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Chapter

Introductory Chapter: Overview on Echinococcosis

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1. Introduction

Echinococcosis is considered the most common parasitic disease and the most widespread zoonotic infection caused by several cestode species belonging to the genus *Echinococcus* [1].

This parasite has 12 different genus types, though four major species are known to cause human parasitic disease: *Echinococcus granulosus* (*E. granulosus*), *Echinococcus multilocularis*, *Echinococcus vogeli*, and *Echinococcus oligarthrus* [2].

E. granulosus is the causative organism of cystic echinococcosis, a small-sized tapeworm measuring approximately 2–7 mm in length with at least 10 different genome patterns and distinct genetic markers [1].

2. Parasitology

The parasitic life cycle involves two mammalian hosts. The intermediate hosts include members of the ungulates such as sheep, goats, and pigs. The definitive host of this parasite is the dog and other members of canids.

The adult parasite inhabits the small intestine of a definitive host and eggs, containing infective oncospheres, are produced. Subsequently, free eggs as well as cestode segments are excreted through the stool of the host into the environment [1].

Following an oral uptake of eggs by an intermediate host, a larval stage (metacestode) develops in several internal organs, mainly the liver and lungs, after a larval migration through the blood and lymph vessels. Typically, numerous protoscoleces are produced by the mature metacestode, each with the potential to develop into an adult cestode after oral ingestion by a suitable definitive host [3].

Humans are accidental or aberrant hosts that are not essential to the natural *Echinococcus* life-cycle. The eggs, after being ingested, develop as a cyst (hydatid cyst) inside the accidental host organs. Larval infection leading to the hydatid disease is characterized by metacestode long-term growth in the intermediate host. The parasitic cystic structure is typically filled with a clear hydatid fluid [4]. The inner germinating membrane of the cyst provides germination and the outer cystic layer features a laminated part. A granulomatous inflammatory reaction leads to a fibrous tissue constitution walling the cyst [5].

Developing cysts are responsible for the morbidity and the mortality related to the disease [3].

Cystic echinococcosis (CE) occurs worldwide. *E. granulosus* is known to occur on all continents but high parasite prevalences are found in endemic areas such as central Asia, the Mediterranean Basin, the Russian Federation, Africa (northern and eastern regions), Australia, and South America [1, 3].

Although Echinococcosis is regarded as an eradicable disease, it leads to serious economic losses for public health systems and agricultural sectors in endemic areas [6]. Therefore, numerous control programs reveal that preventive measures for the interruption of parasitic life cycle may relatively reduce the prevalence and the incidence of the hydatid disease [3, 7].

3. Diagnosis

3.1 Clinic

CE clinical presentation reflects the presence of one or more unilocular fluid-filled cysts. Liver and lungs are most commonly affected: about 70% of cysts involve the liver and the lungs are involved in 20% followed by other organs such as spleen, kidney, brain, breasts, and bone.

The disease course is typically slow and most patients have an asymptomatic disease course for several years. Otherwise, the cystic slow growth rate is estimated at 1–5 mm per year and patients develop symptoms as the cyst gradually grows [5, 8–10].

In the early stage of echinococcosis, clinical manifestations are mild. At later stages, damaged tissues and organs can become dysfunctional with the occurrence of symptoms related to cystic complications.

Symptoms are nonspecific and the diagnosis is often incidental, based on immunological tests and imaging such as standard radiology, ultrasonography (US), computed axial tomography (CT), and magnetic resonance imaging (MRI).

3.2 Serology

Serologic diagnostic methods play an important complementary role. They are used in order to support the radiological diagnosis and for follow-up of patients after surgical or pharmacological management.

There are multiple immunological responses to the disease. Mild immune response is noted with rugged and intact cysts, whereas complicated cysts (leaking or ruptured cysts) tend to show a strong immune response [4].

Serodiagnosis consists of the detection of specific serum antibodies by multiple immunodiagnostic tests. An optimum test should be specific with high sensitivity. There are considerable differences between the various serological tests, concerning the specificity as well as the test sensitivity.

