

Echinococcal Cyst

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July 28th 2011

Case Presentation

- CC: right upper quadrant pain
- HPI: This is a 33 y/o Lebanese male who was diagnosed with an echinococcal cyst in 2009 and was being treated with albendazole. He presented to the ED with a c/o recurrent RUQ / flank pain and was referred to surgery clinic for further evaluation.

- PMHx: echinococcal cyst
- PSHx: none
- Allergies: NKDA
- Meds: albendazole
- SHx: denies smoking, ETOH, IVDA
- FHx: non-contributory

- Radiologic Studies:

CT Abd/Pelvis: 10x10cm right hepatic hydatid cyst without evidence of rupture, 3.5cm cyst within larger cyst, cholelithiasis

Contrast: CONTRAST

Gantry: 0°

FoV: 433 mm

Time: ms

Slice: 3 mm

Pos: -47.1

FFS

F: B

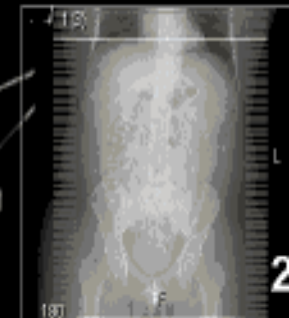
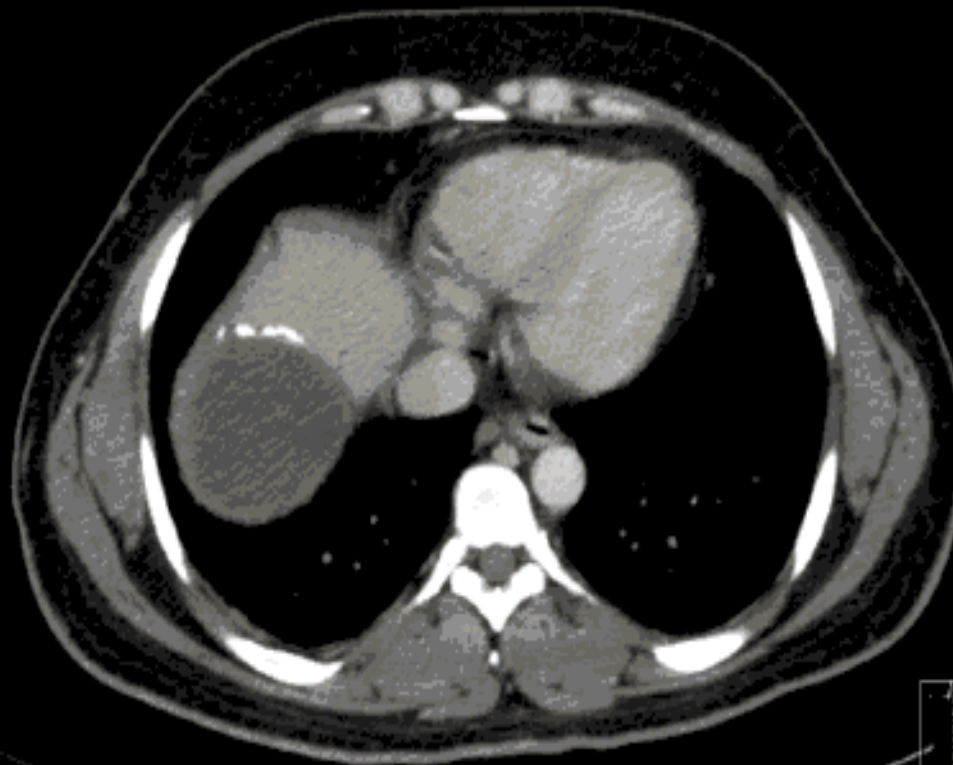
mA: 412

