

نحوه برخورد با خانم 58 ساله مبتلا به کیست هیداتید

ارائه : دکتر امیرحسین زارع

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1403/8/26

History

- منبع شرح حال خود بیمار/ قابل اعتماد ، همکاری مناسب و مطلع
- شکایت اصلی : تمديد داروهای آسم
- شرح حال بیماری فعلی : بیمار خانم ۵۸ ساله مورد شناخته شده آسم (از ۱۰ سال پیش) دیابت تیپ ۲ (از ۸ سال پیش) و فشار خون (از ۳ سال پیش) به علت تشدید علائم آسم پس از تمام شدن داروها مراجعه کرده است. در صورت مصرف منظم داروها شکایتی ندارد.
از درد مکانیکال زانوها با ارجحیت سمت راست شکایت دارد
- بیمار ذکر می‌کند از ۱۰ سال پیش مبتلا به کیست هیداتید است و جهت پیگیری آن سونوگرافی انجام می‌دهد. در آخرین سونوگرافی کبد، کیسه صفرا و مجاری صفراوی به تاریخ 23 آبان:
- تصویر کیست دارای سپتاهای داخلی و محتوای low level echo با ابعاد ۴۹*۶۰ م.م و ۲۱*۲۵ م.م در مجاورت یکدیگر در سگمان ۴ کبد رویت شد. احتمال کیست هیداتید در درجه اول مطرح است.
- درد شکم ، تهوع ، استفراغ ، تغییر وزن اخیر ، تغییراشتهای اخیر ، اسهال و بیبوست : ندارد

History

• PMH: آسم (از ۱۰ سال پیش) دیابت تیپ ۲ (از ۸ سال پیش) و فشار خون (از ۳ سال پیش)

• DH:

Neb symbicort (Budesonide 320/ Formoterol 9) BD

Tab Airokast (Montelukast 10) daily

Tab Melijent-M (Metformin 500/ Linagliptin 2.5) BD

Tab Losartan (unknown dose) daily

• FH , SH : Neg

• Vitals : BP 137/98 , PR 78 , FBS 153

• معاینه : S1 , S2 بدون سوفل / شکم نرم و بدون تندرns / سمع ریه clear / بیمار ill , toxic , icterus

pale نبود/ فورس اندام ها 5/5

INTRODUCTION

- Echinococcal disease is caused by infection with the metacestode (larval) stage of the tapeworm *Echinococcus*.
- prevalent in most areas where livestock is raised in Association with dogs
- The definitive hosts are canines , intermediate hosts sheep,cattle,humans,goats, camels, and horses (*E. granulosus*) mice and other rodents (*E. multilocularis*)
- Four species of *Echinococcus* produce infection in humans;
E. granulosus and *E. multilocularis* : most common, causing cystic echinococcosis (CE) and alveolar echinococcosis (AE), respectively.
E. vogeli and *E. oligarthrus* : cause polycystic echinococcosis , rarely associated with human infection

Clinical manifestations

(E. GRANULOSUS)

Signs and symptoms

- The initial phase of primary infection is always asymptomatic. (Latent periods of more than 50 years reported). The **clinical presentation** of *E. granulosus* infection depends upon the **site and size**. Small and/or calcified cysts may remain asymptomatic indefinitely. However, symptoms due to mass effect within organs, obstruction of blood or lymphatic flow, or complications such as rupture or secondary bacterial infections can result . Hydatid cysts may be found in almost any site of the body(primary inoculation /secondary spread.)
- The liver is affected in approximately two-thirds of patients,
- The lungs in approximately 25 %
- Other organs :brain, muscle, kidneys, bone, heart, pancreas ...

Liver involvement

- Frequently produces **no** symptoms.
- The right lobe is affected in 60 to 85 % of cases.
- Significant symptoms are unusual before the cyst has reached at least **10 cm** in diameter.
- If the cysts become large, **hepatomegaly** with or without associated **right upper quadrant pain, nausea, and vomiting** can result.
- In approximately one-fourth of the cases, *E. granulosus* cysts **rupture** into the **biliary** tree, producing **biliary colic, obstructive jaundice, cholangitis, and/or pancreatitis**.
- Less commonly, cysts **rupture** into the **peritoneal** cavity or other organs, with potential for **peritonitis , anaphylaxis ,** transdiaphragmatically into the pleural space or bronchial tree, causing **pulmonary hydatidosis** or a **bronchial fistula** and **multiorgan failure**.
- **Pressure or mass effect** on the bile ducts, portal and hepatic veins, or the inferior vena cava can result in **cholestasis, portal HTN, venous obstruction, or Budd-Chiari syndrome**.
- Secondary bacterial infection of the cysts can result in liver abscesses. (**Pyogenic liver abscess**)

