THE ROLE OF MULTIDISCIPLINARY TEAM IN MANAGEMENT OF HYDATID DISEASE

Dr/ Rasheed ALEEZI (JBGS, MHPBS)
Department of general surgery,
USThospital
Sana`a-YEMEN
58 years old, female patient, housewife.

Complaints:
severe repetitive attacks of productive cough, brown yellow to dark colour sputum with vomiting associated with night sweating and fever and abdominal pain one month PTA.

O/E:
- conscious, oriented, febrile, not jaundiced.
- Bp 130/80, pulse 95, RR 26, temp 38°C.
- bronchial and harsh breathing in lower zone of Rt. Lung
- mild tenderness in Rt. Hypochondrium.
Cystic lesion in lower zone of right lung.
Investigations

Abdominal and Chest CT.

- Hydatid cyst type 11 seen in vii and viii segment and Rt lobe of the liver.
- Hydatid cyst type 11 seen in the Rt lower zone of the lung with subtle Rt sided pleural effusion.
Treatment

- Pt prepared for operation
- Rt posteriolateral thoractomy
- Rt bronchobiliary fistula with large infected cyst at lower lobe filled with debris material
Fistula through diaphragm disconnected, cleaning and evacuation of both cyst, drain in liver cyst. Closure of diaphragm
Non specific chronic active inflammation with bronchobialiry fistula.
Hepatic hydatid cyst
CASE 2: SPLENIC HYDATID CYST

- Male 40y.
- C/O:
  - Lt upper abd.pain since 2 months, early satiety.
- O/E:
  - Huge splenomegally.
- Abd. CT: Huge spleen with large cystic mass.
- After preparation, total splenectomy done through lt Khocher incision.
AIM

To estimate the burden of Hydatid disease over the society.

Increase the medical awareness about this neglected disease.

Try to formulate an accurate pathway for diagnosis, management and prevention of the disease by review the literatures.

Emphasis on the role of multidispenary team in the management of this disease.
Hydatid disease or echinococcosis is a zoonosis that caused by echinochococcus granulosus and occurs primarily in sheep (intermediate host)-grazing areas of the world but is common worldwide because the dog is a definitive host.

Human is blinded host, infected by eating vegetables contaminated with egg, or plying with dogs.

Echinococcosis is endemic in Mediterranean countries, the Middle East, the Far East, South America, Australia, and East Africa. Humans contract the disease from dogs, and there is no human-to-human transmission.
Hydatid disease still an emerging problem and is a course of challenge to all the medical practitioners.

It is waging a war, with its roots spread deeply in society, it cause considerable morbidity and mortality and socio-economic loses in many developing countries.

Due to its non-specific clinical presentation and lack of awareness regarding the parasite in society, it is being overlooked very commonly.
There still exists a dilemma of obtaining accurate figures on the prevalence of Hydatid disease, as in majority of cases the disease manifests with a very few specific signs and symptoms.

A considerable number of cases present to clinician in an asymptomatic state and the diagnosis will be made incidentally or accidentally.

Hydatid disease is an endemic in Yemen.
This condition was only recently included in the WHO strategic plan (2008-2015) for the control of NTDs and were put under the neglected zoonosis subgroup.

NTD” is a collective term used for diseases which are: -
- more common in tropical countries .
- neglected by researchers and health policy makers .

Although effective chemotherapy and other treatment options are available for many of these diseases, these diseases still prevail amongst the poorest populations in the world where animal husbandry is common but no veterinary control exists.

According to WHO, this disease is called “neglected” as this has not been given enough importance at national or international levels.

neglected NTDs because it doesn't received much attention as compared with other public health problems due to:

- The diagnostic modalities required to detect this condition are not yet widely available and not affordable, especially in the areas where these infections are most prevalent.
- It is chronic conditions and clinical signs may be evident only after many years. Therefore, these conditions are grossly under-reported, ironically in areas where they are most prevalent.
CE has a cosmopolitan spread with cases being seen in both the developed as well as the developing countries.

