

**Disseminated Alveolar Echinococcosis**

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**Abstract**

Alveolar echinococcosis (AE) is caused by the larval form of the tapeworm Echinococcus multilocularis. In humans, E. alveolaris metacestode cells proliferate in the liver inducing a hepatic disorder that mimics liver cancer and can spread to other organs. From 1960 to 1972 mortality was at 70% and 94% after 5 and 10 years of follow-up, respectively. Since then, studies have shown an increasing trend towards improving survival rates [1]. As AE is also spreading to new areas of Eastern Europe, researchers seek to better understand the clinical presentation of pathology, including asymptomatic forms.

**Clinical case:** One 36-year-old woman from Peshkopia has been admitted to the Gastrohepatology department on 20.07.2011 with fatigue, anorexia, dull pain in right hypochondrium, mild epigastric pain, bloating, and weight loss. The epidemiological anamnesis showed that the patient lived in the village and had pets. On physical examination, the patient appeared severely ill with jaundice, massive hepatomegaly, massive mass in the mesogastric area, and anxiety. Laboratory examinations were as follows: Hb 11.1 g/dl; sediment 25 mm/h; leukocytes 6700/mm\(^3\); platelets 127000/mm\(^3\); prothrombin level 60%, uremia 12.7 mmol/l; creatinine 0.78 mmol/l; ALP 127 U/l; AST 15 U/l; ALT 37 U/l; GGT 131 U/l; bilirubin 3.7 mg/l, albumin 2.8 gr /l, total protein 8.1 gr /l, HbsAg negative, anti-HCV negative. Regarding serology, the titer of anti-echinococcal antibodies was positive (22, n = 11)

**Conclusions:** Clinical presentation and radiologic imaging findings of disseminated alveolar echinococcosis can mimic metastatic malignancy, and diagnosis can be challenging in atypically advanced cases. As the incidence of human alveolar echinococcosis appears to be increasing and, physicians should be aware of alveolar echinococcosis, its epidemiology, and its clinical features.

**Keywords:** Alveolar echinococcosis; Echinococcus multilocularis; Zoonosis.

**Introduction**

Alveolar echinococcosis (AE) is caused by the larval form of the tapeworm Echinococcus multilocularis. In humans, E. alveolaris metacestode cells proliferate in the liver inducing a hepatic disorder that mimics liver cancer, and can spread to other organs. From 1960 to 1972 mortality was at 70% and 94% after 5 and 10 years of follow-up, respectively. Since then, studies have shown an increasing trend towards improving survival rate [1]. As AE is also spreading to new areas of Eastern Europe, researchers seek to better understand the clinical presentation of pathology, including asymptomatic forms.

E. Multilocularis is transmitted to intermediate hosts by ingestion of eggs which enter the human body through fruits, vegetables or drinking water and is transmitted to final hosts by eating infected organs. People who are intermediate hosts are accidentally infected by touching the soil, mud or animal hair that contains the parasite eggs [1, 7]. The parasite has a life cycle that requires both intermediate and final hosts. It is the oncosphere membrane that makes eggs tolerate external environmental conditions which can remain contagious at temperatures ranging from
-30 degrees Celsius to +60 degrees Celsius. They are easily destroyed by heat, but can withstand months or years at low temperatures. Carefully follow-up examinations in these patients must continue for at least 10 years.

TREATMENT with Albendazol is mandatory for at least 2 years following in inoperable patients. Depending on the location of the cyst in the body, patients may be asymptomatic, although the cysts may be quite large in size, or symptomatic, although the cysts may be quite small. As a summary of clinical signs, we can mention: mild abdominal pain in the upper right quadrant is the most common symptom and can last for years until the lesions develop, jaundice is observed and may be associated with acute abdominal pain in the upper right quadrant, if parasitic material migrates to the common bile duct, hepatomegaly, and fever in the presence of bacterial infection [9, 12].

Diagnosis of alveolar echinococcosis consists of clinical features and exposure history, imaging examination, and serology. Symptoms and signs are different because of site of infection. The liver is the most common site of infection. Imaging shows a multivesicular solid mass, sometimes with central necrosis. Definitive diagnosis can be decided with histopathology. Of particular importance are serological tests (ELISA or western blood) in the diagnosis of the disease as they tend to be quite reliable as a result of the presence of many specific AE antigens. In this report we are presenting an advanced case with disseminated alveolar echinococcosis [5, 11, 7].

The only way to treat AE is surgical resection of the cysts combined with the treatment with (albendazole and or mebendazole) over 2 years after surgery. However, in inoperable cases, conservative treatment remains the only course and, in these cases, either mebendazole at a dose of 40-50 mg / kg / day in three doses or albendazole at a dose of 10-15 mg / kg / day in two doses can be used. Conservative alone does not guarantee complete elimination of the disease in infected patients, they are usually treated for long periods of time (many years and possibly a lifetime). Liver transplantation is seen as one of the last alternatives in the treatment of the disease although it seems quite dangerous, as it often results in reinfection of echinococcosis in patients [3, 4]. In this report we are presenting an advanced case with disseminated alveolar echinococcosis.