120 kV

Image no: 17

Image 17 of 180

5/25/2011, 6:57:13 PM



Contrast: CONTRAST

Gantry: 0°

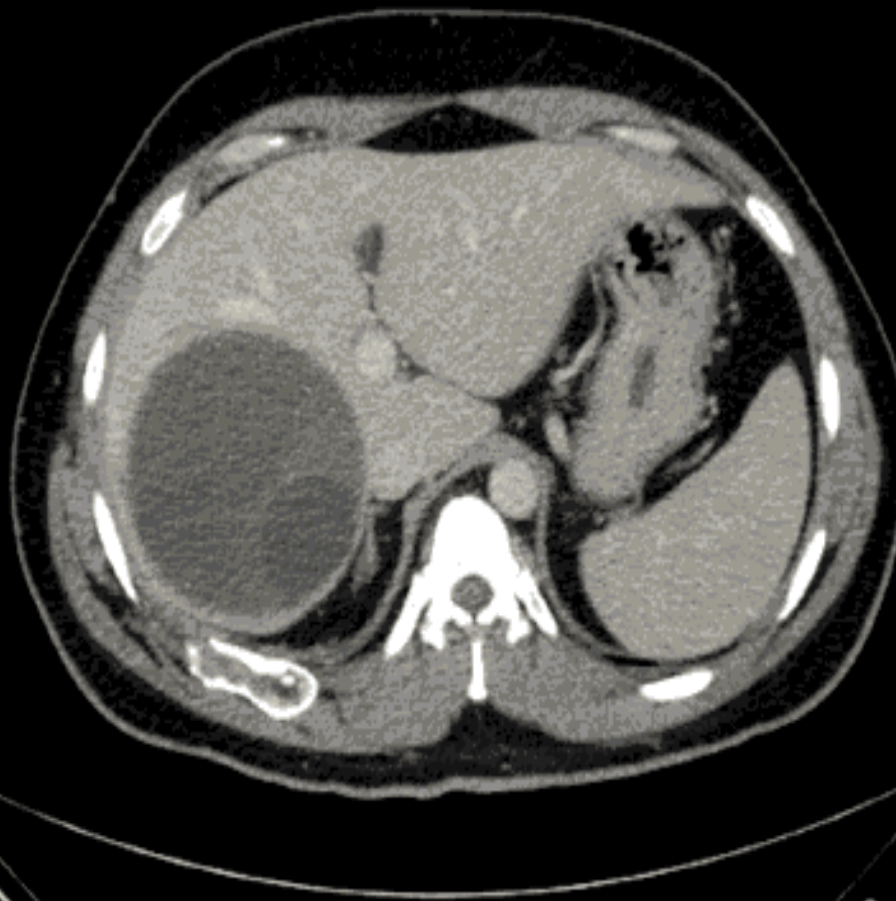
FoV: 433 mm

Time: ms

Slice: 3 mm

Pos: 15.9

FFS



F: B

mA: 412

120 kV

Image no: 38

Image 38 of 180

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- Vitals: Temp 98.8 F BP 115/84 HR 68 RR 18
- Physical Exam:
 - General: AAOx3
 - HEENT: NCAT, EOMI
 - Chest: CTA bilaterally
 - CVS: S1S2, rrr
 - Abdomen: soft, +BS, ND, RUQ/flank fullness
 - Extr: warm to touch, no edema

- Labs:

CBC: 7.3 / 16.3 / 46.8 / 255

Chem: 139 / 4.0 / 102 / 26 / 12 / 0.87 / 84 Ca 10

LFTs: 7.5 / 4.5 / 19 / 28 / 70 / 0.5

Coags: 10.3 / 29 / 1.0

- Intra-op:
 - subcostal incision
 - cholecystectomy
 - mobilization of right lobe of liver
 - aspiration of cyst content
 - infusion of 10% betadine
 - drainage of cyst
 - pericystectomy
 - repair of IVC injury
 - placement of JP drains x 2
 - omentum placed in cyst cavity

- Intra-op:



- Post-op course:

POD #1 – extubated

POD #2 – NGT d/ced, JP drainage bile tinged

POD #4 – clear liquid diet; albendazole restarted

POD #7 – tolerating regular diet, JP output

minimal and serosanguinous, discharged
home

- Pathology:

echinococcal cyst fibrous wall and
laminated membranes present, no scolices
present

