Chapter

Imaging in Echinococcosis

Tuba Apaydın

Abstract

Hydatid disease is a widely common parasitic disease caused by the species; *Echinococcus granulosus* and *Echinococcus multilocularis*. Echinococcosis is a serious medical, economical and social handicap all over the world. Almost any organ of the body can be involved with the zoonosis, liver and lungs are the most frequently involved organs. Imaging has a critical role for diagnosis of the disease. Radiography is the first imaging tool. Although CT aids the detection of size, number, location and imaging of local complications for diagnosis of hydatid cysts, sometimes atypical lesions rise in the imaging tools. MRI may help for differentiation of hydatid lesions from other neoplastic and hypodense infective lesions. MRI is also better for biliary and neurological involvement. Ultrasound is especially helpful for abdominal hydatidosis and peripheral lung lesions. Regarding that clinical evaluation is difficult for hydatidosis; radiological findings should be evaluated well for accurate diagnosis. This chapter will focus on imaging of echinococcosis.

Keywords: *Echinococcus*, MRI, CT, USG, hydatid cyst

1. Introduction

Hydatid disease is a worldwide parasitic infection created by larval phase of *Echinococcus* [1]. Human infection is frequently seen in Europe, Middle East, some places of Canada, Russia, Japan, China, Australia and New Zealand [2].

In the life period, *E. granulosus* stands in the small bowel of its definitive host; carnivores, which are generally dogs and wolves [3]. Then, eggs passed into the feces are ingested with the intermediate host, which is generally sheep [4]. Larval stage occurs in the intermediate host; i.e., ingested eggs outgrows in the small intestine and lets out oncosphere, which invades the intestinal mucosa and come in the lungs, liver or other organs (metacestode larvae). Eggs can survive up to 1 year in the environment. On the next step, definitive host digests the infected organs of the intermediate host. Protoscolices invades the intestinal mucosa and grow up to adult worms [3]. Human being can be more commonly infected by indirect ingestion of contaminated water and food or directly from contact with dogs [4].

Fluid filled cyst is covered with three layers; pericyst, ectocyst and endocyst [5]. Pericyst is created by the protective reaction of the host tissue; the middle laminated layer called ectocyst lets for the transition of nutrients and the germinal layer inside called endocyst generates cyst fluid, blood capsules, scolices and also provides the constitution of ectocyst [5, 6]. Many daughter vesicles remain in the hydatid cysts [6].

Liver is the most commonly affected organ, second one is the lung. Other organs are less commonly affected. Heart is rarely affected, but it's fatal if it's affected. Left ventricule is more commonly affected than the right ventricle possibly due to its

richer blood supply. Additionally, huger myocardial mass in left ventricle supplies better circumstances for parasite's growth. Hydatid cysts of left ventricle are generally located in the subepicardium. Rupture into the pericardial space is seldom. However, habitation in the right ventricle is subendocardial, rupture is more common which results with anaphylaxis, pulmonary embolisation and death [7].

Most common way for myocardial invasion is coronary circulation. Second most common route is pulmonary venous drainage with the rupture of pulmonary cysts. Heart may also be involved with direct contact [7].

Hydatid disease has an evolutionary period; first, cysts grow slowly; then, a differentiation period starts which the parasite dies and forms calcified, solidified cyst behind [7].

The lungs are the most common organ in pediatric patients and the second most common organ for adults. Due to the negative pressure inside the lungs, cysts grow three times faster compared to the growth in the liver [6].

E. granulosus has a broad genetic diversity. Molecular gene analysis has defined 10 genotypes, which are aggregated into four different types: *E. granulosus* sensu strict [G1-G3 complex], *Echinococcus equinus* [G4], *Echinococcus ortleppi* [G5] and *Echinococcus canadensis* [G6-G10 complex]. G1 genotype of *E. granulosus* is reported to infect humans more frequently compared to other subtypes. Genotyping of human CE can help to plan the controlling methods for human hydatidosis. Genetic subtyping also clarifies the diversity in development, antigenicity and response to chemotherapeutic agents [3].

The disease is asymptomatic in the inception and can remain asymptomatic for long years even though the cysts become very huge [1]. For symptomatic patients, symptoms appertain to localization of the cyst. Cough, dyspnea or chest pain can be seen if the cysts are in the lung. Abdominal pain, hepatomegaly, sensibility, fever and icterus are the symptoms for liver cysts. With the rupture of the cyst due to the surgical intervention to the cyst or mechanical trauma, patients are prone to go into anaphylactic shock [1]. Cyst rupture can realise throughout the bronchus and patients can present with cough and sputum with hydatid sand and membrane fragments. If the cyst rupture realises throughout the pleural cavity, patients can present with pneumothorax, effusion and emphysema [6]. If the cyst rupture through vena cava, patient can present with recurrent pulmonary embolism [6].