Lung involvement

- The most common symptoms of pulmonary cystic echinococcosis (CE) :
- **Cough** (53 to 62 %)
- **Chest Pain** (49 to 91 %)
- **Dyspnea** (10 to 70 %)
- **Hemoptysis** (12 to 21 %).
- Less frequent : **malaise**, **nausea** and **vomiting**, and **thoracic deformations** .
- Cysts can break or develop secondary bacterial infection wich can manifest as a **pulmonary abscess** with **poorly defined margins**.
- complication of cyst rupture : pleural cavity involvement can cause **pneumothorax**, **pleural effusion**, or **empyema**.
- 60 % of pulmonary hydatid disease affects the **right lung**, and 50 to 60 % involve the **lower lobes** .
- Multiple cysts are common.
- 20 % of patients with lung cysts also have liver cysts

Other organs

- unusual but can lead to significant morbidity and mortality
- **Heart** : can result in mechanical rupture with widespread dissemination or **pericardial tamponade**
- **CNS** : can lead to **seizures** or signs of **raised** intracranial pressure; infection of the spinal cord can result in spinal **cord compression**.
- Kidney : can cause **hematuria** , **flank pain** , **glomerulonephritis** leading to the **nephrotic syndrome**, and **secondary amyloidosis**.
- **Bone** : growth of the parasite is a very slow , cysts are usually **asymptomatic** until a **pathologic fracture** develops; the spine, pelvis, and long bones are most frequently affected
- Other : Ocular , Subcutaneous cyst
- **Cyst rupture** : **Fever** and acute hypersensitivity reactions, including **anaphylaxis**

Diagnosis

- Generally made by **imaging** in conjunction with **serology**.
- Laboratory findings : Nonspecific leukopenia or thrombocytopenia, mild eosinophilia, and nonspecific liver function abnormalities may be observed but are not diagnostic. Eosinophilia is observed in fewer than 15 % of cases and generally occurs only if there is leakage of antigenic material.
- **Imaging** :

Ultrasonography : most widely used, easy to perform, inexpensive.

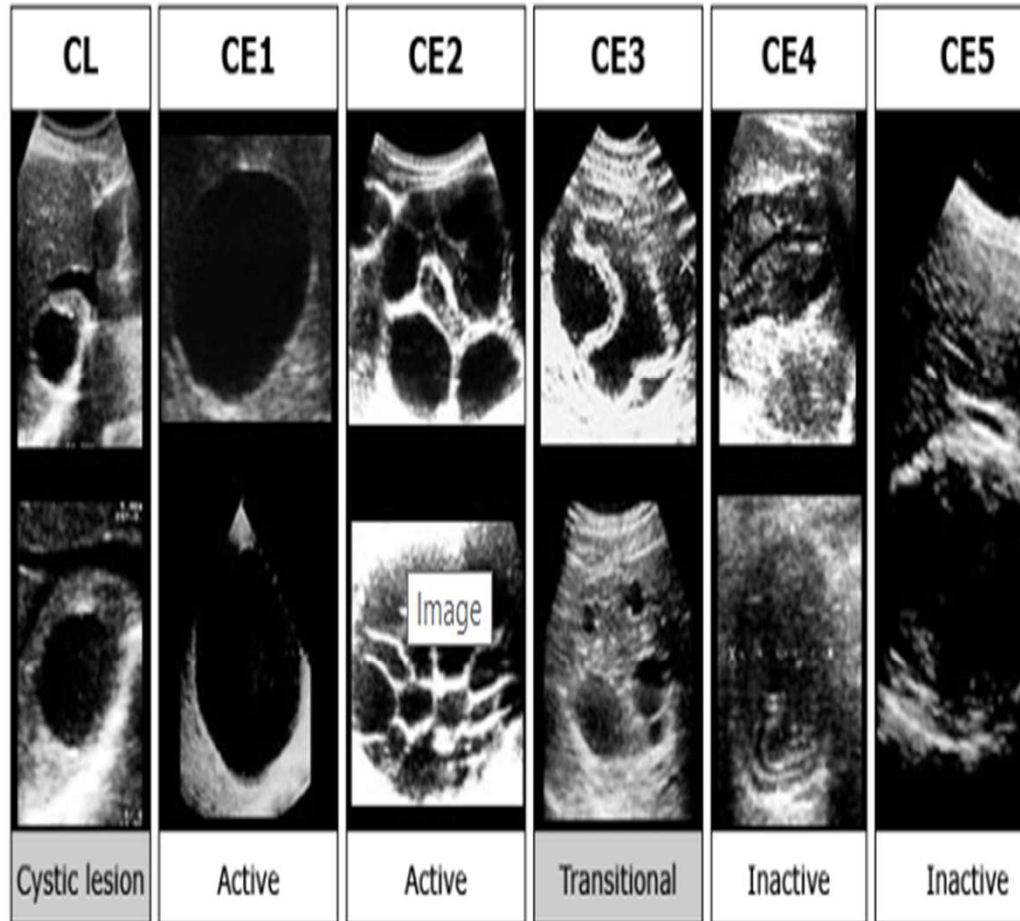
CT or MRI : useful for circumstances in which **greater anatomic detail** is needed to establish the location and number of cysts, the presence or absence of daughter cysts, and presence of ruptured or calcified cysts

