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Case Report

Young Age of Alveolar Echinococcosis Patient in Armenia: A Case Report

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Abstract

Human alveolar echinococcosis (AE) remains a serious public health concern in endemic areas and a challenge for clinicians. Here a confirmed case of human AE in a patient from Armenia who had not visited a known *Echinococcus multilocularis* endemic area is reported. In October 2012, a 12-year-old girl from a little Armenian village, presented with paroxysmal pain in the right lumbar area to the children's medical center (MC). The girl mentioned having close contact with an animal, like a cat. She was admitted to the surgical department with a diagnosis of a malignant liver tumor in the right lobe. In November 2012, the patient underwent laparotomy, removal of the hepatic lesion and abdominal cavity drainage. The histopathological examination of the biopsy material confirmed the main diagnosis of liver AE with suppurative lesions. The patient was given albendazole (ABZ) following 20 days in the hospital, but she stopped receiving the preventive chemotherapy at home and even missed the dispensary visits. It later caused complications, and in July 2016, the child had once again surgery. In January 2017, the child was readmitted to the MC with no content from the external biliary drainage tube in the previous 6 hours. Bile flow improved after flushing the drainage with saline solution and suturing the enterostomy tube. In February 2017, the child visited MC for examination, and the drainage of the bile ducts was blocked, although she had neither discomfort nor jaundice. It was recommended to continue the patient's follow-up, to receive ABZ and to undergo a liver transplant surgery.

Introduction

Human alveolar echinococcosis (AE) is a chronic zoonotic disease that is caused by the larval stage of *Echinococcus multilocularis*, a helminth of the family Taeniidae, Cestoda class. In the life cycle of the helminth, wild or domestic carnivores like foxes, domestic dogs, cats are definitive hosts and their prey, small mammals such as rodents, are intermediate hosts. Humans are accidental (aberrant) intermediate hosts and the dead end of the life cycle (1-3). Humans are infected through direct contact with the definitive host or indirectly through food or possibly water contaminated with parasitic eggs. *E. multilocularis* is mainly distributed in the temperate and cold regions of the northern hemisphere (3). Since 2000, a visible uptrend in AE incidence has been seen in historic endemic areas, such as China, France, Germany and Switzerland, as well as involving new geographic areas (1, 3-5).

The disease has a long asymptomatic incubation period, approximately 5-15 years, and is usually diagnosed accidentally in adults (between 50 – 70 years) and is seen rarely in children. AE has the characteristic of growing like a tumor and often invades other organs (1, 5). The WHO included AE as 1 of 20 neglected tropical diseases and *E. multilocularis* as the food-borne parasite with the third largest global impact of 24 ranked parasites (1).

AE is an emerging threat in Armenia. Since more than a decade ago, Armenia has experienced human AE cases. Active retrospective case finding was carried out at multi-profile medical centers equipped with modern diagnostic means. The medical records of patients with liver surgery were reviewed. Twelve confirmed AE cases were found between 2008 and 2022. The confirmation was based on tissue biopsy and medical imaging results (6). Meanwhile, in European endemic countries, the annual incidence ranges from 0.03 to 0.20/100,000 inhabitants/year (7).

This retrospective study reports a confirmed case of human AE in a patient from Armenia who had not visited a known *E. multilocularis* endemic area.

Case report

Informed Consent

Written informed consent was obtained from the patient's mother, who is the child's only guardian.

In October 2012, a 12-year-old girl from a little Armenian village located in the picturesque valley of Gegharkunik region, near Lake Sevan, presented with paroxysmal pain in the right lumbar area to the children's medical center (MC). Over the past 2 years, the child sometimes reported brief abdominal pains. The girl mentioned having close contact with an animal, like a cat.

The results of all laboratory tests, including the complete blood count (CBC), biochemical blood tests were normal, C-reactive protein was negative except for an erythrocyte sedimentation rate test (ESR) (32 mm/hour; in 12 years and older children-3-15 mm/hour). Both the hepatitis B antigen and rapid plasma reagin (RPR) syphilis tests came out negative. There were no pathological deviations on the chest X-ray. Ultrasonography (US) and abdominal computer tomography (CT) both demonstrated a moderate hepatomegaly, 78x82 mm hepatic foci in the projection of the V, VI, and VII segments with a solid structure in the periphery and 30x32 mm fluid content in the middle. The gallbladder was enlarged, the walls were smooth, the intrahepatic and extrahepatic bile ducts were not dilated, the lymph nodes in the porta hepatis were enlarged. She was admitted to the surgical department with a diagnosis of a malignant liver tumor in the right lobe. In November 2012, the patient underwent laparotomy, removal of the hepatic lesion and abdominal cavity drain-

age. The histopathological examination of the biopsy material from the affected right lobe of the liver confirmed the main diagnosis of liver AE with suppurative lesions.

During the second postoperative US, there was a tiny amount of free fluid in the pelvis, as well as a 3.6x2.6 cm hypoechoic fluid in the projection of the V and VI segments, close to the drainage. The patient was given albendazole (ABZ) following 20 days in the hospital, but she stopped receiving the preventive chemotherapy at home and even missed the dispensary visits.

