Vascular pathology in patients with alveolar echinococcosis: framework for assessment and clinical management – a retrospective case series

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ABSTRACT

Objective Alveolar echinococcosis (AE) is a parasitic liver disease with infiltrative growth similar to solid organ malignancies. Major vascular damage is frequent and often remains untreated until catastrophic events precipitate. Detailed clinical and radiological assessment is required to guide individualised treatment decisions. Standardised radiological reporting templates of malignancies with profiles resembling AE are candidates for adaptation. Our objectives are to describe vascular pathology in AE and establish a framework for structured evaluation as the basis for treatment decisions and monitoring.

Design Retrospective case series.

Results 69 patients (37.1%) had vascular involvement: portal vein (PV) 24.7%, hepatic vein (HV) 22.6% inferior vena cava (IVC) 13.4%. Significant stenosis/occlusion of vessels was present in 15.1% of PV, in 13.4% of HV and in 7.5% of IVC involvement. Vascular pathology needing specific treatment or monitoring was present in 8.6% of patients. The most frequent clinical presentation was high grade IVC stenosis or occlusion which was seen in 11 patients of the cohort.

Conclusion Advanced AE requires early multidisciplinary assessment to prevent progressive impairment of liver function due to vascular damage. The focus at first presentation is on complete evaluation of vascular (and biliary) involvement. The focus in non-resectable AE is on prevention of vascular (and biliary) complications while suppressing growth of AE lesions by benzimidazole treatment to improve the quality of life of patients. We developed a framework for standardised vascular assessment and follow-up of patients with AE to recognise and treat complications early.

INTRODUCTION

Alveolar echinococcosis (AE) is a rare liver disease with infiltrative intrahepatic growth, regional progression beyond the liver and distant metastases. AE exhibits all features of solid organ malignancies and is referred to as 'malignant parasitosis'. Consequently, the Parasitic mass in the liver, Neighbouring organs, Metastases (PNM) classification was suggested in 2006.¹ Detailed clinical and radiological assessment is required to stage patients with AE at presentation and during follow-up to guide individualised treatment decisions in a disease with highly heterogeneous manifestations. The standardised radiological reporting templates developed for various common malignancies with profiles resembling AE—infiltration, stenosis and occlusion of major hepatic vessels—are candidates for adaptation.²⁻⁴

Whereas biliary tree involvement is early recognised by raised cholestasis parameters and in more advanced disease by jaundice,
vascular damage often remains unnoticed until catastrophic events precipitate.

Our objectives are to describe vascular pathology in AE and to establish a framework for structured evaluation as the basis for treatment decisions and monitoring.

MATERIALS AND METHODS

Design
Retrospective case series.

Objectives
► Describing the vascular baseline and follow-up assessments (ultrasound/Doppler/MRI/CT) and the vascular pathology in a large AE patient cohort.
► Developing a framework for identifying, monitoring and treating vascular complications.

Clinical setting and study population
The Division of Infectious Diseases and Tropical Medicine at UKHD runs a clinic for echinococcosis since more than 20 years. Our unit is a national and international clinical reference centre for echinococcosis.

In this retrospective case series of patients treated based on long centre-based experience with the multidisciplinary care for patients with AE by a team which is stable since more than a decade.

We reviewed the database of our prospective cohort and extracted the patients fulfilling the following inclusion criteria:
► Patients with primary presentation at our clinic between April 2000 and October 2021, AE confirmation and at least one follow-up presentation.
► Subdivided into
  - Group A: no involvement of vascular system.
  - Group B: vascular involvement consisting of contact to or encasement of vessels without significant stenosis.
  - Group C: vascular involvement consisting of contact to or encasement of vessels with significant stenosis or occlusion including macroscopic vascular invasion.

In groups B and C, vascular involvement of the portal vein (PV), the hepatic veins (HV), inferior vena cava (IVC) and the hepatic arteries (HA) was identified from radiological MR/CT reports at primary presentation. Significant stenosis was generally considered to be present when a vessel diameter reduction of at least approximately 70% (stenosis with haemodynamic impact) was observed on imaging studies according to the reporting radiologist.

