When Life Gives You Larvae: *Echinococcus Multilocularis* Infection

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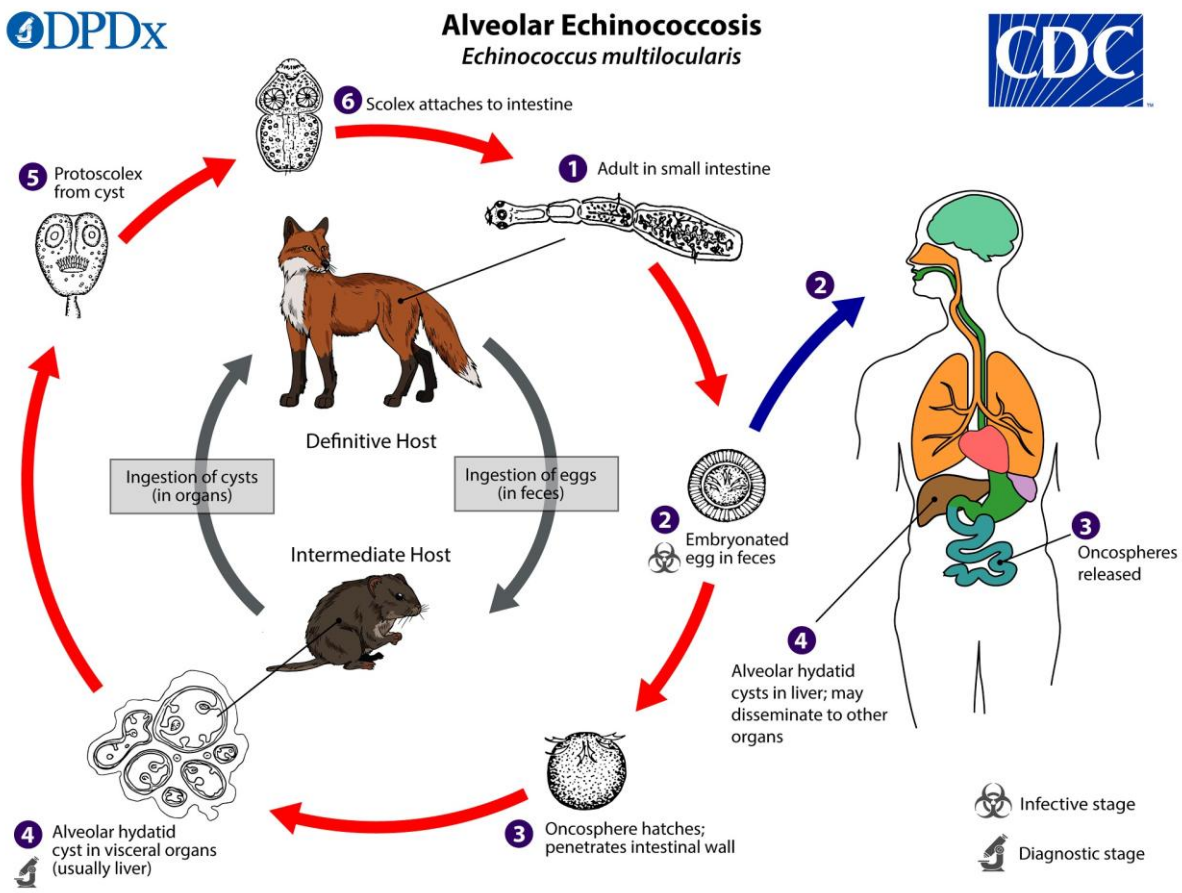
BACKGROUND

- Human Echinococcus or hydatid disease is an infection caused by tapeworms within the genus Echinococcus.
- There are two major groups of Echinococcus with distinct clinical presentations:
  - ***E. granulosis*** – cystic hydatid disease
    - Slow growing cysts in the lungs and liver
  - ***E. multilocularis*** – alveolar hydatid disease
    - Much less common (0.03 to 1.2 per 100,000 in endemic areas)

Goodfellow, M., Shaw, S. & Morgan, E. Imported disease of dogs and cats exotic to Ireland: *Echinococcus multilocularis*. *Ir Vet J* **59**, 214 (2006).