In July 2016, the child once again was admitted to the MC with complaints of skin and visible mucous membrane jaundice, nausea, dizziness, itching, the presence of acholic stools, which had appeared about 2 weeks ago. The tongue was dry and coated, liver palpation was painful. US examination revealed a diffusely inhomogeneous structure of the liver parenchyma, 100x95 mm, round, uneven edged mass is visualized in the VI, VII, VIII segments of the right lobe. Cholestasis and bile duct dilatation were seen, particularly in the porta hepatis and hepatic portal vein zones (Fig. 1).

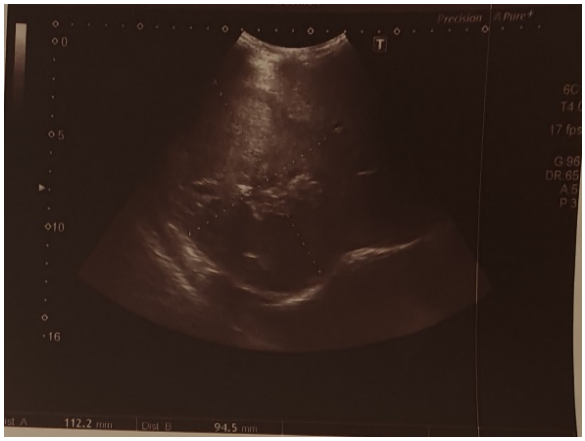

Fig. 1: Liver ultrasound examination

Preoperative diagnostics included a CBC, biochemical blood tests, a coagulation test, and a urinalysis. The patient had successful surgery (cholecystectomy (gallbladder removal surgery), laparotomy, bile duct revision, intraoperative cholangiography, external biliary drainage, jejunostomy with an enterostomy for nutrition, sanitation and drainage of the abdominal cavity). Under a microscope, a cytological examination of the surgical material (a piece of gallbladder- affected lesion) was performed, and erythrocytes, cells of inflammatory infiltrate-leukocytes (lymphocytes, neutrophils, and eosinophils) were discovered after staining with hematoxylin and eosin. The epithelial cells remained unchanged (within the normal range). The post-surgery period was smooth, she was given Amoxiclav, infusion therapy, ABZ. There was no cholestasis, and a tiny quantity of fluid was detected in the Douglas pouch by the US. It was recommended to remain under control in the surgical dispensary of the MC and local pediatrician, ABZ for 6 months (400 mg 2 times per day), CBC and biochemical blood tests once a month were prescribed and to visit a doctor in case of drainage disorder.

In January 2017, the child was readmitted to the MC with acute abdominal pain, vomiting, 36.3 C° temperature, and no content from the external biliary drainage tube in the previous 6 hours. Abdomen palpation was very painful in the epigastric and right lumbar areas. The liver was solid, painful and palpable under the rib. The tongue was dry; the skin was without pathological lesions, normal stool color, urina-

tion was painless. This time, a subtotal alveococcus lesion of the right lobe of the liver, mechanical jaundice was diagnosed, a condition after external drainage of intrahepatic bile ducts. Bile flow improved after flushing the drainage with saline solution and suturing the enterostomy tube. The pain gradually decreased and it was recommended to continue the conservative treatment. She spent the night quietly, there was no temperature, and her hemodynamic indications were stable. The bowels functioned in the evening. Palpating the abdomen in the right lower region was unpleasant. The drainage produced around 45-50 ml of greenish yellow bile. A CT scan revealed a round, cystic, 10.3x8.9x10.5 cm lesion with a solid component in the V, VI, VII segments of the liver right lobe, with air bubbles (a sign of infection) and multiple *dotted* calcifications. The left lobe of the liver is significantly enlarged yet unaffected. There was just a little fluid in the pelvic cavity. There were no enlarged affected lymph nodes. Other organ-systems were without visible pathological lesions. After 15 days, the child was discharged from the hospital for further observation.

In February 2017, the child visited MC for examination, and it was discovered that the drainage of the bile ducts was blocked, although she had neither discomfort nor jaundice. A biliary US examination revealed no signs of cholestasis. It was recommended to continue the patient's follow-up, to receive ABZ and to undergo a liver transplant surgery.

Discussion

AE remains a serious public health concern in endemic areas and a challenge for clinicians. The young age of several AE cases seems like alarming and could indicate a wide spread of infection in the environment or the important role of domestic animals, like dogs and cats, with risk exposure occurring already in early childhood (6). A case in the same age group was also reported in the Slovak Republic, Canada, Kyrgyzstan (7-9). However, a majority of AE cases are diagnosed in older age groups (2, 7, 8, 10-13).

