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A 24-Year-Old Man of Turkish Origin With Jaundice and Cystic Liver Lesions

Vàng da

MARIJA STOJKOVIC

Clinical Presentation

History

A 24-year-old man of Turkish origin presents at a hospital in Germany because of right upper quadrant pain, nausea and vomiting.

The patient is a German resident but visits his family in rural eastern Turkey (Anatolia) every year for about 6 weeks.

Three years earlier the patient had presented with right upper quadrant pain. Then, an ultrasound and a CT scan of the liver had revealed two calcified cystic lesions, one in the right liver lobe, another smaller cyst in the left liver lobe (Figs. 50.1A and B and 50.2). Serology was positive for *Echinococcus granulosus* and a diagnosis of cystic echinococcosis (CE) was made.

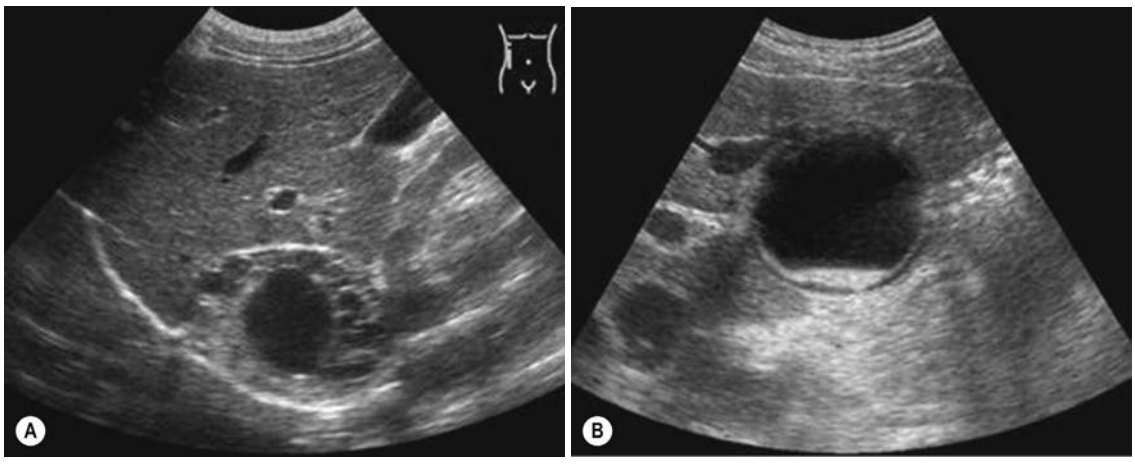
Since both cyst walls were already calcified, anthelmintic treatment was not considered an option as the bioavailability of albendazole in cysts with calcified cyst walls is rather poor. Being free of symptoms, the patient refused surgical treatment. The cysts were monitored regularly by ultrasound for signs of spontaneous involution.

Clinical Findings

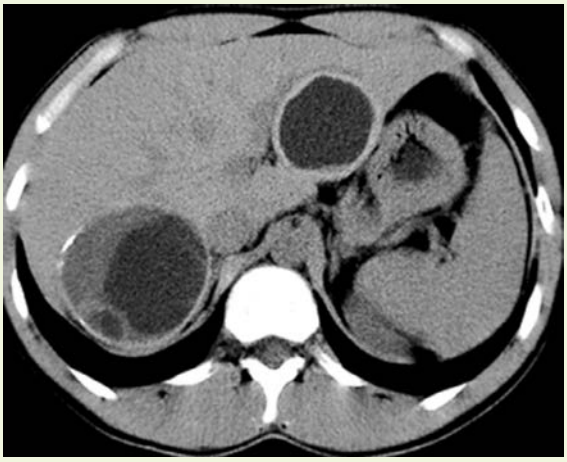
A 24-year-old man in fair general condition with scleral jaundice. Blood pressure 110/70 mmHg, pulse 64 bpm, temperature 36°C (96.8°F). There is right upper quadrant and epigastric tenderness, but no guarding.

Laboratory Findings

The full blood count is normal. Additional results are shown in Table 50.1.



• **Fig. 50.1** Ultrasound scan of the liver at diagnosis 3 years before current presentation reveals two cystic lesions. One lesion is located in the right liver lobe (7 × 6 cm). Cyst content shows a solid cyst matrix containing multiple smaller cysts (A). The cyst wall is partially calcified (WHO classification CE3b). (B) Second cystic lesion in the left liver lobe (5 × 4 cm). The cyst wall is up to 4 mm thick with partial calcification. Cyst content is liquid; there is a double line sign, a feature of WHO CE1, and hydatid sand. (Copyright W. Hosch, Department of Radiology, Heidelberg University Hospital.)



• **Fig. 50.2** Computed tomography of the same patient. Both cyst walls show calcification. (Copyright W. Hosch, Department of Radiology, Heidelberg University Hospital.)

TABLE 50.1 Laboratory Results on Presentation

Parameter	Patient	Reference
AST (U/L)	145	<35
ALT (U/L)	454	<35
GGT (U/L)	394	<55
ALP (U/L)	402	38–126
Total bilirubin (μmol/L)	120	<19.0
ESR (mm/h)	17/40	≤10/20
CRP (mg/L)	49	<5

Questions

1. What is the suspected diagnosis and how would you approach this patient?
2. Which differential diagnoses should be considered in a patient with cystic liver lesions?

Discussion

A young man of Turkish origin presents with right upper quadrant pain and jaundice. He was diagnosed with CE 3 years earlier and has not been on any specific treatment.