Cysts and cyst like hypodense lesions carry diagnostic debate on computed tomography (CT) in some cases. Tumors with cystic degeneration, inauspicious lesions like necrotic lung cancer, metastases and infections like tuberculosis can imitate hydatid cyst [8].

Diagnosis is quite elementary for hydatid cyst (HC) if typical findings like crescent sign, onion peel, combo sign or folded membranes are present. However, atypical findings like solid or more hypodense semblance of ruptured, collapsed or infected cysts are more complicated as they resemble infections like tuberculosis or neoplastic lesions. MRI may help for diagnosis in these conditions [8]. Ultrasound can help to diagnose peripheral lesions and to achieve pleura [6].

2. Imaging in echinococcosis

Imaging tools for hydatid disease are computed tomography (CT), ultrasonography (USG), magnetic resonance imaging (MRI), radiography and urography.

2.1 Radiography

Radiograph is the primary method for imaging in bone and lung disease. An uncomplicated hydatid cyst looks like a well-limited homogenous radio-opacity on

chest X-ray (**Figure 1A**). Cysts quietly resemble carom balls in posteroanterior X-ray and to rugger balls in lateral X-ray [6]. Cysts may look like a strange shaped mass due to pressure of mediastinum, bronchovascular components. Multiple large cysts are also pathognomonic for echinococcosis (**Figure 1B**) [1]. Cysts can present with bilobed appearance due to nicking inside the cysts [6]. The loss of a round shape with a small depressed view points out bronchial rupture, so called "slit sign".

Atelectatic and reactive reactions can result with the loss of well limited borders, thus imitating carcinoma or pneumonia [6].

A radiolucent frame may be seen with the entry of air between pericyst and endocyst due to disruption of bronchus, so called "crescent sign" (**Figure 2**). This sign is not pathognomonic for hydatid cysts and may also be seen in carcinoma, blood clots, mycetoma, and Rasmussen aneurysm. If the entry of air increases, endocyst minimizes and ruptures; an air fluid level is observed in the endocyst, so-called "double curve sign". The natant membranes in the cyst fluid compose "water-lily sign" if there is furthermore collapse of the endocyst. Daughter cysts may look like circular radio-opacities at deep part of the cysts, so-called "rising sun" image. Pericyst can become empty if the membranes are extracted with cough, air-filled cysts can be seen on X-ray, so-called "dry cyst sign". In cases of infection added to the disorder, lung abscess can be imitated (**Figure 3**).

X-rays for abdomen may present with hepatomegaly, elevation of right hemidiaphragm and cyst wall calcification. During healing period of the cyst, all the structures in the cyst calcify and plain radiograph presents a dense calcified mass [5].

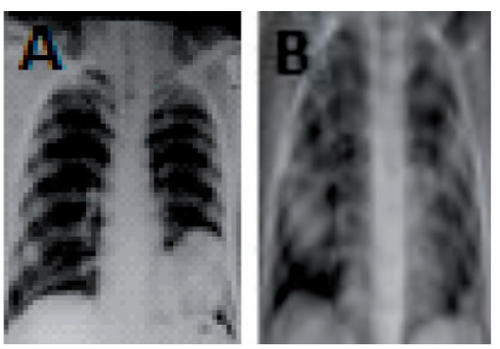

Figure 1.
(A) Posteroanterior view of chest X-ray presenting uncomplicated hydatid cyst of left lower lung and (B) posteroanterior view of chest X-ray presenting multiple, large, circular and well limited masses in both hemithoraces.

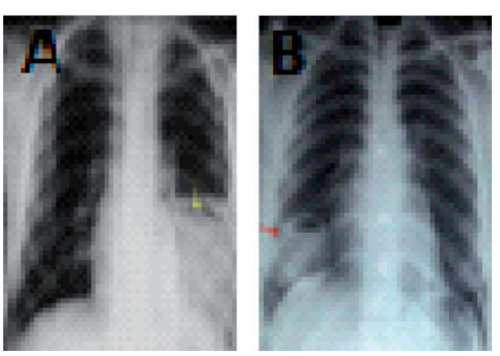

Figure 2.
(A) Chest X-ray showing pulmonary meniscus sign (arrow) indicating crescent shaped containment of air and (B) chest X-ray presenting a well limited lesion (arrow) in the right lower lobe of the lung with air fluid level, indicating a superimposed infection.

