic lung, mediastinum and cardiac involvement

Transdiaphragmatic involvement of lungs and mediastinum occurs in 0.6- 16% of hepatic hydatid cysts. Transdia-

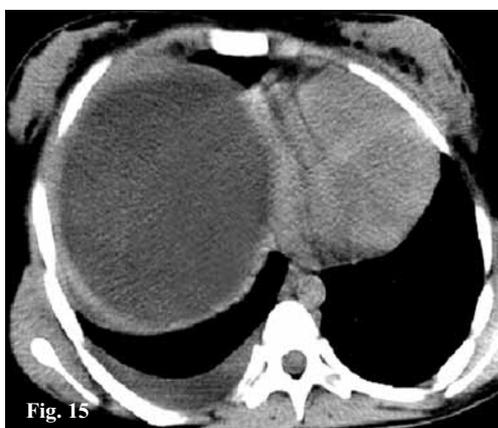
phragmatic migration is most common in HCs located in the 7th and 8th segments of right lobe due to their close proximity to the diaphragm. All HC types can be spread in this way including completely calcified cysts that migrate into the thoracic cavity by adherence to the diaphragmatic surface.

When a HC is located in close proximity to the diaphragm and grows large, initial indirect or reactional findings may be seen on imaging methods, including pleural effusion, atelectasis or consolidation due to decreased diaphragmatic motion and incomplete lung expansion. Elevation of the diaphragm due to mass effect may also be seen with large HCs (Figure 15).

A direct sign of transdiaphragmatic migration is the presence of cystic lesion growing through diaphragm into lung, mediastinum and heart (Figure 16). The cyst typically has a characteristic hourglass shape. Sagittal and coronal MR images are helpful. Migration to the lung is frequent in HCs located in the 7th to 8th segments of the liver while medias-



**Fig. 14**



**Fig. 15**

**Fig. 14—** Non contrast-enhanced CT section shows a hypodense lesion at the 7th segment of the liver with air in it. Surgery revealed its communication with biliary tract. Cyst contained ruptured internal membranes and purulent material.

**Fig. 15—** Type I HC at the liver dome reaches big size, displace the heart to left and cause pleural effusion on CT.



**Fig. 16**— Scanogram shows increased density within the right hemithorax (a). Continuous axial contrast-enhanced CT sections (b-d) reveal Type I HC at the liver dome. Cyst migrate into right lung transdiaphragmatically.

tinal and cardiac involvement is associated with the 4th and 2nd liver segments.

Migration of HCs into lung from liver is seen as a continuation of the main cyst in the liver. Erosion or adhesion to the diaphragm is the cause of cyst migration. If cyst erodes the bronchial wall, as seen in primary lung HCs, internal perforation is considered. Expectoration of cystic membranes can occur with a bronchial fistula. Direct perforation of HCs into lung parenchyma causes prominent parenchymal consolidation and widespread hydatid disease. Perforation of HCs into the pleural cavity causes pleural empyema or multiple pleural cysts.

#### Peritoneal seeding

Peritoneal HD is almost always secondary to hepatic disease. Peritoneal echinococcosis in our series was generally due to previous hepatic surgery or spontaneous or traumatic rupture. Spontaneous rupture is a possibility with HCs located near the hepatic capsule or gastrohepatic ligament that grow exophytic. Peritoneal hydatid disease is generally multiple and remains undetected until cysts are large enough to produce symptoms (Figure 13). The overall incidence of peritoneal disease in cases of abdominal HD is approximately 13%. CT and MR imaging is valuable by imaging the entire peritoneal cavity. All kinds of HCs can be seen in the abdominal cavity.

**Conflict interest statement** The authors declare that they have no conflict of interest to the publication of this article.

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