Alveolar echinococcosis: clinical case presentation

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ABSTRACT
Echinococcosis is a parasitic disease caused by Echinococcus species. Tapeworm Echinococcus multilocularis causes alveolar echinococcosis (AE) which is a significant worldwide public health problem with potential life-threatening outcomes. We present a case report of advanced AE with lesions in liver and dissemination to vena cava inferior and lungs.

Keywords: echinococcus multilocularis, Alveolar echinococcosis, Malignant parasite, Mimicking cancer.

INTRODUCTION
Echinococcosis is a zoonosis caused by a tapeworm species belonging to the genus Echinococcus, family Taeniidae (1, 2). In humans, E. granulosus occurs as cystic echinococcosis, E. multilocularis causes alveolar echinococcosis (AE), and E. vogeli and E. oligarthrus as polycystic echinococcosis (3). E. multilocularis tapeworm is endemic in many regions of the world, but parts of the Northern hemisphere, such as North America, China, central and eastern Europe and the Baltic states, are the most concerning (4, 5, 6). In Lithuania alone between 1997 and 2013, a total of 179 AE cases have been reported (7). Diagnosing this disease is complicated due to extended incubation time, various clinical manifestations and mimicking of differential diagnoses (8). We report a case of advanced AE, which is a rare infection and may even have fatal consequences. Our purpose is to raise awareness of diagnosing this condition among various physicians.

CASE REPORT
A 59-year-old woman referred by a general practitioner to the pulmonology department presented to our hospital with complaints of coughing up phlegm and blood, and weight loss. During a preventative health examination, a front chest X-ray was performed, and it revealed diffuse 13-17 mm diameter high-density formations on both sides in the lungs. The patient was referred to a pulmonologist for a further examination expecting a malignant process. Second X-ray scan was performed showing multiple high-density various sizes with the largest measuring approximately 15 mm mass lesions with clear boundaries, mostly in the periphery of the lungs (Figure – 1). Radiological findings are to be differentiated with primary cancer and metastasis.

The patient claimed to be allergic to iodine and therefore, was hospitalized for computed tomography (CT) scan. On the day of admission, her vitals were normal, vesicular breathing and no crackles were present. A consensus was made to perform a CT scan for suspected lung tumour and spreading during a pulmonologists Concilium. The patient was treated with dexamethasone and clemastine according to the protocol against iodine allergy.

Contrast-enhanced chest and abdominal CT revealed multiple polisegmental heterogenic-density various size mass lesions with a polyclide outline situated along the vessels mostly in the periphery of both lungs, some with mild enhancement. The most prominent lesion is measuring approximately 16 x 11 mm in the right first lung segment (Figure – 2, 3). CT also showed a heterogenic mild enhancement lesion with a polyclide outline, cystic components and calci-
fications measuring approximately 12 x 82 x 78 mm, in the left lobe of the liver (Figure - 4). Vena cava inferior (up to right atrium level) and the vault of the diaphragm were also infiltrated.

A few merging similar structures pathologic nodes measuring approximately 28 x 15 mm and nodules situated along the oesophagus with the largest measuring approximately 12 x 12 mm were present between the left lobe of the liver and the lesser curvature of stomach within the left heart ventricle. With these findings, hepatic echinococcosis with invading to vena cava inferior and hematogenous lung dissemination were suspected radiologically.

Initial laboratory tests showed haemoglobin level 147 g/l, white blood cells count 12,3 x 10^9/l, eosinophils 0,0 x 10^9/l, platelet count 268 x 10^9/l. C – reactive protein, creatinine, urea, alanine and aspartate aminotransferase, and electrolytes were normal. In addition, the patient was positive for the presence of IgG antibodies against Echinococcus spp. 74,2 g/l.

Based on the clinical symptoms, imaging examinations, and serology test, Echinococcus multilocularis infection was diagnosed. Albendazole treatment in 400 mg two daily doses was started, and the patient was referred to an infectologist for an immediate consultation.
DISCUSSION

AE caused by E. multilocularis tapeworm is a worldwide endemic concern. (4). Over the past few decades in Europe emergence of AE in humans was reported in France (509; diagnosed between 1982 and 2011), Switzerland (494; 1956-2005), Lithuania (179; 1997-2013), Germany (114; 2003-2013), Poland (121; 1990-2011), Latvia (43; 1996-2010) Slovakia (26; 2000-2013) and Czech Republic (20; 1998-2014) (7, 9, 10). Moreover, countries previously documented to be free from E. multilocularis infection, including, the UK, Ireland, Malta, Norway and Finland take actions to measure the risk of introducing this parasite into their areas through imported pets and wildlife animals (5).