Plain radiography : may demonstrate calcification within a cyst but **cannot detect uncalcified** cysts so is not adequate for definitive diagnostic evaluation.

Ultrasonography

- The sensitivity is 90 to 95 %
- The most common appearance : **anechoic, smooth, round cyst.**
- In the presence of **daughter cysts, characteristic internal septation** can be seen.
- hydatid sand : hooklets and scolexes from the protoscolices. Hydatid disease is probable in the setting of hydatid sand, inner cyst wall infoldings, and separation of the hydatid membrane from the wall of the cyst observed on ultrasound.
- Ultrasound allows classification of the cyst(s) as active, transitional, or inactive based on biologic activity; such categorizations may influence the choice of treatment.
- Several other classification systems are based upon ultrasound appearance such as WHO's classification

Ultrasonographic classification of cysts due to cystic echinococcosis



World Health Organization classification of cystic echinococcosis and treatment stratified by cyst stage

WHO stage	Description	Stage	Size	Preferred treatment	Alternate treatment
CE1	Unilocular anechoic cystic lesion with double line sign	Active	<5 cm	Albendazole alone	PAIR
			>5 cm	Albendazole + PAIR	PAIR
CE2	Multiseptated, "rosette-like" "honeycomb" cyst	Active	Any	Albendazole + either modified catheterization or surgery	Modified catheterization
CE3a	Cyst with detached membranes (water-lily sign)	Transitional	<5 cm	Albendazole alone	PAIR
			>5 cm	Albendazole + PAIR	PAIR
CE3b	Cyst with daughter cysts in solid matrix	Transitional	Any	Albendazole + either modified catheterization or surgery	Modified catheterization
CE4	Cyst with heterogeneous hypoechoic/hyperechoic contents; no daughter cysts	Inactive	Any	Observation	-
CE5	Solid plus calcified wall	Inactive	Any	Observation	-

Computed tomography

- Many reports suggest that CT has higher overall sensitivity than ultrasonography (95 to 100 percent) .
- best mode for determining the **number, size, and anatomic location** of the cysts and is better than ultrasound for detection of **extrahepatic** cysts.
- CT may also be used for **monitoring** lesions **during therapy** and to detect recurrences.
- CT may be superior to ultrasonography in **assessing for complications** such as infection and intrabiliary rupture.
- **ultrasound** is better than CT in the investigation of the **cyst wall, hydatid sand, daughter cysts**, and **splitting** of the **cyst wall**
- CT superior for detecting **gas** and **minute calcifications** within the cysts, in attenuation measurement, and in **anatomic mapping**

Magnetic resonance imaging

- MRI is usually not required and, in most instances, is not cost effective. Both CT and MRI are useful in diagnosing echinococcal infection **in other sites** such as in the brain.
- Advantages over CT : **defining changes in the intra- and extrahepatic venous system , delineate the cyst capsule** better, at diagnosing complications, particularly for cysts with infection or biliary communication

Serologic tests

- A negative test does not exclude the diagnosis of echinococcosis
- ELISA appears to be the most sensitive and specific of the available assays
- initial screening tests : ELISA and IHA
- Confirmatory tests using specific antigens can then be performed, such as immunoelectrophoresis and immunoblotting.(Detection of antibody to specific echinococcal antigens by immunoblotting has the highest degree of specificity.)
- Additional tests using recombinant or purified species-specific antigens may also be useful diagnosis

Interventional procedures

- **biopsy** : In the absence of a positive serologic test may be required to confirm the diagnosis . Percutaneous aspiration of liver cyst contents is associated with very low rates of complications. This method is generally reserved for situations when other diagnostic methods are inconclusive because of the potential risk for anaphylaxis and secondary spread of the infection .If aspiration is required, it should be performed under ultrasound or CT guidance; complications can be minimized by concurrent administration of **albendazole** and **praziquantel**
- **ERCP** : may be warranted to evaluate for biliary involvement, particularly in patients with cholestatic jaundice.