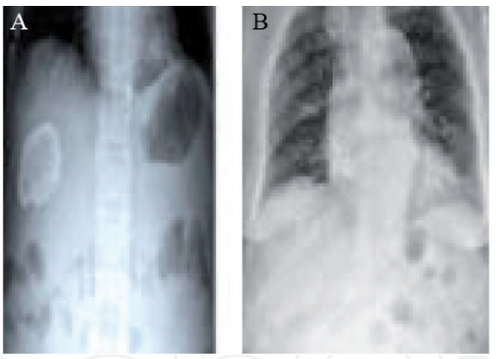

Figure 3.
(A) Abdominal X-ray showing round calcified mass in the right hypochondrium and (B) chest radiograph showing hydatid cyst in the liver.

Figure 4.
Transverse ultrasound of the retroperitoneum showing unilocular anechoic cyst (type CL according to WHO classification).

2.2 CT and ultrasonography in hydatid cysts

Cysts don't always present classical signs above. Obstacles about diagnosis can be overlapped with the help of CT in required cases [6]. CT is a significant diagnostic tool in detecting cyst wall or septal calcification, osseous lesions, cystic component localized posterior to calcification, evaluating complications and in cases which USG is not enough (abdominal wall deformities, excessive bowel gases, obesity and previous surgery) [4].

US is easily accessible and lacks radiation. It's sensitivity is about 100% acquiring it the priority as a screening method for abdominal hydatidosis. USG and MRI are both successful to show hydatid sand, daughter cysts, natant membranes and vesicles inside the cyst (**Figure 4**) [1, 4].

Mobile hydatid sand which points out to the splitted capsules and scolices that moves in the cyst cavity may be presented in a "snowstorm" appearance (**Figure 5**). Most commonly used classifications related to sonographic appearance are classifications of Gharbi et al. (**Table 2**) and the World Health Organisation Informal Working Group classification on echinococcosis (WHO-IWGE) (**Table 3**) [5].

Gharbi type 1 is the most common subtype and it's presented as a pure cystic lesion with or without the entity of hydatid sand (**Figures 4** and **5**). Gharbi type II cysts grows out after trauma, cyst degeneration, host response or drug therapy. Decrease in the

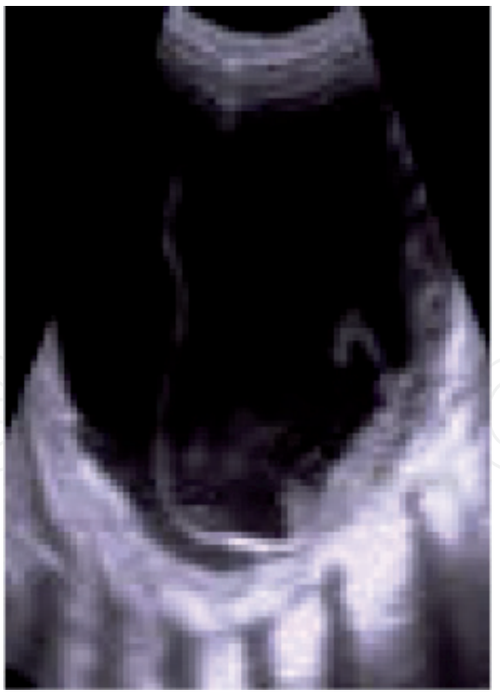

Figure 5.
Longitudinal ultrasound of a liver hydatid cyst in a 8 years old girl presenting the "snowstorm appearance".

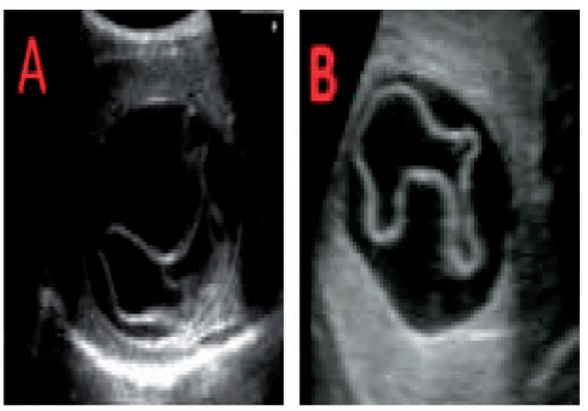

Figure 6.
Ultrasonographic view of Gharbi type II hydatid cysts: (A) "Split wall" sign from splitting of pericyst and endocyst and (B) "water-lily" sign resulting from complete splitting of the membranes.