The definite hosts are wild carnivores – mainly red fox, whereas rodents are intermediate hosts in the life cycle of E. multilocularis (2, 3). Cats and domestic dogs may be involved in a synanthropic cycle of adult tapeworms (2). The eggs with oncospheres produced by the adult parasite living in the small intestine of the definitive hosts are released into the environment with faeces, and the cycle continues with the digestion of contaminated food by the intermediate host (2, 3). Humans are accidental dead-end hosts acquiring the infection through the faecal-oral route (10).

AE is a chronically progressive infection with a high degree of disability, morbidity and mortality in the absence of curative surgery and anti-helminthic treatment (4, 8, 11). The incubation period may take 5 to 15 years without any significant symptoms leading detection by chance or during screening programmes (12). Our patient was also referred to as seek a pulmonologist for further inspection because of suspected metastatic lung lesions after a screening programme. (4, 8, 11).

Although AE is benign, it presents a cancer-like appearance in the liver with destructive tissue growth and metastatic spread in lymph nodes, lung, brain, bone, spleen and other organs through blood vessels and lymphatic vessels (13, 14, 15). Finally, in the advanced stages, a bacterial liver abscess may be mimicked by super-infection of central necrotic cavities (17) which makes the diagnostics even more complicated.

Diagnosis of AE is based on the following aspects: clinical findings and epidemiological data, imaging studies, and histopathology, and serology (11, 12).

Gathering information about patient's history, living space and contact of farm animals that is the source of infection, clinical symptoms such as abdominal pain or discomfort, a mass/masses in hepatic region, jaundice, high fever or diziness, headache, coughing (15) is the first step when suspecting echinococcosis.

Imaging studies for AE include ultrasonography (USG), CT and magnetic resonance imaging (MRI) (13). USG is useful for a screening diagnosis as it shows firm echo lesions with heterogeneous echoes inside when examining the liver (13, 15). No envelope is present around the lesion of hepatic AE, and the margin is irregular so it can be misdiagnosed as primary liver cancer, hepatic hemangioma and focal hyperplasia (15). For further examination after USG, CT and MRI are used (13). CT imaging shows an inhomogeneous solid mass, unclear boundary and no noticeable enhancement in the enhanced scan so lesions may be vesicles, calcification, and liquefaction necrosis. In contrast, on contrast-enhanced CT, the periphery of the mass may be enhanced, and multicentric vesicles may be visible (15). MRI supports the diagnosis by showing multi-vesicular morphology with a clear boundary after enhancement. (13, 15).

The most critical imaging feature of AE is calcification. There should be clusters of microcalcifi-
cations or plaque-like calcific foci with an irregular distribution in any part of the lesion (13). During a histopathologic examination of E. multilocularis the parasitic vesicles have a Periodic-Acid-Schiff (PAS)+ laminated layer. Epithelioid cells, macrophages, fibroblasts, giant multinucleated cells and various cells of nonspecific immune response, usually surrounded by lymphocytes are the composition of periparasitic granuloma (1).

The antigen detection assay with a high diagnostic sensitivity of 90–100% and a specificity of 95–100% is helpful when diagnosing AE (1, 13). The principles of treating AE are to stop the invasion of the lesion into surrounding organs and to prevent dissemination (13). Radical surgical resection followed by benzimidazoles as anti-infec-tive drugs for two years to avoid recurrence is the first-choice treatment, taking into account the patient-specific stage of the disease. (12, 13, 18, 19). Unfortunately, in advanced AE stages, radical surgery is not feasible (20). Our patient had E. multilocularis infiltrations in vena cava inferior, making radical surgery hazardous due to the risks of air embolism or uncontrolled hemorrhage (21).

Inoperable patients must undergo long-term treatment with benzimidazoles, mostly albendazole (ABZ) or alternatively mebendazole (MBZ) when ABZ is not tolerated (18, 21, 22). Recommended dosages of ABZ are 10–15 mg/kg/day, in 2 doses, recommended with a high-fat meal to ensure absorption (18, 20). Alternatively, MBZ is given at a dose of 40–50 mg/kg/day, divided into three doses (20).

All in all, the most crucial factor in improving prognosis of AE is supposed to be early diagnosis following with complete resection (19).

CONCLUSION

Our case report is a reminder for doctors that echinococcosis may present as cancer-like lesions both in clinical manifestation and radiological imaging. It is noteworthy to involve this zoonosis in the differential diagnosis when suspecting cancer and metastases in various organs.
REFERENCES