Answer to Question 1

What is the Suspected Diagnosis and How Would You Approach This Patient?

His presentation should raise suspicion of a cystobiliary fistula with biliary obstruction, which is the most common acute complication in cystic echinococcosis and may be complicated by bacterial cholangitis.

In case of biliary obstruction, the first step is to restore the biliary flow by endoscopic retrograde cholangiography (ERC). ERC is both a diagnostic and therapeutic tool in cases of biliary obstruction; cyst content can be extracted from the hepatic or common bile duct. In case of bacterial cholangitis, the patient should receive antibiotic treatment to cover for Gram-negative bacteria and anaerobes.

Once biliary flow is restored and cholangitis has settled, the cyst can be surgically removed in a second step.

Answer to Question 2

Which Differential Diagnoses Should be Considered in a Patient With Cystic Liver Lesions?

The main differential diagnoses in this patient with cystic liver lesions and liquid cyst content are a congenital simple hepatic cyst and pseudocysts (necrotic cavity) in alveolar echinococcosis (*E. multilocularis*).

Depending on the presence of systemic symptoms and signs and liquid versus solid cyst content, infectious causes like abscesses (bacterial, amoebic) or tuberculoma must be considered.

In addition, benign and malignant liver tumours or metastases are relevant differentials.

The Case Continued…

On ERC, biliary obstruction because of a cystobiliary fistula was found. CE cyst content was removed from the common bile duct and the bile flow was restored. The patient's cholangitis was treated with ciprofloxacin and metronidazole. Once the inflammation settled, the patient was referred for surgery and partial cystectomy was performed on both cysts.

The patient has been followed up for 9 years after surgery and has had no recurrence. He is considered cured and further follow-up visits are not needed.

This case illustrates several learning points. In countries with low CE-endemicity provenance from a hyperendemic region is the single most important risk factor for CE in cystic liver lesions.

CE is generally a benign disease with the exception of complicated cysts; in this case a cyst with biliary fistula. CE cysts are currently classified by the WHO into six stages: CE1 and 2 (active), CE3a and 3b (transitional) and CE4 and 5 (inactive).

A very important question is which cysts can be left untreated and only observed ('watch and wait'). There is fairly solid evidence that inactive CE4 and CE5 cysts can be left untreated if they are not in critical sites.

In this patient the cyst walls of both cysts (CE1, CE3b) were calcified (Figs. 50.1 and 50.2). Previously, calcification was described only as a feature of CE5 cysts; this was later extended to CE4 cysts and has now been shown to occur in all cyst stages.

Retrospectively this case illustrates that surgical treatment was rightly offered to the patient at initial diagnosis.

Cystobiliary fistulas are some of the major reasons for complications in CE of the liver (as are cystobronchial fistulas in pulmonary CE). Fistulating cysts should be surgically treated before complications arise. The key question is how to identify fistulas early to prevent complications. Previously, endoscopic retrograde cholangiography (ERC) has been advocated as the method of choice. However, it has become evident that intracystic pressure may be too high for the contrast medium to enter. If available, MRI with magnetic resonance cholangiography (MRC) is an alternative option, with very good detection rates.

SUMMARY BOX

Cystic Echinococcosis

CE is an infection with the larval form of the dog tapeworm *Echinococcus granulosus*. It occurs worldwide with high endemicity in the Mediterranean basin, the Near and Middle East, North and East Africa, central Asia and Latin America.

Humans become infected by ingestion of eggs of *E. granulosus*.

Symptoms may occur months to years after infection because of mass effects of the growing cyst or because of complications. Liver (70%) and lung (15–30%) are the most commonly affected organs. Cyst complications include fistulas leading to biliary or bronchial obstruction, bacterial superinfection, cyst rupture leading to anaphylaxis, embolism of cyst content and compression syndromes.

Diagnosis of CE is based on imaging, mainly on ultrasound, which is crucial for the diagnosis and classification of disease activity. MRI or CT should be used when cysts are inaccessible by ultrasound.

Serology is hampered by a lack of sensitivity and specificity. In unclear cases, diagnostic cyst puncture and aspiration may be performed by experienced examiners. If CE is suspected, however, albendazole needs to be given peri-interventionally.

Generally, four treatment modalities are available: anthelmintic treatment, percutaneous sterilization techniques, surgery and a 'watch and wait' approach.

Management of CE patients in general depends not only on the individual case but also on local resources and expertise.

Long-term follow-up is important to detect recurrence of disease.

Further Reading

1. Stojkovic M, Gottstein B, Junghanss T. Echinococcosis. In: Farrar J, Hotez P, Junghanss T, Kang G, Lalloo D, White N, editors. Manson's Tropical Diseases. 23rd ed. London: Elsevier; 2013 [chapter 56].
2. Hosch W, Stojkovic M, Jänisch T, et al. The role of calcification for staging cystic echinococcosis (CE). Eur Radiol 2007;17:2538–45.
3. Stojkovic M, Zwahlen M, Teggi A, et al. Treatment response of cystic echinococcosis to benzimidazoles: a systematic review. PLoS Negl Trop Dis 2009;3(9):e524.
4. WHO Informal Working Group. International classification of ultrasound images in cystic echinococcosis for application in clinical and field epidemiological settings. Acta Trop 2003;85(2):253–61.
5. Stojkovic M, Rosenberger KD, Steudle F, et al. Watch and wait management of inactive cystic echinococcosis – does the path to inactivity matter – aalysis of a prospective patient cohort. PLoS Negl Trop Dis 2016;10(12):e0005243.