In routine laboratory applications, the indirect hemagglutination (IHA) is often nonspecific, while the enzyme-linked immunosorbent assay (ELISA) using crude hydatid cyst fluid has a high sensitivity (up to 95%) with low specificity. Concomitant use of both tests (IHA and ELISA) is associated with diagnostic sensitivity over than 85–96% [1, 11].

False-positive serological findings are revealed in 20% of patients with CE, which is mainly related to cross-reactions with other parasitic diseases. Furthermore, seronegativity rate is relatively higher in cases with nonactive disease stages, in patients treated for autoimmune or malignant pathologies and during pregnancy [12]. Cysts in the bone, brain and calcified cysts often show no or low immunological response [5].

Immunoblot analysis is generally used when IHA and ELISA findings are not definitive. The immunological diagnosis is established by the detection of specific serum antibodies. Immunological methods often use the most specific antigens: *E. granulosus*, antigen B, and antigen 5 (Ag5) [1, 13].

3.3 Imaging

US is considered as the first choice imaging modality in the diagnosis and follow-up. It is a convenient and efficient imaging tool that demonstrates the cystic number, location, and size. Furthermore, US can guide the interventional radiology, which is often available, safe, and cost-efficient method with no radiation. Therefore, it can be used in endemic areas with large populations to make a prompt diagnosis. It mainly explores abdominal location.

Typically, the liver involvement is shown as a mixed echogenic, heterogeneous pattern with irregular contours with multiple distributed calcific foci, whereas, the radiologist must be aware of differential diagnosis in less typical appearance. The criteria for ultrasonographic cystic classification of liver was first made by Gharbi in 1981, and then, improved by the World Health Organization (WHO) in 2001 [5]. Cysts in other sites like the brain and the lung are not well demonstrated with US and it require the use of other screening techniques. Conventional radiography can be required as the initial imaging modality to diagnose thoracic and bone involvement. The X-ray findings are nonspecific and are visualized as multiple small opacities [14].

CT and MRI are mainly used in some cases, where US does not provide definitive findings. These imaging tools are necessary for illustrating the lesions morphologic features in intra-abdominal organs, which are involved through a direct invasion from the primary lesion or disseminated disease.

They are also indicated in patients with sub-diaphragmatic location, extra-abdominal sites (in bone and soft tissues involvement), complicated cysts such as cysto-biliary fistulae and abscess, cases with extra-abdominal dissemination. Moreover, CT and MRI can be useful for the preoperative evaluation and for patient's follow-up examination.

The radiologist should be familiar with multimodality imaging findings of this particular disease. Thus, according to the organ involvement, the lesion can be confused with other differential diagnosis including neoplasms, metastatic neoplasms, or abscess [14].

4. Treatment

There are several treatment approaches for echinococcosis, including monitoring (watch and wait strategy), chemotherapy, interventional radiology, and surgery. Indications for one or more management options are complex and depend on the cyst features, medical and surgical equipment availability and the patient's cooperation. Therefore, it is preferable to refer patients to reference treatment centers for echinococcosis in order to get the suitable management [15].

4.1 Medical treatment

The choice of the therapeutic modality is related to cyst stage, size, site, and comorbidities.

Medical treatment using parasitostatic drugs such as benzimidazoles (mebendazole and albendazole) is mainly recommended for inoperable patients and patients with disseminated disease. Drug therapy should be administered continuously for at least 3 months. However, prolonged treatment has a high cost with multiple teratogenic side effects [16]. Moreover, chemotherapy can be used as a complementary therapy to surgery or to prevent metastasis and secondary cystic sites. However, it is not recommended in the treatment of inactive or calcified cysts [17].

The watch and wait strategy is a therapeutic option for uncomplicated cysts with multiple involvements. It requires a regular long-term monitoring of liver function tests and leukocyte counts [17].