Alveolar Echinococcosis (E. Multilocularis)

- Clinical manifestations : usually symptomatic, although the clinical manifestations are frequently nonspecific.
- The most common : **malaise, weight loss**, and right **upper quadrant discomfort** due to **hepatomegaly. Cholestatic jaundice, cholangitis, portal hypertension**, and the **Budd-Chiari syndrome** can also occur. The clinical presentation may mimic that of hepatocellular carcinoma.
- Extrahepatic primary disease is very rare (1 %). Multiorgan disease was described in 13 % of cases
- Immunodeficiency, such as HIV or transplantation, may accelerate the manifestations of alveolar echinococcosis (AE)
- Nonspecific leukopenia or thrombocytopenia, mild eosinophilia, and nonspecific liver function abnormalities may be detected but are not diagnostic. Hypergammaglobulinemia and elevated serum IgE levels are present in more than 50 % of cases

Alveolar Echinococcosis (E. Multilocularis)

Diagnosis

- Generally made by imaging techniques in conjunction with serology.
- **Imaging** : On ultrasound or CT, the lesions usually have an **irregular contour** with **no welldefined wall**, **central necrosis**, and **irregular intralesional and wall calcifications**.
- The WHO Informal Work Group on echinococcosis has developed the **PNM classification system**, which stands for extension of the parasitic mass in the liver (P), the involvement of neighboring organs (N), and metastases (M). The system serves as a benchmark for the evaluation of diagnostic and therapeutic measures.
- **Obstruction** of the **inferior vena cava** or of the **portal venous system** may be evident, which may be more easily appreciated on **MRI**.
- Lung, brain, and bone lesions may also be detected.

PNM classification of alveolar echinococcosis

P	Hepatic localization of the parasite
PX	Primary tumor cannot be assessed
P0	No detectable tumor in the liver
P1	Peripheral lesions without proximal vascular and/or biliar involvement
P2	Central lesions with proximal vascular and or biliar involvement of 1 lobe*
P3	Central lesions with hilar vascular of biliar involvement of both lobes and/or with involvement of 2 hepatic veins
P4	Any liver lesion with extension along the vessels and the biliary tree [¶]
N	Extrahepatic involvement of neighboring organs (diaphragm, lung, pleura, pericardium, heart, gastric and duodenal wall, adrenal glands, peritoneum, retroperitoneum, parietal wall [muscles, skin, bone], pancreas, regional lymph nodes, liver ligaments, kidney)
NX	Not evaluable
N0	No regional involvement
N1	Regional involvement of contiguous organs or tissues
M	The absence or presence of distant metastasis (lung, distant lymph nodes, spleen, central nervous system, orbital, bone, skin, muscle, kidney, distant peritoneum, and retroperitoneum)
MX	Not completely evaluated
M0	No metastasis ^Δ
M1	Metastasis

PNM classification denotes the extension of the parasitic mass in the liver (P), involvement of neighboring organs (N), and metastases (M).

* For classification, the plane projecting between the bed of the gall bladder and the inferior vena cava divides the liver in two lobes.

¶ Vessels mean inferior vena cava, portal vein and arteries.

Δ Chest X-ray and cerebral computed tomography negative.

Alveolar Echinococcosis (E. Multilocularis)

Diagnosis

- **Serology** : The likelihood of a positive serology depends on cyst location and viability. Patients with liver cysts are more likely to be seropositive than patients with lung cysts. Serologic assays are less likely to be positive in the setting of calcified or nonviable cysts. In addition, the sensitivity and specificity of serology is greater for E. multilocularis than for E. granulosus
- **biopsy** If aspiration is required, it should be performed under ultrasound or CT guidance. Complications can be minimized by concurrent administration of **albendazole** and **praziquantel**

Treatment

- Management options for cystic echinococcosis (CE) include **surgery, percutaneous management, drug therapy, and observation**. In general, clinical approach depends on the World Health Organization (**WHO**) **diagnostic classification**
- In general, the approach to treatment of alveolar echinococcosis consists of **surgery** . Infected tissues should be removed as completely as possible, which requires complete excision of parasitic tissue and may also warrant radical resection of host tissue. In general, at least **two years of adjunctive drug therapy** is advisable in conjunction with at least **10 years of follow-up** monitoring for **recurrence**.