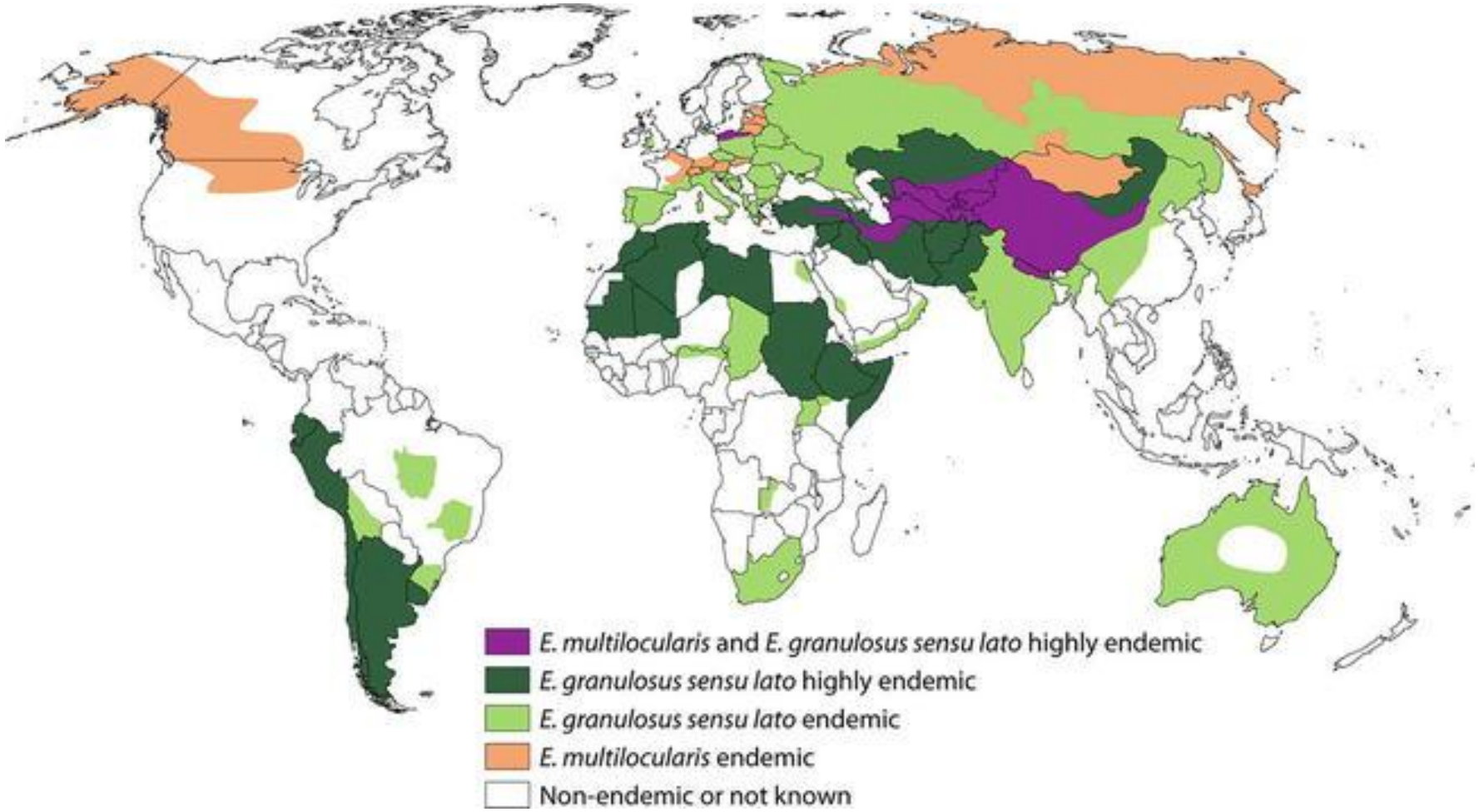
BACKGROUND

- Human *E. multilocularis* infection occurs through:
  - Direct contact with definite hosts:
    - Foxes (most common)
    - Wolves
    - Coyotes
    - Cats and dogs (least common)
  - Indirectly through contaminated water or vegetation.
- Eggs are ingested and hatch in the intestine, releasing oncospheres that pass through the portal and lymphatic vessels to reach the liver where they develop as larvae (metacestodes or hydatid cysts).



https://www.cdc.gov/dpdx/echinococcosis/index.html

BACKGROUND



- The *Echinococcus multilocularis* tapeworm is endemic to the midwestern United States, Alaska, Canada, Japan, central Europe, Asia, and Russia.

Wen H, Vuitton L, Tuxun T, Li JVuitton DA, Zhang W, McManus DP.2019.Echinococcosis: Advances in the 21st Century. Clin Microbiol Rev 32:10.1128/cmr.00075-18

CLINICAL PRESENTATION

- Typically asymptomatic for years with presentation of symptoms occurring between 5-15 years after infection depending on the location of the cyst.
- More than 90% of patients have hepatic infection, but infection can occur in any organ or tissue.
- Patients present with non-specific symptoms including malaise, weight loss, and right upper quadrant pain.
- Mortality is >90% within 10-15 years of diagnosis without treatment.