- Echinococcus

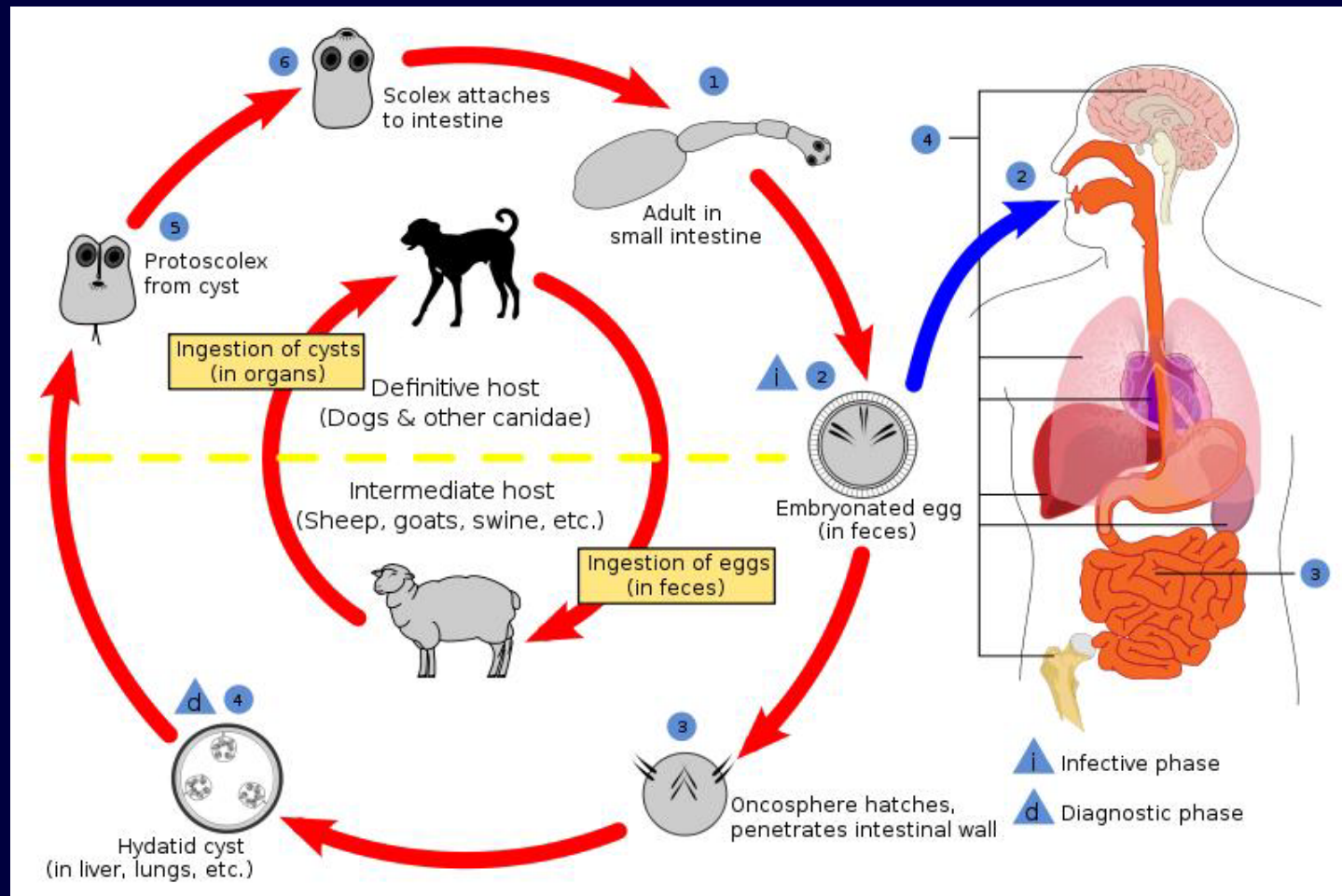
- flat tapeworm; scolex containing 4 suckers and 2 rows of hooklets
- endemic in Mediterranean countries, the Middle and Far East and South America
- annual incidence ranges from 13 to 220 per 100,000 persons

- Echinococcus
 - no racial predilection
 - females more affected than males
 - children have a higher rate of infection
 - life cycle alternates between carnivores and herbivores
 - intermediate host: sheep, cattle, pigs
 - definitive host: dog

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Echinococcal Cyst

- Life Cycle



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Echinococcal Cyst

- species most commonly associated with hydatid disease in humans: *E. granulosus*
- site of infection within the body
 - liver (55-80%)
 - lung (10-40%)
- Characterized by a single cyst, with or without daughter cysts

- Pathology
 - external acellular layer (ectocyst)
 - composed of fibroblasts, forms the pericyst, calcified in 50% of patients
 - inner cellular germinal layer (endocyst)
 - brood capsules that contain protoscoleces; daughter cysts may develop

- Symptoms

- abdominal pain, palpable mass, fever, fatigue, nausea, jaundice

- Diagnosis

- clinical suspicion, epidemiologic data
 - labs: eosinophilia (20-25%); serology (ELISA) and hydatid antigen immunobinding assay (95% sensitive; 90% specific)
 - imaging (U/S, CT, MRI)