intracystic pressure results with splitting of the endocyst and pericyst; natant membranes may be seen in the cyst cavity. Complete splitting of the membranes is called as "water-lily" sign due to its morphological appearance (**Figure 6**). Gharbi type III cysts are multivesicular cysts which septae presenting the neighbour borders of daughter cysts composes a "honeycomb" image (**Figure 7**). "Spoke wheel" appearance may be seen when daughter cysts are splitted with hydatid matrix. Gharbi type IV cysts are composed of an internal echogenic matrix giving them a solid appearance (**Figure 8**). For differential diagnosis, daughter cysts or membranes should be searched to externalize a solid mass. Gharbi type V cysts involve wall calcification and dense distal acoustic shadowing (**Figure 9**). A densely calcified cyst can be supposed to be dead. However, a partially calcified cyst should still be regarded as an alive cyst [5].

Imaging findings for hydatid cyst differentiate from cystic lesions to solid appearing lesions. The cyst may look like a well identified fluid accumulation. There may be the appearance of natant membranes due to cleavage of endocyst from pericyst. Ring like calcification of the cyst can lead to complete calcification during differentiation of the cyst [2].

Figure 7.
Transverse ultrasound of Gharbi type III hydatid cyst of liver multiple daughter cysts in an echogenic matrix results in a "honeycomb" image.

Figure 8.
Ultrasonographic appearance of Gharbi type IV hydatid cyst of the liver.

Specific findings of imaging are visualization of daughter cysts, calcification of the cyst wall and membrane detachment. Diagnosis in early stage is hard [1].

Appearance of hydatid disease may be classified into four subtypes.

Type I is simple cyst with no internal tectonic. On ultrasound, type 1 hydatid cysts seem as well identified unilocular anechoic lesions (**Figure 10**). On CT, they seem as fluid accumulated lesions. MRI demonstrates a fluid accumulated cystic lesion with a T1 isointense and T2 hypointense peripheral border (rim sign) enclosing the homogenous high signal cyst ingredients [2].

Figure 9.
Ultrasonographic appearance of Gharbi type V hydatid cyst.

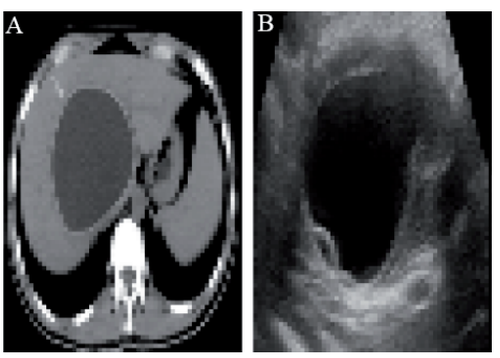

Figure 10.
Type 1 hydatid cyst: (A) axial contrast CT image of a 15 years old female presents a well identified fluid accumulated simple cystic lesion in the liver and (B) oblique sonogram in a 45 years old male demonstrates type 1 hydatid cyst in the liver.

Type II hydatid cysts are cysts with daughter cysts and matrix. They include cleaved natant membranes or daughter cysts (**Figure 11**). Attenuation of the daughter cyst is hypodense/hypointense to maternal matrix on CT and MRI, respectively. If there are multiple cysts, they are covered with a single capsule presenting with a "truckle spoke" appearance [2].

Type II HC are classified to three subtypes according to the age, quantity and setting of the daughter cysts.

Type IIa contain involve daughter cysts organized at the periphery.

Type IIb involve bigger daughter cysts with irregular borders that covers almost all of the capacity of the mother cyst.

Type IIc contains high attenuation round or oval masses with sprinkled calcifications and daughter cysts, demonstrating attrition of the old cyst.

Type III cysts are dead calcified cysts (**Figure 12**).

Type IV hydatid cysts are complicated cysts (**Figure 13**). Most common complications of echinococcosis are rupture and superinfection. Degeneration of parasitic membranes causes the rupture of the cyst (**Figures 14–18**).