Reported symptoms among patients with alveolar echinococcus at diagnosis

Symptoms	Number of reports	%
Abdominal pain	176	56.4
Abscess	11	3.5
Cholangitis	15	4.8
Cholestatic jaundice	85	27.2
Hepatomegaly	65	20.8
Portal hypertension	5	1.6
Pruritus	52	16.7
Other symptoms	68	21.8
One symptom	157	50.3
Multiple symptoms	155	49.7

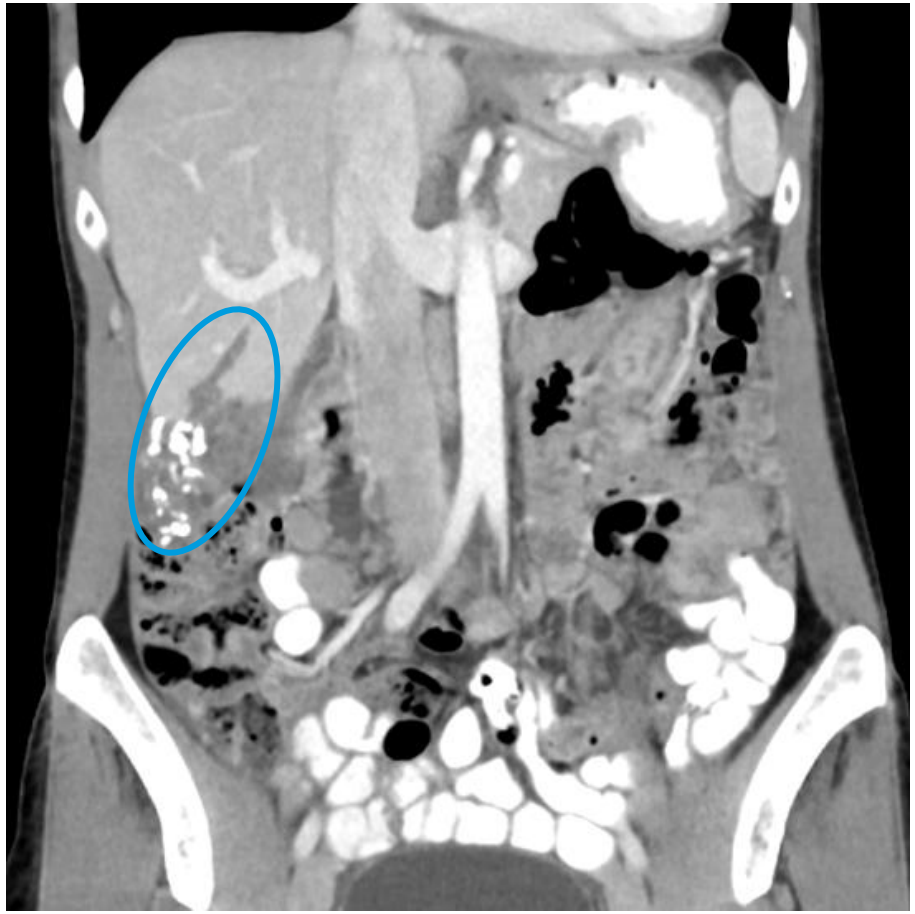
Knapp J, Demonmerot F, Gbaguidi-Haore H, et al. Epidemiological and clinical characteristics of patients in the alveolar echinococcosis registry, France, 1982 to 2021. *Euro Surveill*. 2025;30(32):2500041.

CASE PRESENTATION

- 42-year-old immunocompetent female with 3 months of worsening abdominal and pelvic pain.
  - Denied fever, chills, night sweats, or weight loss.
- She was born in Japan and moved to the United States at age 19.
- She had an extensive travel history throughout Asia, Serbia, and the north and southeastern United States.
- Abdominal imaging was concerning for a liver mass.

CT scout with calcifications within the inferior right hepatic lobe.