*E. granulosus* has been reported from over 100 countries.

CE is of special importance due to its wide geographic distribution and socioeconomic impact, not only with respect to the medical costs associated with it but also with regard to the loss of livestock.

According to statistics given out by WHO, the annual societal cost of CE amounts to US $150 million in the subcontinent of India alone.

*World Health Organisation. Neglected zoonotic diseases (NZD) 2012*
Four species of Echinococcus cause infection in humans; *Echinococcus granulosus* and *Echinococcus multilocularis* are the most common, causing cystic echinococcosis (CE) and alveolar echinococcosis (AE) respectively.

The two other species, *E. vogeli* and *E. oligarthrus* cause polycystic echinococcosis and are less frequently associated with human infection.
The adult form of *Echinococcus granulosus* resides in the small intestine of dogs. The ova from the adult worm are shed through the canine feces into the environment, where the intermediate host sheep and humans ingest the eggs, in humans after entering proximal portion of the small intestine, the larvae burrow through the mucosa, enter the portal circulation and travel to liver. The cycle is completed when dogs eat the carcass of animals infected with the hydatid cysts.
Man is accidental host and is dead end.......
The hydatid cysts are mainly located in the liver as the liver acts as 1st filter in 60-70% of human infections as hexacanth embryos after hatching out penetrates intestinal wall and enter into radical of portal vein.

It can also be present in lungs as sometimes the embryo pass through hepatic capillaries and enter the pulmonary circulation, lung act as 2nd filter.

A few embryos may pass through pulmonary circulation too and enter various organs.

Thus rarely, the cysts may be found in the brain. eye, kidney, muscles, bones, spleen, genital organs.
Theoretically, echinococcosis can involve any organ.

Slowly enlarging echinococcal cysts generally remain asymptomatic until their expanding size or their space-occupying effect in an involved organ elicits symptoms.

Since a period of years elapses before cysts enlarge sufficiently to cause symptoms, they may be discovered incidentally on a routine x-ray or ultrasound study.

Organs affected by E. granulosus are the liver (63%), lungs (25%), muscles (5%), bones (3%), kidneys (2%), brain (1%), and spleen (1%).
LESION (ARROWS) IN BODY AND TAIL OF PANCREASE

b) CT scan showing cystic lesion in spleen and liver

c) Well defined, thin walled unilocular cyst (arrows) in orbit

d) Two thick walled cysts in lungs
1. CLINICAL PICTURE.
2. LAB.
3. IMAGING.
The clinical presentation of a hydatid cyst is largely asymptomatic until complications occur.

1- Local:
   depend on size & site:
   - Liver: Tender hepatomegaly, compression on bile ducts, portal veins.
   - Lung: dyspnea, cough, pain.
   - Brain: symptoms of increased tension, epilepsy.
   - Bone: erosion, fracture.

2- General:
   rupture of the cyst (due to increased tension, trauma or secondary infection) results in:
   - Release of hydatid fluid → anaphylactic reactions.
   - Implantation of germinal cells in other tissues → secondary cysts.
Generally, routine laboratory tests do not show specific results

1. Eosinophilia
2. Hypogammaglobulinemia (25%)
3. Eliza → sensitive (80%)
4. Immune diffusion & immunoelectrophoresis (show antibody to Ag 5)
5. Casoni: (Not used) low sensitivity (70%)

The ELISA test is useful in follow-up to detect recurrence
(3) IMAGING:

1. Plan x-ray calcification
2. U.S → daughter cysts, sand
3. C.T, MRI →
   - accuracy 98%
   - DD: Amebic, pyogenic
   - In AE → may resemble HCC
4. ERCP → in intrabiliary rupture (IBR)
**Figure 5.** Rupture of a type IV hepatic cyst into the biliary tree in a 48-year-old man who presented with acute onset of jaundice and pain.
Ultrasound: currently the primary diagnostic technique and has diagnostic accuracy of 90%.