Clinical data extraction
We extracted
► Demographic variables: gender, age at initial diagnosis, country of origin, profession.
► General clinical variables: duration of treatment, calculated as the time between the first presentation in our clinic and the last follow-up visit; relevant previous illnesses such as malignancies or autoimmune diseases; PNM classification; therapeutic modality (surgery curative/palliative); benzimidazole treatment (albendazole (ABZ)/mebendazole (MBZ)/other); duration of treatment; treatment-related complications and involvement of vessels.
► Vascular-specific variables: signs and symptoms and imaging related to vascular involvement and interventional treatment of vascular pathology.
► Vascular region affected (PV, HV, IVC, HA).
► Follow-up modality (CT/MRI, angiography, colour-coded duplex sonography, oesophagogastroduodenoscopy).
► Vascular thrombosis.
► Anticoagulation.
► Bleeding under anticoagulation.

Data were extracted from paper and electronic patient files—database system SAP IS-H and entered into a REDCap database.

Data analysis
► Description of vascular involvement in our AE patient cohort, including mapping of the course of disease (clinically and/or radiologically evident vascular complications) and interventions across time (patient biographies).
► Analysis of the vascular baseline and follow-up examinations (ultrasound/Doppler/MRI/CT) in relation to vascular complications.

RESULTS
A total of 186 patients with AE were included in the study. A total of 136 were on active follow-up at the time of evaluation. Of 50 patients not under follow-up anymore, 5 had R0 resection and completed 10 years of follow-up, 16 received treatment in another clinic, 10 patients had died and 20 patients were lost to follow-up. Mean duration of follow-up at the time of evaluation is 3.7 years (range 1 month to 15.4 years). The mean age of patients at diagnosis was 52.3 years (range 13–85 years), with a gender distribution of 112 (60.2%) female and 74 (39.8%) male patients.

Overall, 42 (22.6%) patients had underlying illnesses. Twenty-one patients (11.3%) had a history of cancer, of which 8 patients (4.3%) were under chemotherapy when AE was diagnosed. Thirteen patients (7.0%) had an autoimmune disease with 9 (4.8%) receiving immunosuppressive treatment. Five patients (2.7%) were diagnosed with a chronic inflammatory disease, one patient was diagnosed with HIV and one had liver transplantation before being diagnosed with AE.

Staging identified 57 patients (30.6%) with extrapathic growth of AE into neighbouring organs. Distant metastases and/or extrahepatic manifestations were present in 16 patients (8.6%) with 14 cases of (7.5%) lung metastases. Other organs involved were bone (4), pectoral muscle (1) and breast (1) and brain (1).

A total of 73 patients (39.2%) were treated by surgery with 41 (22.0%) R0 resections (histopathology). A total
of 113 patients (60.8%) were exclusively treated with benzimidazole. Forty-eight patients (25.8%) had to pause ABZ at least once due to drug toxicity. Nine patients died during the period of observation.

Over all 69 patients (37.1%) fulfilled radiological criteria for vascular involvement. PV was affected in 24.7% (15), HV in 22.6% (42), IVC in 13.4% (25) and HA in 5.9% (11).

Significant stenosis or occlusion of vessels was present in 15.1% (28) of PV involvement, in 13.4% (25) of HV involvement and in 7.5% (15) of IVC involvement. Infiltration and stenosis of the HA was seen in 1.6% (3) of patients.

Vascular involvement needing specific treatment and/or close monitoring (clinically manifest vascular involvement) was present in 8.6% (16) of patients. Eleven patients showed clinically manifest high grade IVC stenosis or IVC occlusion mainly manifesting with dyspnœa on exertion and significantly reduced physical fitness, leg oedema and/or newly occurred varicosities of the lower extremities, collateral vascular circulation of the abdominal wall, ascites. Nine of these patients received prophylactic anticoagulation. Four patients anticoagulated presented various severity bleeding (recurrent epistaxis, gingival, lower limb varicose or gastrointestinal bleeding). Three patients with IVC stenosis were treated with percutaneous transluminal angioplasty and stent insertion (not possible in one patient). One patient underwent surgery with resection of the IVC.

