Alveolar Echinococcosis Presenting as Hepatocellular Carcinoma: A Rare Case Report

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Introduction

- Alveolar Echinococcosis (AE) is a rare zoonotic disease caused by the helminthic parasite *Echinococcus multilocularis*.[1,2]
- *E.multilocularis* is endemic to the northern hemisphere affecting approximately 0.03-1.2/100,000 individuals annually.[2,3]
- Although more prevalent in Europe and Asia, the rates of AE have increased in North America in recent years.[4]
- AE is marked by visceral hydatid cyst formation, most commonly in the liver, with expansive, tumor-like growth and invasion.[1,2,5]
- Similar to malignant neoplasms, AE infections are able to spread to adjacent or distant sites through direct, lymphatic or hematogenous pathways.[1,3]
- Usually asymptomatic for a 5-15 year period and most commonly presents with abdominal pain, fatigue and weight loss.[3,5,6]
- Often misdiagnosed as primary liver malignancies due to their tumor-like growth pattern.[2,3,5]
- If inadequately treated or left untreated AE has a >90% fatality rate.[5,7]

Clinical History

- A patient in their 30s presented with right upper quadrant pain, worsened after eating, along with concurrent shoulder pain.
- Ultrasound, CT and MRI scans revealed an ill-defined heterogeneous mass (10.5x 10.5 x15 cm) in the right lobe of the liver consistent with possible hepatocellular carcinoma.
- A right hepatectomy was performed to remove the mass along with portal and retroperitoneal lymph nodes.

Results

Gross Examination

- Sectioning revealed a light tan, ill-defined, variegated mass (17.1 x 10.2 x 9.7 cm) associated with a light tan nodular area of liver capsule.
- Inconsistent with the typical presentation of hepatocellular carcinoma, the mass consisted of tan to yellow solid components and mucinous to milky cystic components.
    - Hepatocellular carcinoma usually presents as a nodular (unifocal or multifocal) or diffusely infiltrative mass often arising in the background of cirrhosis.[8]
- The mass grossly abuts the liver capsule, resection margin and hepatic tubular structures.
- The remaining liver parenchyma had a nutmeg appearance.
- Representative sections of the mass and potential lymph nodes (0.7 and 2.7 cm in maximum dimension) were submitted for histological examination.

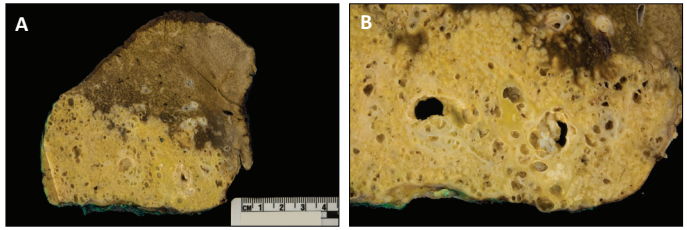

Fig. 1. Gross sections of the hepatic mass. **(A)** Solid and multicystic components on the cut surface of the mass with nutmeg appearance of uninvolved liver parenchyma. **(B)** Magnified view of solid and cystic hepatic mass.

Histology & Ancillary Studies

- Microscopic examination of the liver mass revealed the presence of laminated membranes and scolices consistent with echinococcal infections.
- A portion of fibroadipose tissue was also involved by echinococcosis.
- A final diagnosis of AE was determined after serology and microbiology were performed and confirmed an infection with *E.multilocularis.*

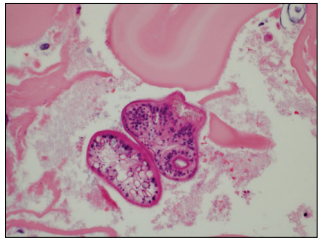

Fig. 2. Histological section of liver mass demonstrating *E.multilocularis.* (H&E, 40x magnification)

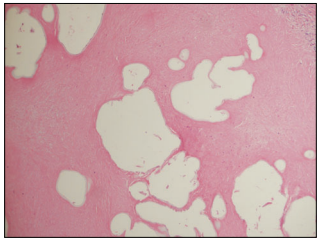

Fig. 3. Histological section demonstrating cystic spaces in liver. (H&E, 10x magnification)

Discussion

- Staging and classification of AE is based on the European Network for Concerted Surveillance of Alveolar Echinococcosis and the WHO Informal Group on Echinococcosis PNM system.[6,9]

Table 1: PNM classification of alveolar echinococcosis.[6,9]

P	**Hepatic localization of the parasite**
Px	Primary tumor cannot be assessed
P0	No detectable tumor in liver
P1	Peripheral lesion without proximal vascular and/or biliary involvement
P2	Central lesions with proximal vascular and/or biliary involvment of one lobe
P3	Central lesions with hilar or biliary involvement of both lobes and/or with involvement of two hepatic veins
P4	Any liver lesion with extension along the vessels (inferior vena cava, portal veins and arteries) and the biliary tree
N	**Extra-hepatic involvement of neighbouring organs and regional lymph nodes**
Nx	Not evaluable
N0	No regional involvement
N1	Involvement of neighbouring organs or tissues
M	**Absence or presence of distant metastasis**
Mx	Not completely assessed
M0	No metastasis on chest radiograph and computer tomography
M1	Metastasis present

Table 2: PNM stage grouping of alveolar echinococcosis.[6,9]

Stage	Criteria		
I	P1	N0	M0
	P2	N0	M0
IIIa	P3	N0	M0
IIIb	P1-3	N1	M0
	P4	N0	M0
IV	P4	N1	M0
	Any P	Any N	M1

- Accurate descriptions of mass location, invasion to adjacent structures and lymph node involvement are all important for determining AE stage which directly impacts patient treatment.
- Treatment for AE may include antiparasitic drugs therapy, surgical resections, liver transplantation or combinations of the three.[6]

Conclusion

- AE is a rare zoonotic disease associated with a poor prognosis if left untreated.
- The characteristic tumor-like growth and dissemination patterns makes proper initial diagnosis difficult in areas with low prevalence.
- Careful grossing on echinococcosis cases by pathologists' assistants is of utmost importance for accurate staging and subsequent treatment.

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