It is important to treat this serious infection properly. The fatality rate in untreated or inadequately treated persons is above 90% (1, 3, 5). Almost 98% of AE cases are located in the liver and further spread into other organs (13). After a radical or sparing surgical treatment, the patient requires preferably two-year albendazole administration and appropriate follow-up (2, 10, 14). According to the WHO, the disease is frequently discovered at an advanced stage in many patients. Surgical treatment without or with incomplete anthelmintic drugs often leads to relapses. The study highlights the usefulness of the biopsy diagnosis in countries where serological tests are not available, like Armenia.

In many endemic countries, the total number of AE cases is considered underestimated (3, 10). The majority of clinicians consider cystic echinococcosis to be the only type of human *Echinococcus* spp. in Armenia. Even experienced clinicians may face difficulties in the differential diagnosis due to their lack of knowledge about this illness and a sizable number of cases are likely to remain misdiagnosed. AE can easily be misdiagnosed as cirrhosis or liver cancer (4).

Within the framework of the "One Health" concept, there is a need to implement surveillance and prospective research for *Echinococcus* spp., both in humans and animals. The prospective registry of all AE cases should be a priority. A differential diagnosis of AE should be considered for patients with tumor-like lesions of the liver. Awareness campaigns among clinicians as well as among the general population potentially at risk should be implemented, to enhance earlier diagnosis, treatment and preventive measures. The risk of infection in humans should be decreased by the routine deworming of domestic animals.

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Conflicts of interest

The author declares no conflicts of interest.

References

1. Baumann S, Shi R, Liu W, et al. Worldwide literature on epidemiology of human alveolar echinococcosis: a systematic review of research published in the twenty-first century. Infection. 2019; 47(5):703-727. doi:10.1007/s15010-019-01325-2.
2. Dezsényi B, Dubóczki Z, Strausz T, et al. Emerging human alveolar echinococcosis in Hungary (2003–2018): a retrospective case series analysis from a multi-centre study. BMC Infect Dis. 2021;21(1):168. doi:10.1186/s12879-021-05859-5.
3. Torgerson PR, Keller K, Magnotta M, Ragland N. The Global Burden of Alveolar Echinococcosis. Brooker S, ed. PLoS Negl Trop Dis. 2010;4(6):e722. doi:10.1371/journal.pntd.0000722.
4. Vuitton DA, Demonmerot F, Knapp J, et al. Clinical epidemiology of human AE in Europe. Vet Parasitol. 2015;213(3-4):110-120. doi:10.1016/j.vetpar.2015.07.036.
5. Jonaitytė E, Judickas M, Tamulevičienė E, Šeškutė M. Alveolar Echinococcosis in Children. Case Rep Pediatr. 2020 ;2020:5101234. doi: 10.1155/2020/5101234.
6. Manukyan A, Avetisyan L, Sahakyan G, et al. A review of human alveolar echinococcosis in the Republic of Armenia from 2008 to 2020. Parasite Epidemiol Control. 2022;17:e00246. doi:10.1016/j.parepi.2022.e00246.
7. Houston S, Belga S, Buttenschoen K, et al. Epidemiological and Clinical Characteristics of Alveolar Echinococcosis: An Emerging Infectious Disease in Alberta, Canada. Am J Trop Med Hyg. 2021;104(5):1863-1869. DOI: 10.4269/ajtmh.20-1577. PMID: 33755579; PMCID: PMC8103444.
8. Kinčeková J, Hrčková G, Bober J, et al. A rare case of alveolar echinococcosis in a 14-year-old child. Helminthologia. 2008;45(1):28-31. doi:10.2478/s11687-008-0005-4.
9. Paternoster G, Boo G, Wang C, et al. Epidemic cystic and alveolar echinococcosis in Kyrgyzstan: an analysis of national surveillance data. Lancet Glob Health. 2020; 8(4):e603-e611. doi:10.1016/s2214-109x(20)30038-3.
10. Cambier A, Leonard P, Losson B, et al. Alveolar echinococcosis in southern Belgium: retrospective experience of a tertiary center. Eur J Clin Microbiol Infect Dis. 2018 ;37(6):1195-1196. doi: 10.1007/s10096-018-3233-7.
11. Delbecque K, Detry O, Hayette MP, et al. A case of hepatic alveolar echinococcosis contracted in Belgium. Acta Gastroenterol Belg. 2002 ;65(1):55-60. PMID: 12014318.
12. Dezsényi B, Strausz T, Makrai Z, et al. Autochthonous human alveolar echinococcosis in a Hungarian patient. Infection. 2017 ;45(1):107-110. doi: 10.1007/s15010-016-0918-7.
13. Yamamoto Y, Sakamoto Y, Kamiyama T, et al. A case of alveolar echinococcosis in the liver that ruptured into the pericardium treated by a combination of hepatectomy and albendazole. Surg Case Rep. 2022; 8(1):63. doi:10.1186/s40792-022-01417-6.
14. Pielok Ł, Karczewski M, Cierach W, Zmora P, Lenartowicz E, Stefaniak J. Portal hypertension as a result of the incomplete surgically treated advanced alveolar echinococcosis: a case description. BMC Gastroenterol. 2020;20(1):176. doi:10.1186/s12876-020-01320-0.