All patients with HV involvement had concomitant IVC or PV involvement. One patient who showed a subtotal stenosis of a single remaining HV after surgery had a history of deep vein thrombosis and Factor-V-Leiden mutation. The patient was started on prophylactic anticoagulation.

Three patients with advanced PV involvement had oesophageal varices. All three patients received prophylactic anticoagulation. One patient had oesophageal variceal bleeding. Two patients received prophylactic or therapeutic endoscopic band ligation of oesophageal varices.

One patient showed extensive involvement of the arterial system (Aorta, HA, coeliac artery, mesenteric arteries) with encasement of the vessels and radiological signs of vascular wall infiltration. The case was discussed in the interdisciplinary team and aspirin 100 mg was added to ABZ treatment, no interventional treatment was considered necessary. Out of 10 overall deaths during the period of observation, 4 occurred in the group of patients with clinically manifest vascular involvement.

Exemplary case reports

Summary case 1 ‘significant PV stenosis’
► A 29-year-old male patient.

Complications
► Cholestasis and cholangitis.

► PV occlusion.
► Oesophageal varices grade III.
► Acute variceal bleeding November 2020.

Clinical management
► Endoscopic retrograde cholango-pancreatography (ERCP), stenting and drainage of biliary duct.
► Albendazole 2×400 mg.
► Non-selective-beta-blocker (NSBB), carvedilol.
► Endoscopic band ligation of varices.
► Antibiotic prophylaxis for recurrent cholangitis (amoxicillin/clavulanic acid).
► direct acting oral anticoagulant (DOAC), apixaban 2×2.5 mg.

Discussion case 1
This patient first presented with jaundice and cholangitis and was diagnosed having AE. Due to recurrent cholangitis, long-term antibiotic prophylaxis was installed without further episodes of cholangitis. Furthermore, the patient had chronic, non-cirrhotic high-grade PV stenosis due to infiltration by AE with portal hypertension (figure 1). Portal hypertension was complicated by oesophageal varices and variceal bleeding. Secondary prophylaxis with a combination of NSBB and endoscopic band ligation. Recommended follow-up intervals after successful treatment of varices were 3 months and, thereafter, 6 monthly. No progression of AE under long-term ABZ treatment was observed. After weighing risks and benefits we decided in favour of long-term prophylactic anticoagulation with apixaban.

Summary case 2 ‘significant stenosis of single HV’
► A 30-year-old female patient.
► Diagnosis AE September 2004, P3, N1 (diaphragm), M0.

Complications
► Non-curative liver surgery.
  ‒ Right hemihepatectomy September 2004.
► Intervention used in cystic echinococcosis (CE).
► Residual AE hepatic lesion encasing single remaining left HV.
► Growth of AE lesion.
► Cholestasis.
► Factor-V-Leiden mutation.

Clinical management
► Albendazole 2×400 mg → dose doubled 2×800 mg due to disease progression (growth of lesion under treatment).
► Albendazole-sulfoxide (ASOX) intensive drug level monitoring.
► DOAC, rivaroxaban after DVT maintained as long-term prophylactic anticoagulation.
► ERCP+stent for cholestasis.
Discussion case 2

In this patient, AE was misinterpreted as CE. Differential diagnostic difficulties in distinguishing AE from CE have been described. The initial misdiagnosis explains the non-curative R2 surgical resection and PAIR attempt prior to the referral to our centre. The latter is reserved for CE and must not be employed in AE. Additionally, there was disease progression under ABZ treatment. ASOX levels were within the therapeutic range but the patient admitted that she had not taken ABZ regularly. Lack of compliance and substantial weight-gain may have contributed to failing AE growth suppression. We, therefore, increased the dose of ABZ to 800 mg two times per day on June 2014.

