

EDITORIAL

Controversial issues in the management of echinococcal liver disease

Karaciğer hidatik kist tedavisinin sorunları

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In this issue of The Turkish Journal of Gastroenterology, Bülbüller et al. (1) presented their experience on the results of surgical treatment of hepatic Echinococcal (hydatid) cysts. Echinococcal disease continues to be a major health problem in developing countries in sheep-raising areas, which makes a large portion of the world. Public education directed at the proper disposal of cyst containing viscera of slaughtered animals (preventing their consumption by carnivores) can play an important role in the eradication of the disease. However, public health measures do not seem to work in a lot of countries and second to the fact that large populations of humans are on the move, physicians everywhere nowadays are faced with patients with the disease having to decide on a management strategy. The liberal use of sonography, CT and MRI has also increased the number of asymptomatic patients who come to the attention of physicians.

A review of recent medical literature discloses 3 alternative strategies for the management of cystic echinococcosis of the liver. The proposed modalities are; surgery, percutaneous drainage and chemotherapy. A follow-up without intervention strategy can also be implemented on asymptomatic patients based on the follow-up findings by Frieder et al on a small group of patients (2).

The issue of classification: The availability of more than one therapeutic modality necessitates the use of a comprehensive classification system to triage patients to different therapeutic modalities and analyse results. The Gharbi classification (3) which is extensively used in our country is based on the sonographic morphological appearance of liver lesions and does not take into account the phenomenon of rupture which is the most important event in the natural history of the disease. It is a well known fact that the majority of hydatid complications occur secondary to rupture and rup-

ture causes the disease to become symptomatic in the majority of patients.

The treatment of complications second to rupture or trying to protect the patient from rupture is the main rationale for the treatment of echinococcal liver disease. So, an ideal hydatid classification should also distinguish lesions that are prone to rupture from lesions that are not. Radiological signs which may indicate that a lesion is prone to rupture include large lesion size, rapid lesion growth on follow up examinations, a ruptured endocyst (floating membranes), thin walls, small biliary communications and weakness in the pericyst which is manifested by an exophytic component (reminiscent of a saccular aneurysm in a blood vessel) (4).

The WHO classification which was proposed in 2002 (WHO informal working group) has the same drawback as the Gharbi classification of not taking into account the phenomenon of rupture and being applicable solely to the liver and the sonographic examination. The Lewell and Mc Corkell classification of rupture (5) is an excellent classification for rupture but lacks detail in the classification of nonruptured lesions. A very useful classification would be a hybrid classification combining the WHO classification with the Lewell and Mc Corkell classification.. We reported such a classification which is applicable to all imaging modalities, in 1993 (6) Grade 1: Purely cystic unilocular lesion. Grade 2A: Lesion with complex morphologic appearance (CMA), predominantly fluid. Grade 2B: Dilatation of biliary canaliculi around a lesion with CMA. Grade 3: Lesion with intrabiliary rupture. Grade 4: Lesion with direct rupture into surrounding organs and spaces. Grade 5: Solid appearance or densely calcified lesion. This classification is also applicable to other organ systems. Biliary involvement can be substituted with airway involvement in the lungs and pelvicalyceal involvement in the kidneys.

MANAGEMENT STRATEGIES

Follow-up without intervention: The Frider paper published in 1999 reported the results of the follow-up of 33 asymptomatic hydatid patients followed-up for 10-12 years. 21/28 patients remained asymptomatic and most of the lesions had static cyst size. Although the number of followed patients is small, this paper suggests that asymptomatic echinococcal liver disease is not an aggressive disease. Therefore a treatment strategy with high morbidity and mortality rate is not justified.

Medical therapy: Benzimidazole compounds especially albendazole (ABZ) is known to be an effective agent against Echinococcal disease. However, medical treatment seems to be effective in less than 50% of treated patients when the goal of therapy is defined as complete cure manifested by disappearance of the lesion (7). We all know that the natural history of Echinococcal disease is very slow. As the main rationale of treatment in asymptomatic patients is the prevention of complications, success may alternatively be defined by lack of complications in medically treated patients.