CT ABDOMEN/PELVIS WITH CONTRAST

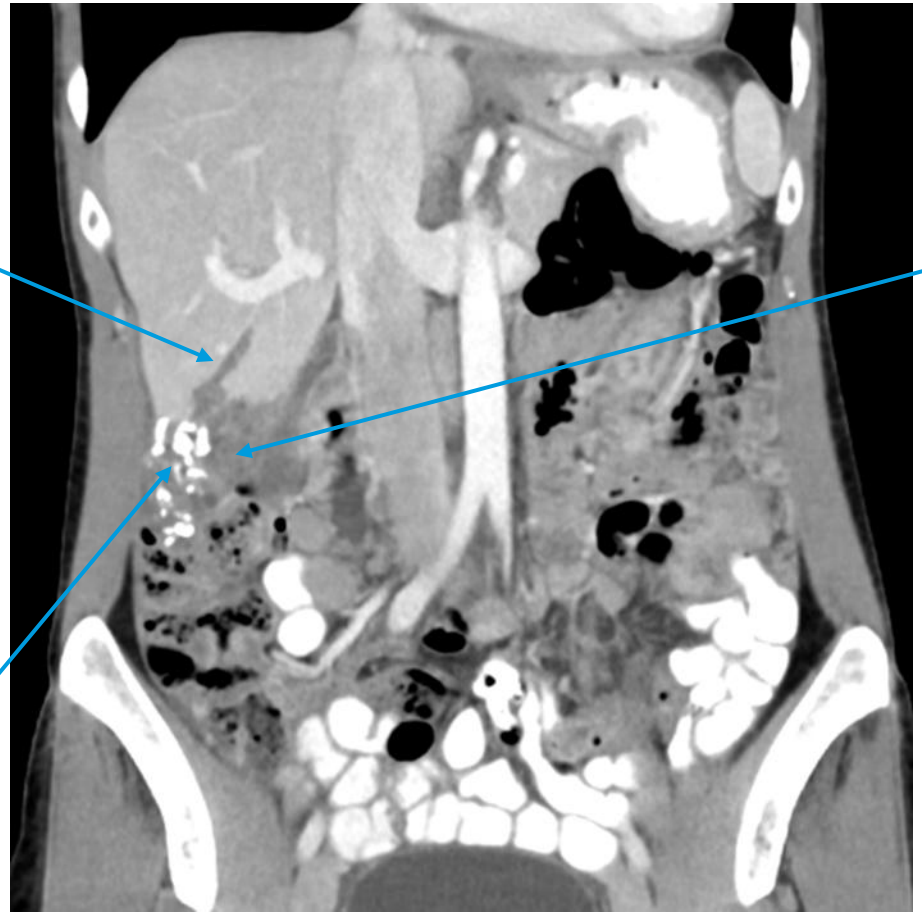


- Non-enhancing, partially calcified multi-cystic lesion at the inferior right hepatic lobe with intrahepatic biliary duct dilation.

TYPICAL CT IMAGING FINDINGS

An infiltrative mass with calcifications and the absence of contrast enhancement suggests the diagnosis.

Irregular, ill-defined lesions resembling neoplasm or metastasis.