From the perspective of hepatic vasculature, the single remaining left HV and IVC are circularly encased by AE (figure 2) with significant stenosis. Additionally, the patient has Factor-V-Leiden mutation and a high risk of progression of stenosis and/or development of thrombosis (figure 2). In order to prevent further vascular pathology and preserve technical transplantability with respect to orthotopic liver transplantation (OLT) the patient was started on long term prophylactic anticoagulation with low dose DOAC. The optimal point in time for OLT is controversially discussed. The high recurrence rate of AE after OLT must be considered. Life-threatening developments in progressing AE disease marked by a succession of obstructive jaundice refractory to interventions, recurrent cholangitis, liver abscesses, recurrent bleeding caused by portal hypertension and chronic Budd-Chiari syndrome, are indications for OLT. This patient was listed for OLT after she developed obstructive jaundice.

Summary case 3 ‘asymptomatic significant IVC stenosis’

- A 39-year-old female patient.
- Diagnosis AE July 2017, stage P3 N0 M0.

Complications

- Occlusion of DHC with recurrent cholestasis and cholangitis.
- Liver abscesses.
- Albendazole toxicity with myelosuppression and alopecia.
- Extensive intrahepatic IVC walling with stenosis.

Clinical management

- Repeated ERCP with biliary balloon dilation and stenting.
- Cholangioscopic drainage of liver abscess.
With MBZ at a reduced dose with 2×500 mg, we managed to maintain antiparasitic treatment. In view of the relatively young age of the patient and the degree of complications, a surgical opinion was sought. The AE lesion is resectable but extensive surgery including resection/reconstruction of PV and IVC would be required. The patient sought several surgical opinions and felt that the risk of surgical treatment was too high.

Summary case 4 ‘IVC occlusion’
► A 69-year-old male patient.

Complications
► Drainage of hepatic necrotic cavity which was mistaken for malignant tumour with histopathological diagnosis of AE.
► Occlusion of the hepatic IVC segment with ascites and peripheral oedema at primary presentation.
► Albendazole toxicity with repeated myelosuppression and alopecia.

Clinical management
► Albendazole at reduced dose 2×200 mg with ABZ drug levels in therapeutic range
► Low-dose anticoagulation with DOAC, rivaroxaban 2×2.5 mg since March 2020.

Discussion case 4
Before the diagnosis of AE was made the patient experienced a period of 18 months with deteriorating general condition, fatigue, dyspnoea and weight loss of 12 kg. He attributed the complaints to depression due to the recent death of his spouse. In the further course, he developed oedema of the lower extremities up to the pelvic region and workup of the patient revealed a necrotic mass in the liver with ascites and occlusion of the IVC (figure 4). Malignancy was suspected, a liver biopsy and drainage of the necrotic cavity was done, histopathology showed AE. After starting diuretic and ABZ treatment peripheral oedema and ascites receded steadily over the initial 2 years of treatment. At the start of ABZ treatment, the patient experienced toxicity with myelosuppression (pancytopenia) and complete alopecia. ABZ at half the usual dose (2×200 mg) is well tolerated by the patient and shows sufficient ASOX levels. The clinical situation and radiological imaging have been stable over a period of 4 years with, however, reduced physical/cardiovascular fitness.

We started the patient on prophylactic anticoagulation with rivaroxaban due to the risk of Budd-Chiari-Syndrome. As the patient had no clinical signs of portal hypertension with normal platelet counts so far, no oesophageal varices screening was recommended.

**DISCUSSION**
Classification systems of AE are largely descriptive and lack focus and depth on the critical biliary and blood vessels. The PNM classification stratifies P and addresses...
critical areas determining the quality of life and life expectancy of patients with AE with advanced disease. The strength of our study is the long centre-based experience with and the multidisciplinary care for patients with AE by a stable team on which we base the lessons learnt and the strategies suggested to improve the outcome of these patients.

Given the similarities of AE and malignancies of infiltrative growth and vascular involvement, adapting tools and concepts developed in oncology to AE management is appealing and appears promising: standardised radiological reporting templates, multidisciplinary decision-making and management based on (repeated) highly standardised and stringent assessments (ID specialists, radiologists, gastroenterologists/hepatologists, haematologists, liver surgeons).