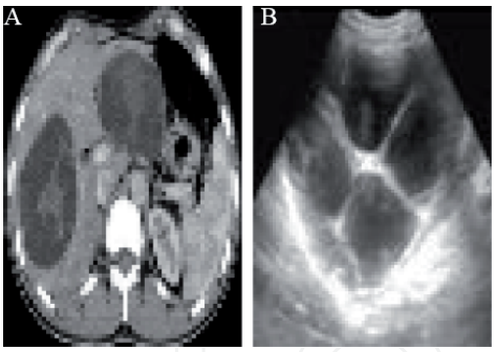

Figure 11.
Type 2 hydatid cyst: (A) contrast enhanced computed tomography demonstrates multiple daughter cysts with irregular borders covering almost all of the volume of the mother cyst-so called "rosette appearance" and (B) oblique ultrasonogram in a 39 years old male demonstrates type 2 hydatid cyst.

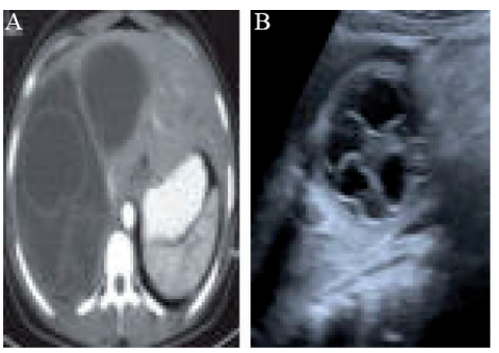

Figure 12.
Type 3 hydatid cyst: (A) contrast enhanced computed tomography presents type 3 hydatid cyst in the right lobe of the liver and (B) oblique ultrasonogram shows anechoic lesion with membranes floating on the cyst fluid.

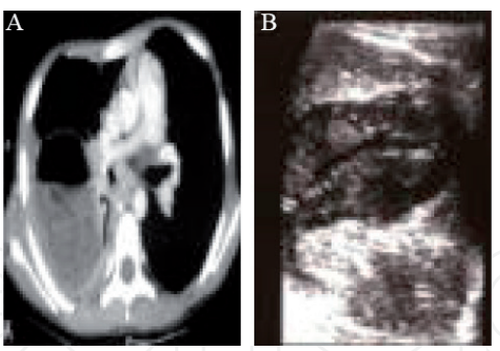

Figure 13.
Type 4 hydatid cyst: (A) sagittal contrast enhanced CT images of thorax show ruptured type 4 hydatid cyst in the lower lobe of the right lung with wrinkled floating endocyst-so called "lily sign" and (B) oblique ultrasonogram shows type 4 hydatid cyst.

Various secondary complications possibly happen according to site of rupture in the body. Secondary complications of echinococcosis according to different locations are listed in **Table 1**.

Gharbi et al. preferred classification for differentiation of subtypes of hydatid cysts on ultrasonogram of the liver (**Table 1**) [5].

Another global classification used for sonographic imaging of hydatid cysts wad presented by World Health Organization Informal Working Group (**Table 2**) [5].

Type	Description
Type I	Pure fluid accumulation
Type II	Fluid accumulation with splitted membrane
Type III	Fluid accumulation with septa and/or daughter cysts
Type IV	Heterogenous echo pattern (hyperechoic with high internal echoes)
Type V	Cyst with calcified thick borders

Table 1.
Gharbi et al. classification of hydatid cysts related to ultrasonographic features [5].

Type	Description
Type CL	Unilocular, cystic lesions with uniform anechoic ingredient, not absolutely confined with hyperechoic frame.
Type CE1	Unilocular, simple cyst with uniform anechoic ingredient. Cyst can present with echoes because of moving capsules, so called ‘’hydatid sand’’(flack sign)
Type CE2	Multivesicular and multiseptated cysts; cyst septations cause tire like architecture, daughter cysts present ‘’rosette-like’’ or ‘’honey-comb’’ like architecture.
Type CE3	Anechoic ingredient with splitted laminated membrane from the cyst wall seen as natant membrane or as ‘’waterlily sign’’ which points out more membranes natant in the cyst fluid.
Type CE4	This may present with a ‘’ball of wool’’ sign presenting retrograded membranes.
Type CE5	Thick calcified wall of the cyst is belt shaped and produces a cone shaped shadow.

Table 2.
Types of hydatid cysts observed on ultrasound examination of liver [5].

