Alveolar Echinococcosis

Clinical course

After infection with E. multilocularis, there is always an asymptomatic incubation period of 5e15 years, except in immunosuppressed patients. Infected individuals typically develop a silently progressing hepatic disease that clinically behaves like a tumor with infiltrative growth and the potential for metastasis.

Clinical Presentation

Initial clinical symptoms can be abdominal pain (mostly epigastric or right upper quadrant) or cholestasis with or without jaundice. In at least one-third of the cases, AE is diagnosed incidentally during a medical workup for abnormal laboratory tests or symptoms and signs. AE symptoms and prognosis are primarily dependent on location and secondarily on the size of the lesion. Dependent on the location of the lesions, not on different morphologic features, the Alveolar Echinococcis is classified in PNM-stages I-IV.

Diagnostic tools/ Confirmation of diagnosis

A diagnosis of AE is based on clinical findings, epidemiological data, imaging techniques (Sonography, MRI, CT/PET CT), histopathology and/or nucleic acid detection, and serology (“national reference center” UZH Parasitology department).

Therapy options

Therapeutic management of patients with Alveolar Echinococcosis (AE) clearly requires a multidisciplinary approach, in which drug-therapy with BMZ (Albendazole, Mebendazole) is the backbone of treatment for all patients. A complete evaluation of the disease process (including thoracic and brain CT) is necessary before any therapeutic decision and to serve as a basis for follow-up. Duration of therapy ranges from 2 years in radical surgery to lifelong therapy in persistent active Echinococcosis.

Therapy should be performed in an expert center. AE can be cured by radical surgery if detected at an early stage (in Europe between 20-50% of patients). Nonsurgical interventions should be pursued in patients with lesions that cannot be completely resected but need palliative intervention. Liver transplantation remains a rescue measure in selected cases. All therapy-options for AE are available in the University Hospital Zurich and are discussed at the interdisciplinary weekly tumor board.

Drug Therapy

For most patients 2x200 mg Albendazole (Zentel) daily is an appropriate starting dose. The dose of second choice medication Mebendazole (Vermox) is 40-50 mg/kg/day split into three doses. All Benzimidazoles are given with fat-rich meals.
Monitoring of blood drug levels is indicated to confirm adherence to treatment, to ensure adequate therapeutic drug levels and to avoid toxic reactions. BMZs are embryotoxic and teratogenic. Therefore, the use of these drugs should be avoided in pregnancy, and contraceptive measures are mandatory for women of reproductive age.

Prognosis

The prognosis depends on making the correct diagnosis, the stage of disease and treatment options. With adequate therapy, the prognosis is good. If left untreated alveolar echinococcosis is progressive and fatal.

Follow Up

Follow up in alveolar echinococcosis is recommended long-term (in most cases lifelong) by imaging and serology with intervals of 6 months in the first years after diagnosis.

Reference: Echinococcus and Echinococcosis, Part B, First Edition, 2017, 259e369)