- Ultrasound

- first line diagnostic imaging tool
- inexpensive, noninvasive, readily available
- cysts appear as thick-walled with calcifications
- specificity to rule-out hydatid disease: 90%
- ultrasonographic classification of hydatid disease
 - Gharbi's classification
 - WHO standardized classification
- helpful as staging determines therapy

- Gharbi's classification of cystic hydatid disease**

Type	Ultrasound classification
I	Pure fluid collection
II	Fluid collection with a split wall /detached membrane (water lily sign)
III	Fluid collection with septa (honeycomb sign) or daughter cyst
IV	Heterogeneous echographic patterns
V	Reflecting thick +/- calcified wall

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Echinococcal Cyst

- WHO Working Group on Echinococcus classification

Type of Cyst	Status	Ultrasound Features	Remarks
CL	Active	Signs not pathognomonic, unilocular, no cyst wall	Usually early stage, not fertile
CE 1	Active	Cyst wall, hydatid sand	Usually fertile
CE 2	Active	Multivesicular, cyst wall, rosette-like	Usually fertile
CE 3	Transitional	Detached laminated membrane, “water lily” sign, less round, decreased intracystic pressure	Starting to degenerate, may produce daughter cyst
CE 4	Inactive	Heterogenous hypoechogenic or hyperechogenic degenerative contents; no daughter cyst	Usually no living protoscolices
CE 5	Inactive	Thick, calcified wall, calcification partial to complete; not pathognomonic, but highly suggestive of diagnosis	Usually no living protoscolices

- CT/MRI

- provides structural details; location and depth within liver
- presence of daughter or exogenous cysts

- ERCP

- indicated for patients with jaundice or cholangitis
- determine if there is a communication between biliary tract and echinococcal cyst

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Echinococcal Cyst

- Treatment: once disease is confirmed the principle of treatment centers on elimination of the Echinococcus and prevention of recurrence
- Medical Management
 - chemotherapy with benzimidazole compounds: mebendazole, albendazole
 - parasitostatic activity as patient may develop recurrent disease with cessation of drug
 - medical management alone may result in 30% clinical and radiographic resolution of disease

- Medical Management

- recommended that drug therapy should primarily be used in conjunction with percutaneous drainage or surgery
- WHO guidelines: preoperative administration
 - albendazole 1-4 days
 - mebendazole 3 months
- mebendazole: poorly absorbed in the intestine, does not penetrate cyst well
- albendazole: better intestinal absorption, rapid first pass metabolism, higher cyst fluid concentration, drug of choice

- Percutaneous Aspiration, Injection, and Reaspiration (PAIR)
 - transhepatic percutaneous drainage under CT or U/S guidance
 - injection of protoscolicidal agent
 - most commonly used is 20% hypertonic saline
 - reaspiration of cyst content
 - benzimidazole therapy 1 week before and 28 days after procedure

- Percutaneous Aspiration, Injection, and Reaspiration (PAIR)
 - following procedure cyst fluid can be assessed for viable protoscolices
 - complications: infection, leakage (fever, anaphylactic shock)
 - indicated in pts who refuse surgery, multiple cyst or relapse after surgery
 - contraindicated: inaccessible to puncture
 - high rate of recurrence and fistula formation

Echinococcal Cysts: Surgical Agents

Use of albendazole sulfoxide, albendazole sulfone, and combined solutions as scolical agents on hydatid cysts (*in vitro* study)
Gokhan Adas et al.; Okmeydani Training Hospital, Okmeydani/Sisli, Istanbul 34715, Turkey

World J Gastroenterol. 2009 January 7; 15(1): 112–116.