To set up a diagnostic pathway and follow-up of vascular involvement, we developed a standardised template for imaging evaluation with a senior radiologist (TFW) experienced in the diagnosis of hepatic malignancies and AE (online supplemental appendix 1). This tool facilitates the primary detection of early vascular involvement and of subtle progression at follow-up. The latter allows early adjustment of treatment (eg, AE growth suppressive benzimidazoles) and follow-up intervals.

Advanced AE requires early multidisciplinary planning to prevent progressive impairment of liver function due to vascular damage. Thereby, liver transplantation can be significantly deferred. The need for prophylactic anticoagulation, surgical resectability (R0, R1, R2) or transplantability must be repeatedly carefully assessed to prevent complications. The OLT indications for AE differ from a wide range of other chronic progressive liver diseases. If vascular and biliary complications are identified and treated early non-curatively resectable AE patients do surprisingly well with AE-growth suppressive benzimidazole treatment.

Radiology
As in oncological patients, multiphase contrast-enhanced imaging studies represent the backbone of AE assessment in general and evaluation of vascular involvement in particular. Imaging can be done either by CT, MRI or enhanced ultrasound. PET/CT is suggested to play a role for evaluation of parasitic activity.16

In addition to the lack of radiation exposure, MRI depicts AE-typical hepatic lesions consisting of varying proportions of cystic, necrotic and fibrotic tissue, much better than CT. We suggest performing a ‘one-stop-shop MRI’ including general non-contrast sequence protocols, dynamic contrast-enhanced imaging (DCE) and MR cholangiography (MRCP).

Regarding vascular involvement, we suggest additional acquisition of a balanced steady-state free-precession sequence protocol. This MRI technique is characterised by high spatial resolution and rapid acquisition times. On the one hand, it provides a non-contrast-enhanced bright blood effect allowing to diagnose vascular lesions

‘vessels’. More detail is required, however, to assess and follow-up critical vascular involvement to prevent complications and postpone OLT in inoperable patients.

We developed a framework for standardised vascular assessment and follow-up of patients with AE to recognise and treat complication early in our centre. The cases presented were selected to illustrate the vascular problems and interdisciplinary solutions.

In our AE cohort, we found an overall vascular involvement with contact to, stenosis or infiltration of vessels in over one-third (37.1%) of patients and clinically manifest vascular infiltration in 8%.

Our retrospective analysis of advanced non-resectable AE patients cannot give a representative estimate of the proportions of the various types and combinations of vascular involvement in AE. Among other reasons, very severe and very advanced AE cases may be over-represented in a tertiary referral centre. The cohort, however, confirms the high morbidity and mortality in patients with vascular involvement and highlights the

Figure 4  IVC occlusion MRI performed at baseline demonstrates a large AE mass in the atrophic left hemiliver (asterisk in A, B). The mass extends from the anterior liver surface to the orifices of the hepatic veins into the IVC. The lesion consists predominantly of liquefactive necrosis. There is complete obstruction of the IVC at the level of the hepatic vein orifices with evidence of macroscopic vascular invasion (arrows in A, C). The intravascular component is confined to the intrahepatic IVC segment. Alternative venous drainage over accessory hepatic veins into the subhepatic IVC is present (open arrows in D, E). There are large venous collateral pathways draining the blood coming from the lower body half into lumbar paravertebral veins (azygous system) and veins of the abdominal wall (open arrows in F). Thus, flow reversal in the subhepatic IVC is supposed to be present in order to drain the hepatic venous blood into the azygous system. AE, alveolar echinococcosis; IVC, inferior vena cava.
Table 1  Evaluation and follow-up of vascular pathology in alveolar echinococcosis compensated advanced chronic liver disease (cACLD)—asymptomatic advanced fibrosis or cirrhosis