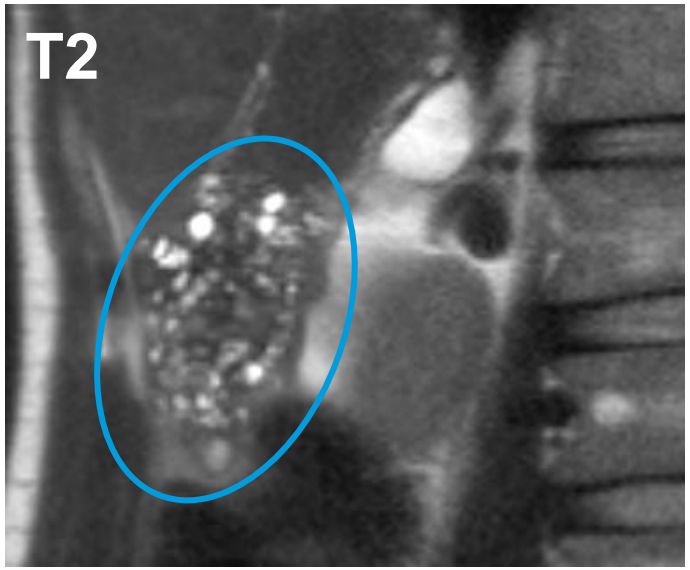


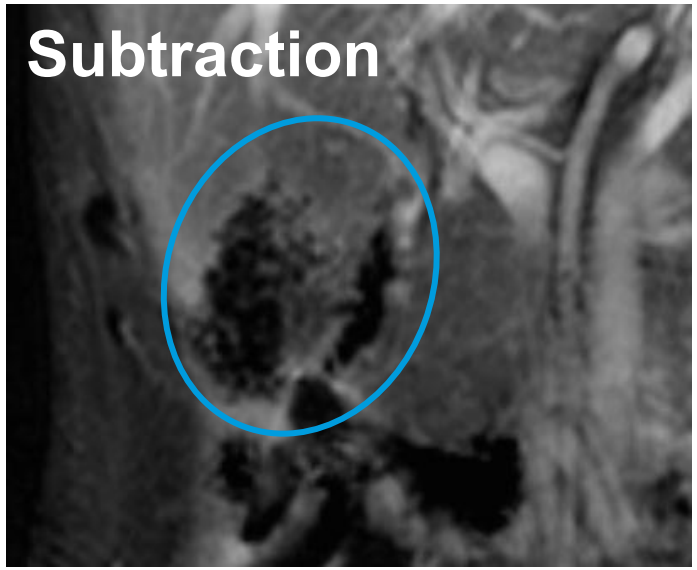
Central liquefactive necrosis with poor or no enhancement due to invaded vasculature.

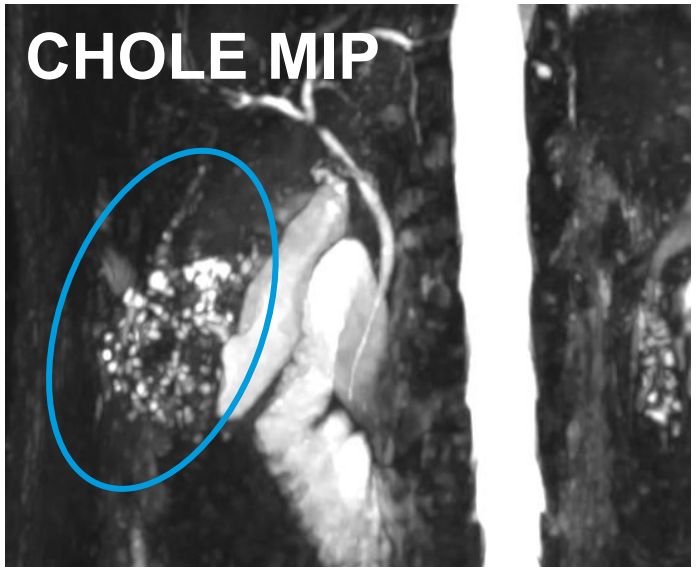
90% have central irregular microcalcifications, which can be better seen on CT than MR.

- 50% have hilar infiltration → intrahepatic bile duct dilation or portal or hepatic vein invasion → hepatic atrophy.
- Cyst rupture or direct invasion → spread to peritoneum, omentum, and other organs.