Table 2: Different scolical agents in literature

Author	Agents	Results
Caglar et al[26] (2008)	20% silver nitra (20 min)	100% death
	50% Dextroz (30 min)	100% death
	20% NaCl (45 min)	100% death
	20% Mannitol (45 min)	100% death
Frayha et al[27] (1981)		
	Cetrimide 0.5%-1% (10 min)	100% death
Kayaalp et al[28] (2002)		
	10%-30% NaCl (3, 6, 75 min)	100% death
	Sonişik et al[11] (2002)	
Besim et al[16] (1998)	10%-30% NaCl (3, 6 and 75 min)	100% death
	20% Saline (15 min)	100% death
	95% Ethyl alcohol (15 min)	100% death
	10% Polyvinyl pirrolidone iodine (15 min)	100% death
	3% H2O2 (15 min)	100% death
Erzurumlu et al[20] (1998)		
	Albendazole sulfoxide 20 µg/mL	5% death
	Albendazole sulfoxide 50 µg/mL	50% death
	Albendazole sulfoxide 100 µg/mL	100% death
Adas et al (our study 2008)		
	20% NaCL (5 min)	98.2% death
	3% H2O2 (5 min)	90.3% death
	Cetrimide (5 min)	86.9% death
	Albendazole sulfone (5 min)	97.3% death

- Surgical Management

- options: cyst drainage and partial pericystectomy with or without omentoplasty, total pericystectomy, partial hepatectomy or hemihepatectomy

- surgical approach is based on cyst size, site, type (classification system) and surgeon's expertise

- Surgical Management: WHO Classification Guidelines
 - CL, CE1, CE3: chemotherapy (CT); however PAIR is required due to lack of degenerative changes with CT alone; surgery if CT and PAIR fail
 - CE2: PAIR or surgery based on possibility of cyst drainage, number of daughter cysts and location
 - CE4, CE5: surgery is treatment of choice unless completely calcified and inactive

- Surgical Management: simple cyst drainage
 - pack operative field with scolicedal soaked lap sponges
 - aspirate cyst content with a closed system
 - infuse scolicedal agent
 - evacuate cyst content
 - unroof cyst and remove cyst wall
 - explore cavity; may be filled with omentum

- **Surgical Management: total pericystectomy**
 - open technique: removal of the pericyst wall with electrocautery after drainage
 - recommended when cyst is large and may rupture
 - closed technique: plane is identified outside the pericyst and en bloc resection is performed without manipulation of cyst content
 - technically demanding as hepatic parenchymal transection is required

- Surgical Management: liver resection
 - indicated if previous surgical therapies have failed
 - should only be performed if complete cyst excision can be achieved
 - contraindicated in centrally located or deep seated cysts

Complex hydatid cysts of the liver: a single center's evolving approach to surgical treatment.

Botrugno I, Gruttadauria S, Crino F, Anastasi D, Gridelli B ; *Am Surg* 2010 Sep; 76(9):1011-5.

- retrospective study; 38 patients; complex hydatid cyst $\geq 10\text{cm}$, multiple or recurrent, resistant to conservative management
- aim: analyze patients who underwent surgery for complex hydatid cysts to determine the evolution of the surgical treatment and the risk factors for recurrence
- 14 women; 24 men
- mean follow-up: 24 months

Complex hydatid cysts of the liver: a single center's evolving approach to surgical treatment.

Botrugno I, Gruttadauria S, Crino F, Anastasi D, Gridelli B ; *Am Surg* 2010 Sep; 76(9):1011-5.

- all pts given albendazole (10mg/kg/day) 15d preop; 2 mos postop
- partial pericystectomy – 2 pts (5.3%); total pericystectomy – 20 pts (53%); liver resection – 15 pts (39%); wedge resection and pericystectomy – 1 pt (2.6%)
- no deaths; 1 patient had signs of recurrence during follow-up
- conclusion: surgery most effective treatment for complex hydatid cyst; pericystectomy (partial or total) may have eliminated need for liver resection

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Echinococcal Cyst

- Postoperatively WHO recommends continuation of albendazole for 1 month or mebendazole for 3 months
- Uncomplicated disease
 - morbidity 20%; mortality 1%
- Complicated disease
 - morbidity 30-50%; mortality 5%
- Recurrence rate approximately 10%
- Follow-up: long-term; serology and radiographic imaging due to risk of reinfection

References

1. Cameron et al. Current Surgical Therapy 9th Ed. pgs. 331-335
2. Townsend, Beauchamp et al. Sabiston Textbook of Surgery 17th Ed.
3. Besim H et al. Scolicidal agents in hydatid cyst surgery. HPB Surg 1998;10(6):347-51.
4. Gokhan Adas et al; Use of albendazole sulfoxide, albendazole sulfone, and combined solutions as scolicidal agents on hydatid cysts (*in vitro* study) World J Gastroenterology 2009 Jan 7; 15(1): 112-116
5. Brunetti et al; Echinococcosis Hydatid Cyst Treatment and Management <http://emedicine.medscape.com>