<table>
<thead>
<tr>
<th>Infectious diseases</th>
<th>Radiology</th>
<th>Gastroenterology</th>
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<tr>
<td>▶ Start benzimidazole treatment with a low dose (albendazole 200 mg/day; mebendazole 500 mg/day) if suspected cACLD. Close monitoring for drug toxicity (liver function tests, full blood count, hair loss, drug levels). Stepwise (2–4 weeks) dose increase if feasible.</td>
<td>▶ ‘One-stop-shop MRI’ including general non-contrast sequence protocols, dynamic contrast-enhanced imaging and MR cholangiography balanced steady-state free-precession sequence protocol.</td>
<td>▶ IVC or PV obstruction</td>
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</table>
| ▶ Monitor for development of cACLD by liver stiffness measurement with transient elastography (TE). Refer suspected cACLD patients (TE 10–15 kPa) to hepatologist for further workup. | ▶ Consider contrast-enhanced CT in case of hilar infiltration and/or hepatic artery involvement. | ▶ Evaluate for oesophageal varices—set follow-up intervals
| ▶ Evaluate surgical treatment options to prevent long-term complications of vascular pathology even if R0 resection is not possible. | ▶ Use structured reporting for assessment of vascular and biliary involvement. | ▶ No varices: screening at diagnosis and then yearly till 2 years after diagnosis.
| | ▶ Imaging guides determination of R0 resectability | ▶ ≥Grade I: yearly follow-up.
| | ▶ Monitor hepatic vasculature and IVC by colour-coded duplex ultrasonography at 3–6 months intervals. | ▶ ≥Grade II
| | | ▶ Primary prophylaxis of portal hypertension-related bleeding: NSBB for grade II and III varices.
| | | ▶ Secondary prophylaxis of portal hypertension-related bleeding: NSBB and band ligation.
| | | ▶ IVC or hepatic vein obstruction.
| | | ▶ Evaluate for secondary Budd-Chiari-Syndrome.
| | | ▶ Discuss indication for anticoagulation and weigh anticoagulation benefit against bleeding risk.

IVC, inferior vena cava; NSBB, non-selective beta-blocker; PV, portal vein.

when DCE is indeterminate or limited by artefacts. On the other hand, it allows simultaneous evaluation of the biliary ducts comparable to classical MRCP sequence protocols.

Hilar infiltration and, thus, involvement of the HA and the main PV may be difficult to identify using MRI when stenosis is absent, because the lesion-to-background contrast is frequently poor in fat-saturated T1-weighted images used for DCE. Because of its assumed superiority for demonstrating hypoenhancing perivascular hilar extension of AE, contrast-enhanced CT may be performed in addition to MRI.

Comparable to malignancies, the HA is the most likely also more resistant to AE vessel wall infiltration than venous vessels. Thus, arterial stenoses are rarely seen even in extensive AE lesions. Pancreatic cancer frequently shows a perivascular growth pattern like AE. In the former, an encasement of a vessel of at least 180° reliably indicates infiltration, there are, however, no data available for AE supporting the differentiation between vessel contact and infiltration.

Involvement of the main PV must trigger careful analysis of signs of portal hypertension, such as presence of portosystemic collateral pathways, splenomegaly and ascites. There are data available suggesting that oesophageal varices with diameters of at least 4 mm at CT have a high bleeding risk and that performance of imaging for identification of oesophageal varices is reasonably good to help to avoid unnecessary endoscopic procedures in patients with liver cirrhosis. However, in standard clinical practice, presence of significant PV pathology generally should trigger endoscopy for gold standard assessment of oesophageal varices.

In analogy to the examination of liver transplants, haemodynamic impact of main PV stenoses may be assessed by colour-coded duplex sonography. The relationship of AE lesions to the left or right branch of the PV is important for surgical planning, because involvement of one of these vessels generally requires corresponding hemihemipatectomy.

HV frequently represent a guiding anatomic structure for intrahepatic AE growth. This results in a territorial or wedge-shaped morphology commonly observed in larger AE lesions. Nevertheless, stenoses and occlusion of HVs occur regularly.

Macroscopic vascular invasion can be seen especially in cases of HV infiltration and may contribute to lung metastases.

Involvement of all HVs or involvement of the IVC at the level of the HV orifices may lead to Budd-Chiari syndrome.