As stated earlier most hydatid complications occur secondary to high pressure in the cavity. Pressure is caused by the production of hydatid fluid by the metabolically active parasite. It has been shown that 3 weeks of pre-operative Albendazole therapy reduces pressure in the cavity (8). In another study no viable protoscolices were found at surgery in 94% of patients following 10 mg/kg Albendazole for 3 months (9). When less rigid criteria are used to define success such as >25% reduction in cyst size, membrane separation, or cyst calcification, 82% of cysts out of 882 evaluable cysts were found to respond to Albendazole therapy (10). With only 6% of side effects requiring stopping treatment (10) medical therapy seems to be a safe and effective mode of therapy, especially for asymptomatic patients.

Percutaneous therapy: Several techniques for percutaneous therapy for echinococcal cysts with very high success rates and very low complication rates have been described in the literature (6). However, unlike most of the surgical series there is a lesion selection bias in percutaneous therapy. All series reported in the literature have used a selection bias and performed therapy on lesions that are predominantly fluid and nonruptured. Furthermore, most patients in the reported series

have received chemotherapy in addition to percutaneous therapy. We may conclude that percutaneous therapy is a hybrid treatment combining the therapeutic effect of the injected scolecidal agent and drainage and the systemically administered chemotherapeutic.

Surgery: Surgery combined with chemotherapy using various technical approaches (including laparoscopic surgery) remains the gold-standard therapeutic modality for echinococcal liver disease. Surgery was defined as the only definitive and curative modality by the WHO informal working group. However, surgery in its various forms (radical or nonradical and laparoscopic) has a considerable morbidity and mortality rate. A meta-analysis comparing the clinical outcomes for 769 patients treated with percutaneous aspiration-injection-reaspiration (PAIR) plus albendazole or mebendazole with 952 era-matched historical control subjects undergoing surgical intervention found a higher cure rate, lower rates of morbidity and mortality, lower recurrence rate and shorter hospital stay in the PAIR group (11). Therefore, surgery should be reserved for symptomatic patients who are not good candidates for percutaneous therapy.

Conclusion: Surgery has been accepted as the treatment of choice for cystic echinococcal liver disease for several years. Although numerous surgical procedures have been described; some of them could not find a clinical application. This means that there is no completely satisfactory surgical method.

The main purposes of surgical treatment are to inactivate scolices, to prevent spillage of scolices into the abdomen and to obliterate the residual cavity. Several surgical methods have been introduced to eliminate the cavity problem which was thought to be the predisposing factor of postoperative complications. This is the reason why surgical methods such as marsupialisation or simple drainage have been abandoned. An unobliterated cavity is unavoidable following a PAIR procedure and it usually does not cause any complications (12, 13).

Rupture into the biliary tract is a severe complication of the disease (14, 15). Formerly, choledocoduodenostomy and T-tube drainage were effective methods used to solve this serious complication. Nowadays, ES is the preferred method to treat this complication.

Postoperative biliary fistula is another problem. In the authors' series, this was observed in two cases. ES was not used in any of these cases with postoperative biliary fistula or with cholangitis due to biliary rupture of the cyst. In this situation, ERCP and ES might have been preferred instead of the surgical intervention (16, 17).

The incidence of asymptomatic patients reaches 60% in some series. In present series it is reported as 11% although the authors state that "most of their cases were asymptomatic". As stated above we believe that a more conservative approach

(medical therapy and/or percutaneous therapy) should be preferred in asymptomatic patients.

Another treatment option which should be mentioned in the management of hydatid cysts is laparoscopic surgery. Although there are some disadvantages in centrally located cysts and cysts close to the major biliary ducts, encouraging results have been achieved in some series. Despite the relatively limited indications in comparison with open surgery, it is expected that laparoscopic hydatid cyst surgery will improve and will take its place amongst the therapeutic modalities of hydatid cysts (18, 19).

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Ali EMRE¹, İzzet ROZANES², Koray GÜVEN²

Department of ¹General Surgery, ²Radiology, Istanbul Faculty of Medicine, Çapa, Istanbul