4.2 Percutaneous treatment

The percutaneous treatment modality is defined as an option for the management of cysts in the liver and other abdominal locations. This procedure is divided into two techniques: (1) PAIR technique is the best known, consisting of the destruction of the germinal membrane following the use of a scolicidal agent and (2) modified catheterization techniques aiming at the entire endocyst evacuation. This approach is a mini invasive procedure comparing to the surgery, but cysts containing daughter vesicles are not the best indication for percutaneous treatment because of the high risk of spread into the peritoneal cavity and into the biliary tract [5].

4.3 Surgical treatment

Surgery should be carefully evaluated. The decision-making is based on the characteristics of the hydatid cyst as the cyst type, number, size, location, and the presence or not of associated complications. It is well recommended for large cysts containing multiple daughter vesicles, symptomatic and complicated cysts, cysts with superficial location that may be ruptured spontaneously or following a benign trauma, infected cysts and cysts with close contact with vessels or adjacent vital organs. Furthermore, it can be an option for patients not suitable for percutaneous treatment.

The surgical approach is aiming to parasite inactivation, evacuation of the endocyst with the prevention of contamination, germinal layer removal, and residual cavity management [18].

Regarding the abdominal sites (typically the hepatic involvement), there are two surgical approaches consisting of a conservative surgery and radical procedure using open or laparoscopic surgery [19].

Conservative procedures consist of the parasitic cyst contents removal such as daughter vesicles and germinative membrane, whereas the pericyst is retained. The residual cavity is carefully explored to research any evidence of cystic biliary tract communication and then managed according to different techniques: capitonnage, omentoplasty, or external drainage.

Surgeons should cover the operating field with a scolicidal agent in order to avoid the parasites spillage and the peritoneal cavity contamination.

Conservative surgery is simple, safe with relatively reduced operative time, but has high morbidity and recurrence rates [19].

Radical surgery is the first therapeutic choice suitable for total excision of the entire parasitic lesion. Whenever possible, complete resection is required because radical surgical procedures are superior to conservative surgical methods and may cure definitively the patient [18].

The radical surgery target is the removal of the whole cyst with the parasitic contents and pericystic tissue.

Radical procedures includes: partial cystectomy, total cystic removal through an open or closed pericystectomy, and hepatic resection.

During the pericystectomy, a proper cleavage plane between the inner layer and the outer cystic layer may limit the liver parenchyma damage.

Hepatic resection is more difficult, takes longer operative time with higher risk of blood loss but lower cystic recurrence rates.

Comparing to conservative treatment, radical surgery is preferable due to lower morbidity, mortality, and reoccurrence rates [19].

Laparoscopic management of hydatid cysts with abdominal (hepatic or extra-hepatic sites) has its place among management approaches. It is a mini invasive surgery that can be safely achieved in particular cases after patients' selection.

Laparoscopy has several advantages including: better visualization of the peritoneal cavity and internal organs, prompt postoperative discharge, limited postoperative morbidity, and good esthetic outcomes [18].

There is lower frequency of hydatid disease in extra-hepatic sites. The management and the proper therapeutic approach depend essentially on cystic site and the organ involved. Generally, when the removal of the cysts is complete, the prognosis is good with low rate of recurrence [20].

5. Follow-up

A long-term follow-up for patients managed for hydatid disease should be planned because recurrences may occur in some cases. The patient monitoring is mainly based on imaging techniques (US, CT, and MRI) at short intervals. Serological tools and specific serum antibodies dosage support imaging techniques and can reflect metastode viability. Moreover, the monitoring of parasitostatic plasma level is necessary to adjust the therapeutic range and to prevent long-term treatment side effects [20].

Currently, planning for the echinococcosis control relies on the interruption of parasitic life cycle. The disease can be prevented by hygiene improvement in the slaughtering of livestock, public education campaigns, periodic deworming of dogs, and adequate destruction of infected offal. Vaccination of dogs with recombinant proteins provides encouraging prospects for prevention and control [21].

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