Gharbi’s Classification

- **Type I**: pure cystic fluid Collection (spherical-oval, thick-walled)
- **Type II**: fluid Collection with membrane separation
- **Type III**: Fluid collection with septa
- **Type IV**: heterogeneous (hypoechoic-hyperechoic-intermediate) pattern
- **Type V**: completely calcified (Reflecting) walls
Although concept of management of liver hydatidosis is changing, surgery is still gold standard for complete cure. There is still controversy regarding the appropriate surgical technique.

**Available Options:**

1. Surgery
2. Chemotherapy
3. Intervention
   a) PAIR
   b) ERCP

- **Calcified cyst** are dead cyst. They are left alone.
- **Symptomless, small cyst** can be left alone. Once symptomatic or if the size is more than 5 cm they may be treated.
(1) Surgery:

Benefit: immediate, radical cure

**Indications:**
1. large liver cysts, with multiple daughter cysts.
2. Superficial single liver cyst (Rupture fear)
3. liver cyst with biliary communication
4. infected

**Contraindications:**
1. multiple cysts in multiple organs
2. difficult access, very small
3. dead & calcified,
Risks:

1. 2\textsuperscript{ry} anaphylaxis
2. Operative mortality
3. Recurrence

Technique:

- **Radical**:
  Total pericystectomy or partial affected organ resection.

- **Conservative**:
  Open cystectomy or simple tube drainage for infected and communicating cysts.
(1) **Surgery:- Laparoscopy**

- Laparoscopic approach has recently been applied for the treatment of uncomplicated liver hydatid cysts.
- Controversies about the role of LAP. In the management of HC have not been resolved; these controversies include selection of the patients and surgical techniques.
Benefit:
- Oral - easy

Indications:
1. inoperable liver or lung cysts
2. two or more organs
3. peritoneal cysts

Contraindications:

A) Absolute:
1) Early pregnancy
2) Bone Marrow suppressive
3) Chronic liver disease
4) large cyst (Rupture risk)
5) inactive (calcific) cysts.

B) Relative: bone cyst (low response)
1. **Albendazole**

Albendazole is administered in a dose of 10 – 15 mg/kg/day in adults or a fixed dose of 400 mg twice daily. The treatment is given in cycles of 28 days with two weeks treatment free periods between the cycles.

**The different schedules for the treatment are:**

1. **Inoperable cases** - as primary treatment - 3 cycles
2. **Pre-operatively** – to reduce the risk of recurrence 6 weeks continuous treatment
3. **Post-operatively** to prevent recurrence in cases of intraoperative cyst spillage – 3 cycles.

Response to treatment is best assessed by serial imaging studies, with attention to cyst size and consistency.
PAIR (Puncture, Aspiration, Injection, Re-aspiration), was proposed in 1986 by the Tunisian team that first used it in a prospective study.

PAIR is a relatively recent and minimally invasive therapeutic option, that complements or replaces surgery which was long considered as the only treatment for CE.
Confirmation of diagnosis as scolicidal agent is injected and aspirated.
(3) **INTERVENTION**

A) **PAIR**

**Benefits**

1. Minimal invasiveness
2. Reduced risk compared with surgery
3. Confirmation of diagnosis
4. Removal of large numbers of protoscolices
5. Improved efficacy of chemotherapy
6. Reduced hospitalization time
7. Cost of the puncture and chemotherapy usually less than that of surgery or chemotherapy alone

*WHO 2001*
Direct cholangiography, (Endoscopic or percutaneous) may be required in suspected intrabiliary rupture and bile duct obstruction. ERCP is also a valuable method for detecting post-operative complications involving the biliary tree following surgical intervention.