MR ABDOMEN



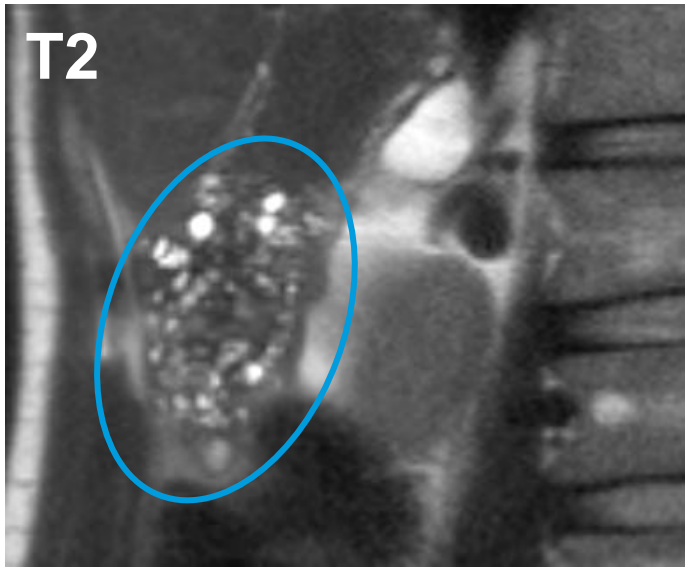


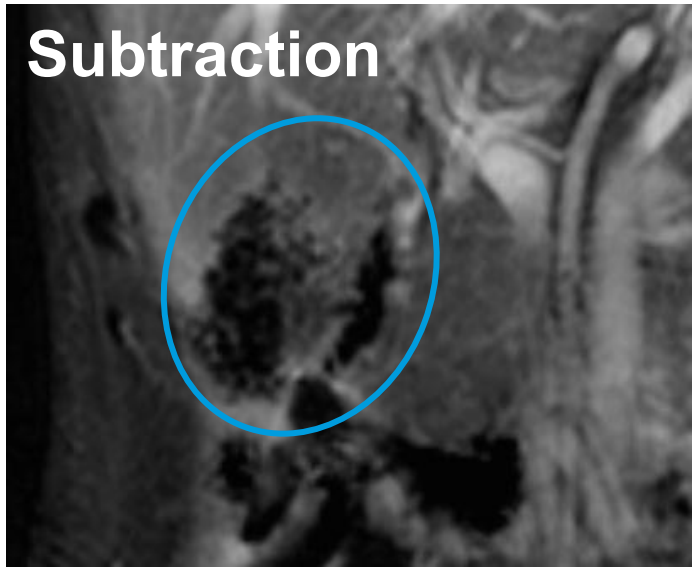


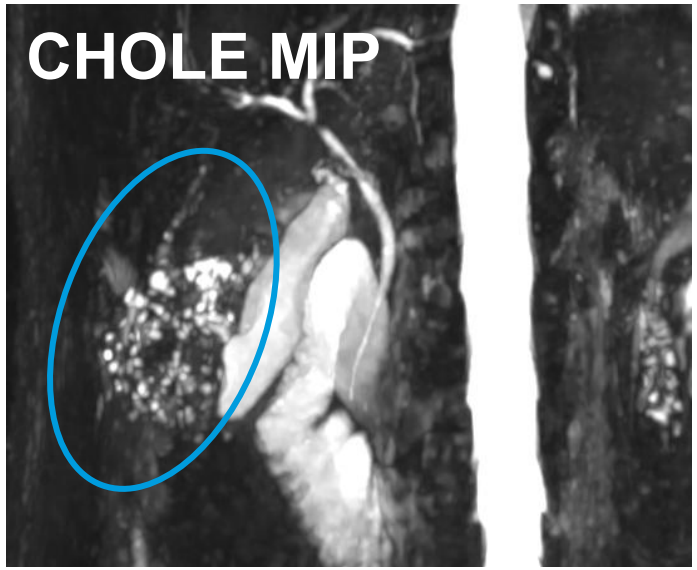
- T2 hyperintense, multiloculated, partially cystic lesions involving the inferior right hepatic lobe measuring 7.7 x 3.3 x 7.3 cm with multifocal calcifications and focal intrahepatic biliary duct dilation.
- MRCP – Cystic lesion with calcifications in segments V and VI, mild prominence of the right intrahepatic ducts, and patient hepatic vasculature.

TYPICAL MR FINDINGS

MRI provides more precise characterization. Typically, a heterogenous infiltrative hypovascular mass with mixed cystic and solid tissue.







T2-weighted MRI:

High signal - microcysts and liquefactive necrosis

Low signal - granulomatous tissue, coagulative necrosis, and calcification

MRI Post Contrast:

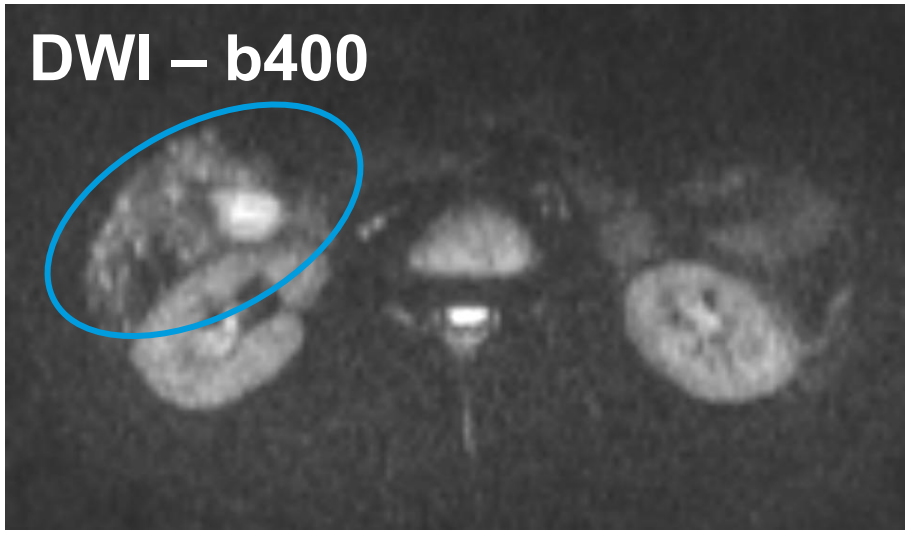
Central liquefactive necrosis with poor or no enhancement due to invaded vasculature.