2.3 Magnetic resonance imaging in hydatid cysts

MRI is better to show cyst wall defect, biliary and neurological involvement. Cysts are hyperintense on T2W images and are covered by a low signal frame possibly because of the collagen rich pericyst [1]. If available, daughter cysts are hypointense compared to the intracystic fluid on T1 weighted imaging and hyperintense on T2-weighted images [5]. DW MRI makes the differentiation of CE1 hydatid cysts from other simple cysts with their hyperintense image. Apparent diffusion coefficient (ADC) of the hydatid cyst is lower compared to ADC of simple cyst because of internal viscous contents [1]. Also, ADC values of simple cysts and type I to III hydatid cysts are higher than ADC values of abscesses because diffusion of protons throughout thin fluid is limited. ADC values of type IV hydatid cysts and abscesses present no considerable statistical difference [5].

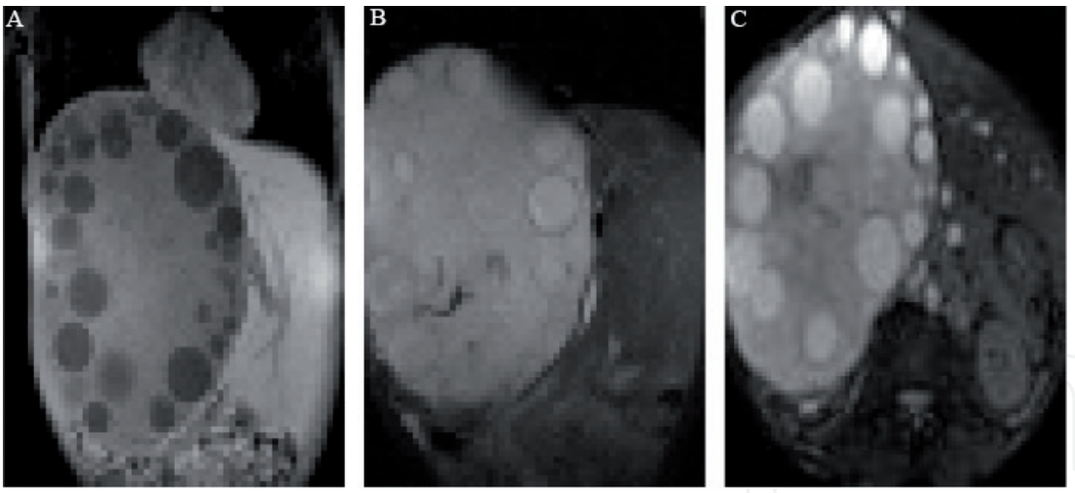

Figure 14.
MRI showing hydatid cyst in the liver of a 45 years old male: (A) coronal T1 weighted image, (B) coronal T2 weighted scan and (C) axial T2 weighted scan.

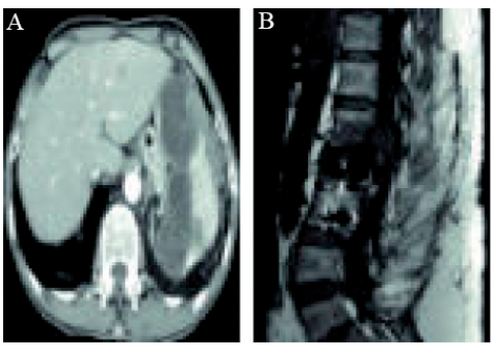

Figure 15.
(A) Axial T2 weighted MRI showing splenic hydatid cyst and (B) coronal T2 weighted scan of hydatid disease in L4-5 vertebra.

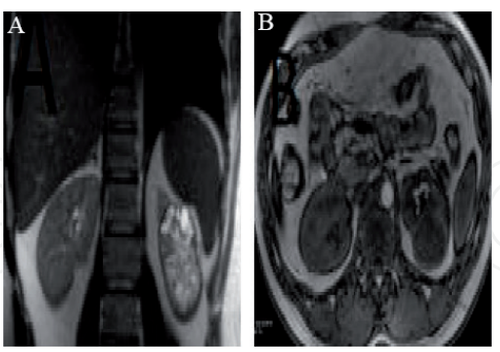

Figure 16.
MRI showing hydatid cyst in the lower pole of the kidney: (A) coronal T1 weighted scan and (B) axial T2 weighted scan.

MRI demonstrates the degree of cyst degeneration with secession of the wall, collapsed membranes are observed as bent linear compositions inside the cyst. Wall calcification is demonstrated better on MRI compared to CT, besides MRI is more successful than CT to represent irregularities of borders that points out inchoative segregation of membranes [5].

MRI is also better to differentiate liver hydatid cysts from other simple cysts [4] (**Figures 14–18**).