- biliary fistula
- hydatid material within the bile duct
- bile duct stenosis
- hydatid membrane on the gall bladder
- extrnsic compression to bile duct
Operative specimen of opened hydatid cyst showing multiple daughter cysts

**Follow Up**

**Chemotherapy:**
- Postoperative treatment with benzimidazoles is continued for 1 month in patients with CE who have undergone complete resection or PAIR successfully.
- The treatment is continued for 3-6 months for patients with resected AE, incompletely resected CE, spillage during surgery or PAIR, and metastatic lesions.

**Laboratory tests:**
- Patients on benzimidazoles should have a CBC count and liver enzyme evaluation performed at biweekly intervals for 3 months and then every 4 weeks to monitor for toxicity.
- ELISA or indirect hemagglutination tests are usually performed at 3-, 6-, 12-, and 24-month intervals as screening for recurrence of resected disease or aggravation of existing disease.

**Imaging:**
- Ultrasonography and/or CT scan are used in follow-up at the same intervals as the laboratory tests or as clinically indicated.
WHO/OIE Manual on Echinococcosis in Humans and Animals: a Public Health Problem of Global Concern

Edited by
J. Eckert, M.A. Gemmell, F.-X. Meslin and Z.S. Pawłowski

- Aetiology
- Echinococcosis in humans
- Echinococcosis in animals
- Diagnosis
- Treatment
- Ethical aspects
- Geographic distribution
- Surveillance
- Epidemiology
- Control
- Prevention
- Methods
In endemic areas, echinococcosis can be prevented

- by administering praziquantel to infected dogs,
- by vaccinating sheep.
- Limitation of the number of stray dogs is helpful in reducing the prevalence of infection among humans
- by denying dogs access to infected animals.

Strict hygienic measures are essential.

- Raise the awareness of modes of transmission of the disease to human being may be helpful through health media
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- control of dogs (responsible ownership, registration, movement control and collection of stray dogs with euthanasia if dogs are not claimed)
- treatment of imported dogs with praziquantel
- prevention of smuggling of food animals
- safe destruction or deep burial of carcasses or offal of food animals
- safe slaughtering
- continuing education
- special measures in infected villages (see below).
Hydatid disease is still an important health problem in Yemen, with its expansion in various parts of Yemen. It is still raging a war over the communities from low socioeconomic status who are unaware about this demonic parasite.

Thus,

there is a quite an impact of morbidity due to Hydatid disease over the society due to its higher infliction rate in farmers and housewives.
Female are more prone to develop hydatid disease than males due to they stay in their home more than male which give them more chance to be in contact with animal and food staff that my transmit the disease.

Rural are more affected than urban as our social and religions concepts doesn't accept keeping dogs in their homes but in rural area they use dogs as a guard in addition there is a lot of such animals outside home.
The true picture of hydatidosis in YEMEN IS difficult to be established because of:

- many cysts remain asymptomatic and some of the patients never seek medical advice.
- Furthermore, most data are not available in the medical records.
- Limitation of published studies about this subject
Figure 1. Frequency of hydatid cyst cases during the period 2001 – 2008 in 5 hospitals in Sana’a city
We conclude that the risk of hydatidosis is still high in Yemen, where street or stray dogs move freely down town and the population should be aware about the role of dogs in the transmission of this disease.

Hospital records provide a useful indication of infection expressed as annual rate of hospital cases.
In conclusion, Hydatid disease is still an important health problem in Yemen which needs to be studied further. Therefore, accurate information on the distribution of the disease should be the first step for the control and prevention of the disease.

Hydatid disease still an emerging problem and is a course of challenge to all the medical practitioners.

According to WHO, this disease is called “neglected” as this has not been given enough importance at national or international levels.

Although concept of management of liver hydatidosis is changing, surgery is still gold standard for complete cure.

Accurate information on the distribution of the disease should be the first step for the control and prevention of the disease.

Emphasis on the role of multidisciplinary team in the management of this disease.

Finally, the collaboration of Public Health Authorities, the Veterinary Medical Authorities and the Environmental Affairs Authorities is a must to control this disease.
THANK YOU