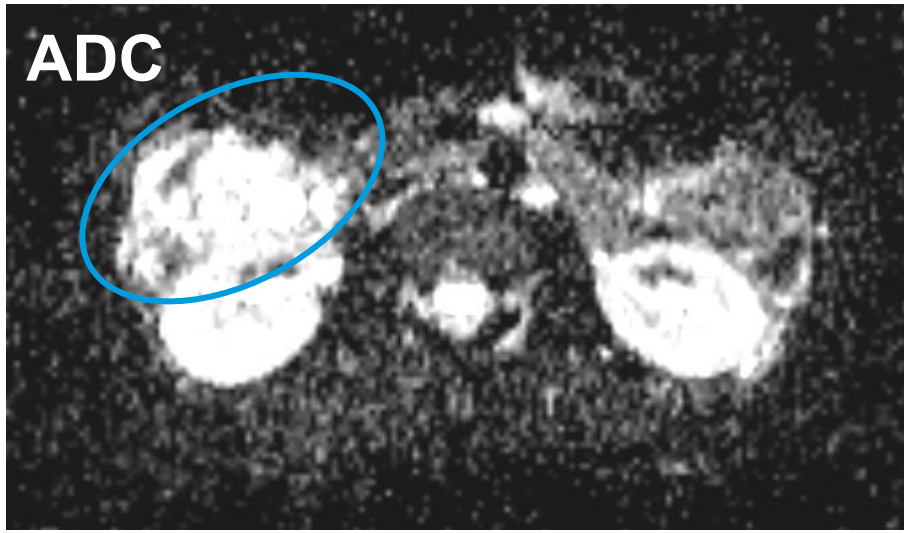
MRCP:

Useful to determine potential invasion of the biliary tree.

Lesions described as a “bunch of grapes” or “honeycomb”.

TYPICAL MR FINDINGS

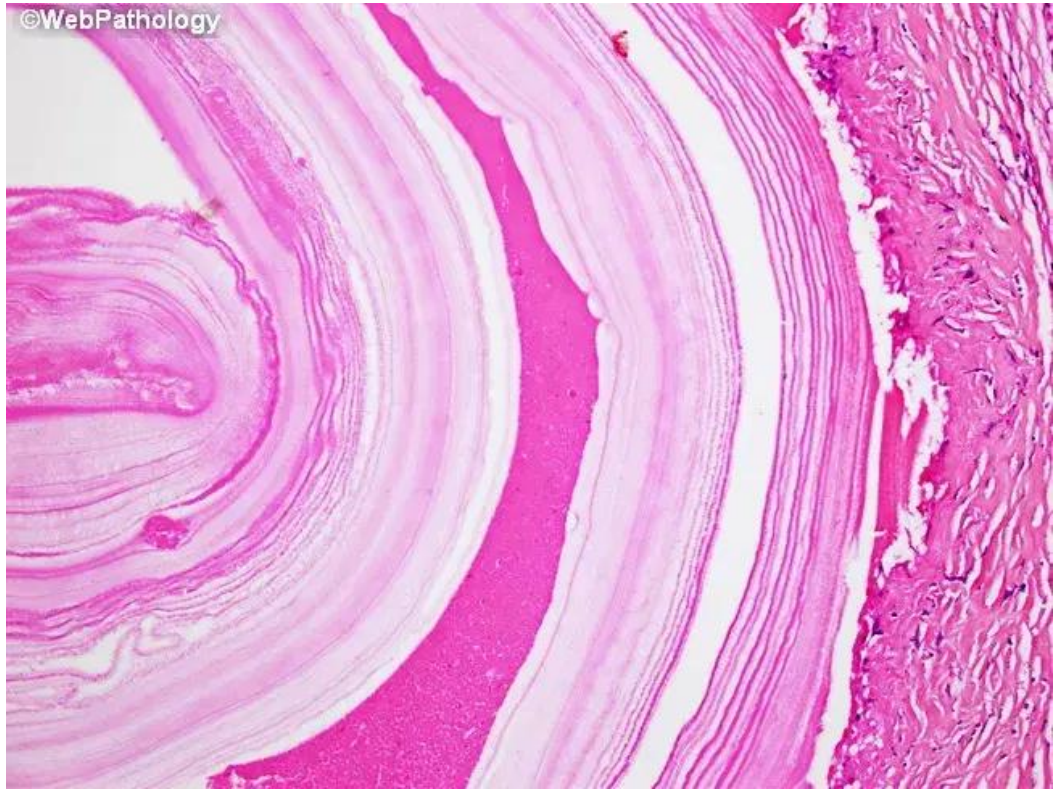




- Diffusion Weighted Imaging and Apparent Diffusion Coefficient map both show hyperintensity within the right lower hepatic lobe.
- This can be helpful in differentiating echinococcal infection from cholangiocarcinoma, which will be hypointense on the ADC map.