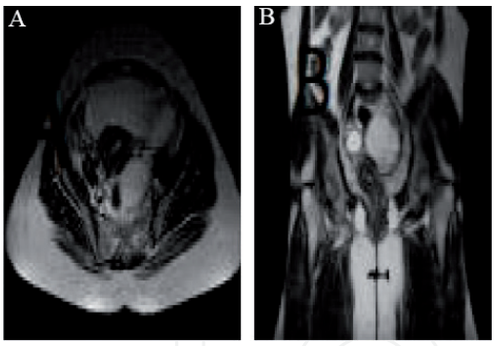

Figure 17.
MRI showing hydatid cyst in the cyst hydatid in the mesosalpinx adjacent to the left ovary: (A) axial T2 weighted image and (B) coronal T2 weighted image.

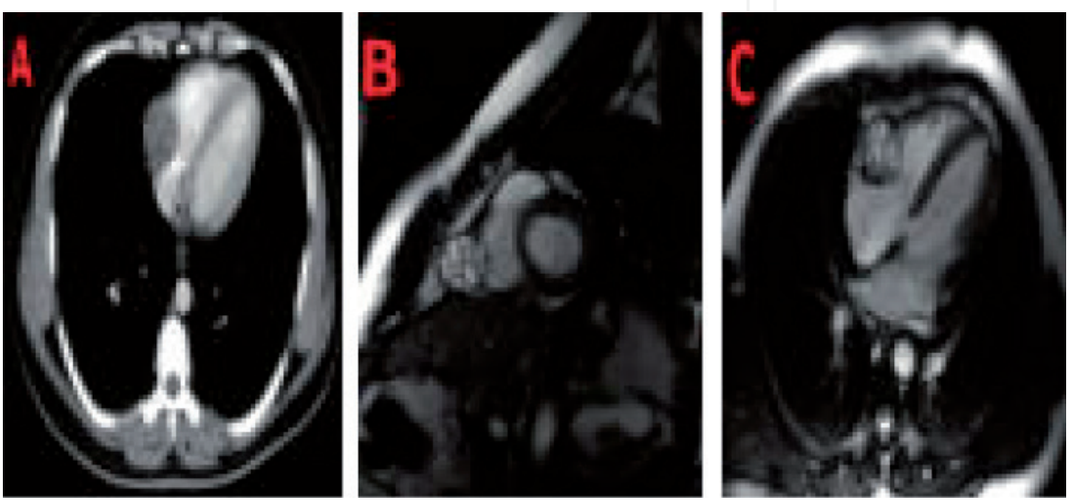

Figure 18.
Case of cardiac cyst hydatid: (A) computed tomographic image, (B) magnetic resonance imaging of transverse section, and (C) magnetic resonance imaging of longitudinal section showing daughter cysts.

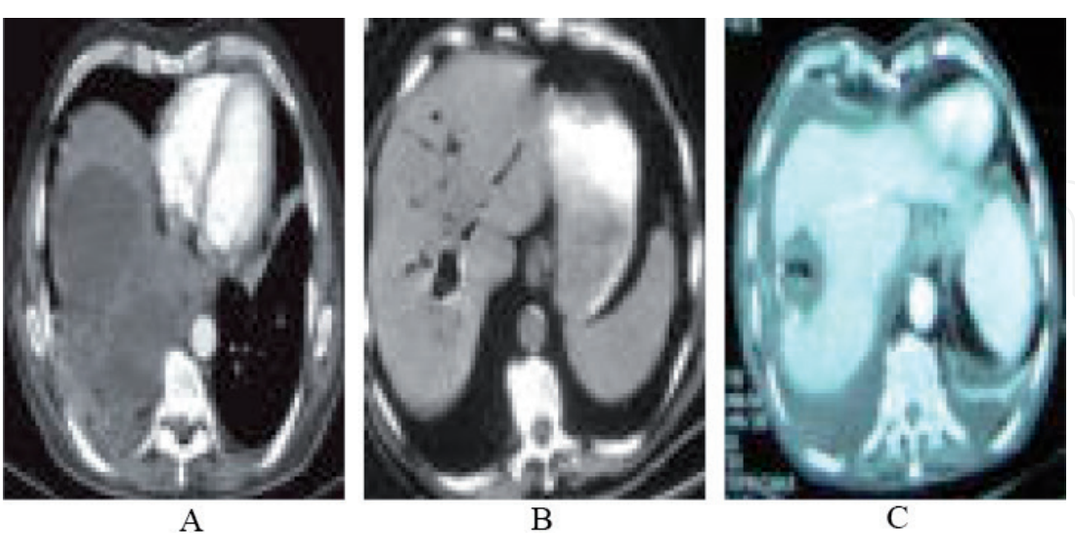

Figure 19.
(A) CT image of rupture of hydatid cyst in the lung, (B) CT scan of intrabiliary rupture of hydatid cyst. (C) CT scan showing intraperitoneal cyst rupture with diffuse peritoneal effusion.