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CE5	Solid plus calcified wall	Inactive	Any	Observation	-

Albendazole is dosed 10 to 15 mg/kg per day in two divided doses; the usual dose for adults is 400 mg twice daily. Duration of therapy is discussed in the text.

WHO: World Health Organization; CE: cystic echinococcosis; PAIR: puncture, aspiration, injection, reaspiration.

Data from:

1. Junghans T, da Silva AM, Horton J, et al. Clinical management of cystic echinococcosis: state of the art, problems, and perspectives. *Am J Trop Med Hyg* 2008; 79:301.

Stage-specific treatment approach to alveolar echinococcosis

WHO Classification	Surgery	Interventional treatment	Drug therapy	Suggested	Resource setting
P1N0M0	X		X	Radical resection (R0) BMZ for 2 years PET/CT controls	Optimal
				Radical resection (R0) BMZ for 3 months	Minimal
P2N0M0	X		X	Radical resection (R0) BMZ for 2 years	Optimal
				Radical resection (R0) BMZ for 3 months	Minimal
P3N0M0			X	BMZ continuously PET/CT/MRI scan initially and in 2 years intervals	Optimal
				BMZ continuously	Minimal
P3N1M0		X	X	BMZ continuously plus PET/CT/MRI scan initially and in 2 years intervals	Optimal
				Surgery, if indicated	Minimal
P4N0M0		X	X	BMZ continuously plus PET/CT/MRI scan initially and in 2 years intervals	Optimal
				Surgery, if indicated	Minimal
P4N1M1		X	X	BMZ continuously plus PET/CT/MRI scan initially and in 2 years intervals	Optimal
				Surgery, if indicated	Minimal

R0: no residue following resection; BMZ: benzimidazoles; PET: positron emission tomography; CT: computed tomography; MRI: magnetic resonance imaging.

Reproduced from: Brunetti E, Kern K, Vuitton DA, Writing Panel for the WHO-IWGE. Expert consensus for the diagnosis and treatment of cystic and alveolar echinococcosis in humans. *Acta Tropica* 2009; 114:1. Table used with the permission of Elsevier Inc. All rights reserved.

Primary prevention

cystic echinococcosis :

avoiding close contact with dogs hand washing after handling dogs, and before handling food

Restrict home slaughter of sheep and other livestock

preventing dogs from consuming infected sheep viscera

Elimination of stray dogs

Administration of **praziquantel** treatment to infected dogs

Vaccination of sheep may also be useful for prevention

no available vaccine against the adult echinococcus infection in dogs



Primary Prevention

alveolar echinococcosis

avoidance of contact with foxes and other potentially infected definitive hosts (cats)

Wash hands with soap and warm water after handling dogs, and before handling food

Reducing contact between pets and rodent prey

intermittent prophylactic treatment of canids with praziquantel (Praziquantel-impregnated bait)

No effective vaccine against *E. multilocularis* has been developed

(Even if a vaccine became available, the primary cycle is almost always sylvatic, which makes a vaccination approach to control unlikely to be fully effective)

Secondary Prevention

- For asymptomatic individuals in endemic areas, screening with imaging and serology has been found useful in epidemiologic studies ; outside of endemic areas, the role of screening for asymptomatic individuals is uncertain
- The methods most frequently employed for initial screening tests (using crude antigens such as hydatid fluid or protoscolex extracts) are ELISA and IHA
- Simple, heat-stable, inexpensive tests, such as hydatid antigen dot immunoassays, are often used for field testing and population screening
- Portable ultrasound for screening patients in communities in which *E.granulosus* infection is endemic, sometimes with confirmatory serologic testing to maximize the diagnostic yield

Tertiary Prevention

- انتخاب روش مناسب برای درمان (رژیم دارویی و تکنیک جراحی مناسب)
- توجه به عوارض روش های تشخیصی و درمانی تهاجمی (بیوپسی ، ERCP ، PAIR) و انجام آن توسط افراد ماهر

Quaternary Prevention

- Antiparasitic agents should not be used in patients with significant underlying liver disease or bone marrow suppression



THANK YOU
FOR YOUR
ATTENTION