DIAGNOSIS

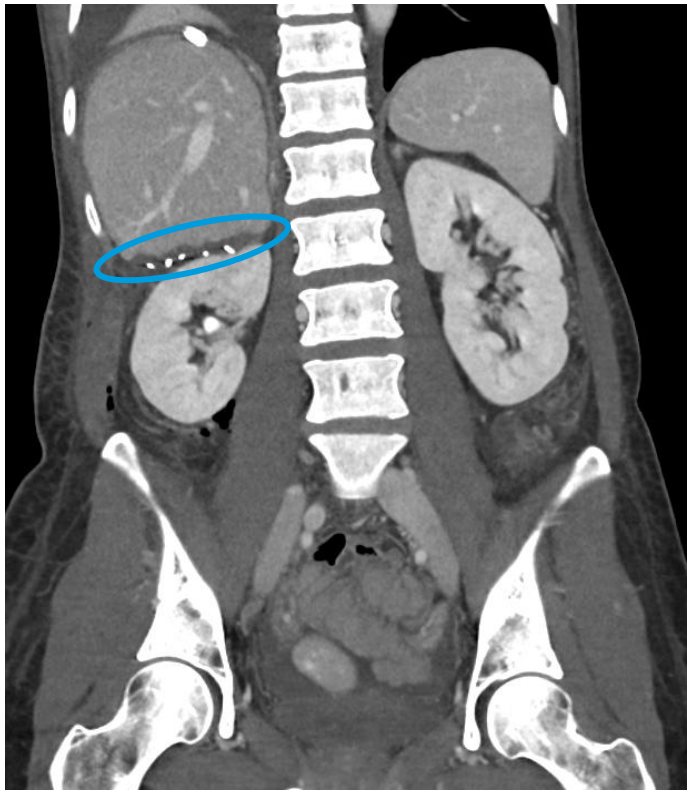
- Patient underwent exploratory laparotomy.
  - Laminated membranes with necrotizing granulomatous inflammation were present in samples from liver, peritoneal implants, and omentum.
- Echinococcus IgG level was elevated to 4.8.
- Metagenomic testing was positive for *Echinococcus multilocularis.*



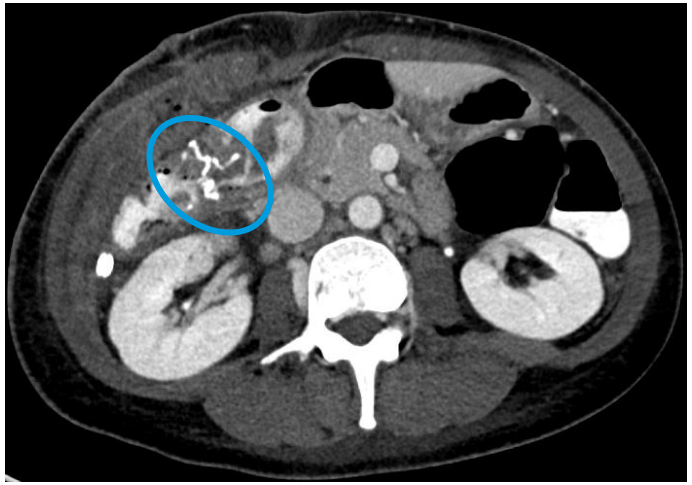
https://www.webpathology.com/images/infectious-disease/parasites/tissue-cestodes-tapeworms/46238

TREATMENT

- CT chest and MRI brain showed no dissemination of infection.
- She was treated with 400mg Albendazole twice daily for 2 months prior to surgery.
- Debulking surgery included a right hepatectomy of segments IVb, V, and VI, cholecystectomy, peritoneal stripping, and transverse colon resection due to infarction and devitalization.
- Albendazole was continued for 2 years following surgery without disease recurrence.



Right hepatectomy of segments IVb, V, and VI.



Transverse colon resection.

TAKE AWAY POINTS

- *Echinococcus multilocularis* causes an aggressive and invasive hepatic hydatid disease, which can take over a decade to present after direct exposure to a definitive host (fox, wolf, coyote, dog).
- Hepatic lesions can be mistaken for malignancy, but imaging features can help distinguish the infection including:
  - Central irregular microcalcifications best seen on CT.
  - Non-enhancing central liquefactive necrosis on both CT and MR contrast-enhanced imaging.
  - Characteristic microcysts on T2-weighted imaging.
- Treatment often includes a combination of albendazole and surgical resection.

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