In cardiac cystic echinococcosis, trans-thoracic echocardiography, CT and MRI can demonstrate the cystic structure of the lesion and it's correlation with the cardiac chamber [8] (**Figure 18**).

Location	%	Complications
Liver	71.9	Cyst rupture, liver abcess, inferior vena cava compression and thrombosis, portal hypertension, Budd Chiari syndrome, cholangitis, pancreatitis, biliary peritonitis, peritoneal spread
Lung	19,6	Pneumothorax, lung abcess, biliptysis, lung embolism, pneumonia
Peritoneal &pelvic cavity	3,6	Mass effect
Spleen	2,15	Rupture into peritoneal cavity
Skin&muscles	1,2	Pain, movement disorder
Bones	0,65	Morphologic deformities, pain
Brain	0,56	Seizures, headache, tumor like symptoms

Table 3.
Frequency and location specific complications of hydatid disease [2].

ERCP and MRCP show cystobiliary relationship, daughter vesicles & germinative membranes of cysts in bile ducts, dilated bile ducts. However, because of high intracystic pressure, communication of cyst with bile ducts can't be shown via ERCP& MRCP effectively [1].

2.4 Imaging of complications

Complications like rupture into the peritoneal cavity, biliary cavity and the pleura are presented in **Figure 19**. Incidence and location specific complications of hydatid disease is presented in **Table 3** [2].

3. Conclusion

Echinococcosis is a disorder of larval incursion by echinococcus tapeworm, prevalent in various continents in the world. Lungs and liver are the most commonly affected organs. Imaging has a significant role to implement the right treatment techniques. US is easily accessible and lacks radiation. MRI, CT and ERCP are also used for complicated cases.

Acknowledgements

No contribution by any other author or no funding declared.

Conflict of interest

The authors declare no conflict of interest.

DOI: http://dx.doi.org/10.5772/intechopen.88708

Author details

Tuba Apaydin
Department of Thoracic Surgery, SBU Istanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Training and Research Hospital, Istanbul, Turkey

*Address all correspondence to: tubaapaydn72@gmail.com

References

[1] Devi MA, Venumadhav T, Sukanya B. Role of imaging in diagnosis, predicting biological activity and in treatment plan of hydatid disease. Open Journal of Internal Medicine. 2018;**8**(3):177-195. DOI: 10.4236/ojim.2018.83018

[2] Srinivas MR, Deepashri B, Lakshmeesha MT. Imaging spectrum of hydatid disease: Usual and unusual locations. Polish Journal of Radiology. 2016;**81**:190-205. DOI: 10.12659/PJR.895649

[3] Sarkar M, Pathania R, Jhobta A. Cystic pulmonary hydatidosis. Lung India. 2016;**33**(2):179-191. DOI: 10.4103/0970-2113.177449

[4] Mehta P, Prakash M, Khandelwal N. Radiological manifestations of hydatid disease and its complications. Tropical Parasitology. 2016;**6**(2):103-112. DOI: 10.4103/2229-5070.190812

[5] Banderker E, Cox S, Gaxa L. Imaging and management of abdominal echinococcosis in children. Journal of Pediatric Infectious Diseases. 2017;**12**(01):030-039. DOI: 10.1055/s-0037-1599118

[6] Garg MK, Sharma M, Gulati A. Imaging in pulmonary hydatid cysts. World Journal of Radiology. 2016;**8**(6):581-587. DOI: 10.4329/wjr.v8.i6.581

[7] Carita P, Verdecchia M, Ferro G. Multimodality imaging in cardiac echinococcosis for diagnosis and follow-up of an untreatable cyst. Internal Journal of Cardiology. 2016;**221**:468-470. DOI: 10.1016/j.ijcard.2016.06.317

[8] Tandur R, Irodi A, Chacko BR. Magnetic resonance imaging as an adjunct to computed tomography in the diagnosis of pulmonary hydatid cysts. Indian Journal of Radiology and Imaging. 2018;**28**(3):342-349. DOI: 10.4103/ijri.IJRI_121_18