The presence and degree of IVC stenosis can be hard to assess by CT or MRI, because the diameter of the IVC is variable and depends, for example, on the respiratory phase. As the IVC diameter decreases during inspiration, the presence of IVC stenoses may be overestimated in CT or MRI scans, typically acquired during deep inspiration. Ultrasound is the primary tool to analyse haemodynamics of the IVC including assessment of the respiratory distensibility.
Gastroenterology

Splanchnic vein occlusion through infiltration of vessels is the main vascular pathology caused by hepatic AE. The resulting secondary Budd-Chiari-Syndrome (BCS) or PV occlusion is most similar to vascular pathologies caused by malignancies. The main difference with regard to malignancies being that AE growth can be reliably halted over decades with benzimidazole treatment. This means that the prognosis of disease is highly dependent on optimal management of vascular (and if present biliary) pathology. This highlights the need for a thorough workup of the extent of vascular damage. Treatment of splanchnic vascular complications due to AE should be managed according to treatment recommendations for vascular diseases of the liver and adapted where needed.

Nevertheless, the decision of the type of anticoagulant medication and the duration of anticoagulant therapy in vascular complications in AE are mostly based on case-by-case decisions and best discussed in the multidisciplinary treatment team.

CONCLUSIONS AND RECOMMENDATIONS

Too often the primary treatment focus in AE is on antiparasitic treatment. Since parasitic growth is slow, there is no need to rush starting antiparasitic treatment, however. The focus at first presentation is on complete evaluation of vascular (and biliary) involvement assessing curative resectability. If this is not possible, we suggest the approach detailed in table 1. The focus in non-resectable AE is on prevention of vascular (and biliary) complications while suppressing growth of AE lesions by carefully monitored benzimidazole treatment to defer OLT and improve the quality of life of patients.

Contributors

Conception of the work: MS and TJ. Acquisition and interpretation of data: PG, UM, TJ, TFW and MS. Drafting and revising the work: PG, UM, TJ, TFW and MS. Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved: PG, UM, TJ, TFW and MS. MS is the guarantor, accepts full responsibility for the conduct of the study and controlled the decision to publish.

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Competing interests

None declared.

Patient consent for publication

Consent obtained directly from patient(s).

Ethics approval

The study was approved by the Ethical Board of the University of Heidelberg (UKHD) (S039/2013).

Provenance and peer review

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Data availability statement

All data relevant to the study are included in the article or uploaded as online supplemental information.

Supplemental material

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REFERENCES


STRUCTURED REPORTING: ALVEOLAR ECHINOCOCCOSIS (AE)

› GENERAL INFORMATION

» AE treatment context

» Comparison

· Most recent imaging: DD/MM/YYYY

› FINDINGS: ALVEOLAR ECHINOCOCCOSIS

» Description (free text)

» Extrahepatic involvement

· Infiltration of neighbouring organs: YES/NO
· Involvement of lymph nodes: YES/NO
· Distant metastases:
  Lung YES/NO – imaging not done/available
  Brain YES/NO – imaging not done/available

» Relation to biliary vessels

· D. hepatocholedochus: -
· D. hepaticus sin.: -
· D. hepaticus dex.: -
· Changes compared to previous imaging: -
· Free text: -

» Relation to portal vein

· Portal vein: -
· Left portal vein: -
· Right portal vein: -
· Changes compared to previous imaging: -
· Free text: -

» Relation to hepatic veins

· Left hepatic vein: -
· Middle hepatic vein: -
· Right hepatic vein: -
· Inferior vena cava: -
· Changes compared to previous imaging: -
· Free text: -

» Relation to hepatic arteries

· A. hepatica (communis/propria): -
· Tr. coeliacus: -
· A. mesenterica superior: -
· Aorta: -
· Changes compared to previous imaging: -
· Free text: -
REFERENCE MEASUREMENTS

- Lesion 1:
- Lesion 2:

FINDINGS: OTHER

- Description (free text)

SUMMARY

- Alveolar echinococcosis
- Category: Stability of disease - Progression of disease
- Reference imaging date: DD/MM/YYYY
- PNM classification: Px Nx Mx
- Free text:

- Other findings

-