Clinical case

One 36-year-old women from Peshkopia has been admitted in the Gastrohepatology department on 20.07.2011 with fatigue, anorexia, dull pain in right hypochondrium, mild epigastric pain, bloating and weight loss. The epidemiological anamnesis showed that the patient lived in the village and had pets. On physical examination, the patient appeared severely ill with jaundice, massive hepatomegaly, massive mass in the mesogaster, and anxiety. Laboratory examinations were as follows: Hb 11.1 g/dl; sediment 25 mm/h; leukocytes 6700 / mm3, platelets 127000 / mm3, prothrombin level 60%; uremia 12.7 mmol/l, creatinine 0.78 mmol/l; ALP 127 U/l; AST 15 U/l; ALT 37 U/l; GGT 131 U/l; bilirubin 3.7 mg/l; albumin 2.8 gr/l; total protein 8.1 g/l; HbsAg negative; anti-HCV negative. Regarding serology, the titer of anti-echinococcal antibodies was positive (22, n = 11)

Examination of ascitic fluid: glucose 0 mg/dl, amylase 12U/l; cholesterol 212 mg/dl; triglycerides 26.6 mg/dl; total protein 6.9 gr/l, albumin 2.2 gr/l; 1400/mm3 cells dominated by neutrophils mainly eosinophils as well as lymphocytes.

Imaging examinations: Abdominal ultrasound: the enlarged liver, in the right lobe is observed a large formation 20 x 18 cm, with many cysts inside, surrounded by a membrane that goes in favor of the echinococcus. The structure of the rest of the liver appears to be preserved. Cholecyst and bile ducts were normal. Lien and pancreas without lesions. A mesogastric formation with cysts is observed in the mesogastric area. There was free fluid around the liver and in the hypogastric area.

Magnetic Resonance Imaging: showed diffuse peritoneal, cystic formations (largest 19x15 cm, located in the right lobe of the liver), with fluid content, with septa, slightly injected by KIV and a cystic formation 6 cm in diameter. Other small peritoneal, sub hepatic dexter and anterior abdominal formations. Liquid in the abdomen (Fig. 1, a, b, c).

Fig. 1.a. The coronary T2 sequence evidences a multicamera cystic formation that occupies the right lobe of the liver and other similar, perihepatic and peritoneal formations. Evidence of ascites.
After 10 years (14.05.2019), she did relapse and admitted to our clinic at age 46 with fatigue, abdominal distention, light abdominal pain, weight loss, and vomiting time by time.

In CT abdomen (09.01.2019): liver without dilatation of intrahepatic bile ducts. Postoperative changes in the dexter lobe with omentum deposition. Cystic lesion on the edge of the resection and other lesions in the dexter and sinister lobes are evident, aspects in favor of echinococcal cysts. Multiple peritoneal cystic lesions with echinococcal cyst aspects are evident.

CT abdomen (16.05.2019): liver post segmentectomy dexter. Multiple echinococcal cysts with dimensions 115 x7 0 mm are observed. Subhepatic cystic mass is observed which has a close relationship with the renal parenchyma and the diaphragm with dimensions 75x 50mm. Intrahepatic bile ducts are not dilated. Lien homogeneous. Sub-lienal echinococcal cysts with dimensions 72 x 53mm with thickened walls are noticed. Intraperitoneal multiple echinococcal cysts are observed the largest in the median line with dimensions 110 x 50mm. Anteriorly fundus uteri cystic formation with dimensions 106 x 43 mm is observed. No free fluid in the abdomen. Serological tests for echinococcus this time was negative: IgG 6.5. The situation was not so good for surgery but with her insisted, she did the Surgery.

The surgical intervention was performed where intra-abdominal cysts were resected, echinococcal cyst at the level of the gastric ligament with a diameter of 15x9cm, sub-lienal echinococcal cyst with a diameter of 9x8 cm, supra-lienal echinococcal cyst 7x6 cm, supra-hepatic echinococcal cyst 12 cm at the level of the celiac trunk 8x6 cm, subhepatic echinococcal cyst with a diameter of 12x11 cm, echinococcal cyst in the pelvis between the uterus and the bladder with a diameter of 17x15 cm, echinococcal cyst at the level of the sigmoid segment 11x12cm and multiple miliary cysts with a diameter <1 cm. She continuous to be under the treatment with Albendasol 400 mg on day, and is in a good condition until now, tow year after the second surgery.

Clinical presentation and radiology imaging of disseminated of alveolar echinococcosis, and diagnosis can be very difficult in atypically advanced cases. Patient went to surgery, where extensive laparotomy was performed and it was seen that there was diffuse spread of echinococcus in the peritoneal cavity and all cystic formations were resected. The biopsy was then performed which resulted in echinococcosis. After the surgery patient was treated with Albendazole 2 x 400 mg on day during 2 years, and she was normal for many years.

**Discussion**

Alveolar echinococcosis should be established as an unexplained chronic inflammatory tumor regardless of its localization. Some studies have suggested that AE may be linked to genetic factors but familiar cases are considered quite rare. Often this disease is asymptomatic for years, but depends from the site of location. The surgery is the only best way of treatment, but it can relaps after some years after the surgery [8]. Ultrasound monitoring should be recommended to patients’ relatives in order to detect small lesions to be treated surgically. Studies have shown that in addition to clinical and radiological findings, researchers must rely on serological tests to confirm the diagnosis and treat their patients without using invasive diagnostic
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In the process of treating these patients, the only way to address the lesions is to remove as much of the lesion as possible. This is typically done using a combination of surgery and chemotherapy. However, it has been shown that even with aggressive treatment, relapse can occur in up to 10% of cases. It is possible for the disease to recidivate even after long periods of apparent remission. This highlights the importance of ongoing surveillance and treatment for patients with a history of echinococcosis.

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