Progressive pericardial echinococcosis with intra- and extra-cardiac complications: a multidisciplinary approach

Jason L Walsh[1]
Carine J Moezinia[2]
Amit KJ Mandal[1]
David Lawrence[3]
Constantinos G Missouris[1,4]

Author details can be found at the end of this article

Correspondence to:
Jason L Walsh;
jason.walsh2@nhs.net

Introduction

Echinococcosis or hydatid disease is a zoonotic infection caused by the larval stages of the *Echinococcus* genus of tapeworms, commonly *E. granulosus*. *E. granulosus* is found in every continent except Antarctica. Within Europe, the European Centre for Disease Prevention and Control (2022) reported 243 confirmed cases of human *E. granulosus* infection in 2020, with the highest reported prevalence in Bulgaria.

Infection manifests as slow-growing cysts, typically in the liver and lungs, but can affect numerous other sites. Symptoms may result from compressive effects of the cystic lesions at the infected site or from cyst rupture, which may cause pain, fever, urticaria, eosinophilia and/or anaphylaxis.

This article presents a rare case of progressive pericardial echinococcosis. The magnetic resonance and echocardiographic images presented are unique, demonstrating the potential compressive and haemodynamic effects of pericardial echinococcosis, leading to venous stasis, intra-cardiac thrombus formation and pulmonary embolism. Further, hydatid fluid embolism is highlighted.

Discussion

This case highlights the compressive and haemodynamic effects of advanced pericardial hydatid disease leading to venous stasis, intra- and extracardiac thrombus formation and

Case report

In October 2019 a 31-year-old woman presented to the authors' institution with fever and rigours after a short history of right loin pain. Her medical history was significant for disseminated echinococcosis, which had been diagnosed in rural Romania in 2010. She had undergone craniotomy and resection of a cerebral cyst followed by a 2-year course of albendazole, 400 mg twice daily. Since then, she had been symptom free and had been resident in the UK since 2011. She was taking no regular medications.

Computed tomography of the thorax, abdomen and pelvis at the time demonstrated liver, kidney, spleen and pericardial hydatid cysts. Imaging supported rupture of a right renal cyst, thought to be the cause of her symptoms. She underwent right ureteroscopy with removal of the infected material, which cultured *Escherichia coli*. Repeat blood and urine cultures were also positive for *E. coli* and her C-reactive protein level peaked at 418 mg/litre. She was treated for *E. coli* sepsis with ciprofloxacin as per culture sensitivities to good effect.

Transthoracic echocardiography demonstrated a polycystic mass in the pericardium compressing the right atrium and inferior vena cava (Figure 1). Expectant observation with surveillance transthoracic echocardiography was deemed appropriate.

She was lost to follow up during the COVID-19 pandemic. Transthoracic echocardiography in December 2021 showed enlargement of pericardial cysts with increasing inferior vena cava and right atrium compression. Cardiac magnetic resonance imaging was subsequently performed to further characterise the pericardial lesions and their compressive effects on the right atrium and inferior vena cava (Figure 2). This confirmed cysts within the pericardial space and increasing compression of the right atrium with upstream superior and inferior caval dilatation. Discussions between cardiologists and cardiac surgeons lead to a decision for urgent elective surgical management with median sternotomy and prophylactic excision of the large pericardial cyst, pending advice from infectious disease physicians regarding adjunctive perioperative pharmacotherapy.

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Case report (continued)

In January 2022 the patient presented with pleuritic chest pain and dyspnoea. Clinical examination revealed tachypnoea and tachycardia. Computed tomography pulmonary angiography demonstrated bilateral segmental and subsegmental pulmonary emboli. She was anticoagulated with dalteparin 11 500 IU subcutaneously once daily. Multidisciplinary team discussions between cardiologists, cardiac surgeons and infectious disease physicians again confirmed the decision for surgical resection, but with surgery delayed for 6 months to allow some resorption of the pulmonary thrombi. A hydatid enzyme-linked immunosorbent assay was positive and Western blot confirmed *E. granulosus*. Praziquantel 1 g twice daily was prescribed preoperatively, with a plan to start lifelong albendazole 800 mg twice daily postoperatively. Initiation of albendazole was not recommended preoperatively because of the increased risk of cyst rupture resulting from its direct softening effect on cystic membranes (Usluer et al, 2014).

In August 2022, shortly before the planned surgery, the patient represented with paroxysmal episodes of dyspnoea accompanied by widespread pruritus. Clinical examination was unremarkable. Laboratory investigations were notable for significantly elevated eosinophil count (19.3×10^9/litre) compared to baseline (2.8×10^9/litre). Computed tomography of the thorax, abdomen and pelvis with pulmonary angiography showed progression of hydatid disease burden since 2019. Some pulmonary filling defects were of low to intermediate attenuation, supporting hydatid fluid emboli. Transthoracic echocardiography demonstrated further enlargement of the pericardial cysts with a mobile mass, likely thrombus, within the right atrium (**Figure 3** and **Video 1**). The patient was promptly transferred for definitive surgical intervention.

At operation, a cluster of intrapericardial hydatid cysts (**Figures 1–3** and **Video 1**) was found densely adherent and fistulating to the right atrium with compression of right atrium, inferior vena cava and right ventricle. Subsequently, the pericardium and right atrium were opened, intrapericardial cysts were excised and the right atrium defect repaired. The patient had an uncomplicated postoperative recovery. On outpatient review in September 2022, she felt well and has resumed normal activities of daily life. She has been prescribed lifelong albendazole therapy and long-term anticoagulation with a novel oral anticoagulant. An annual cardiology review with repeat transthoracic echocardiography and 6-month infectious disease clinic review have been planned.

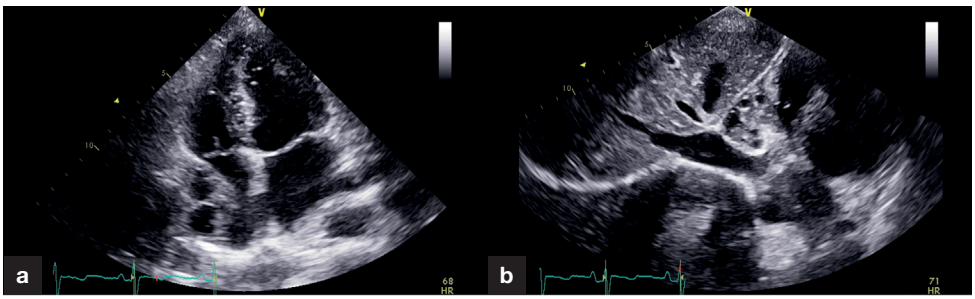

Figure 1. Transthoracic echo in 2019. a. Four-chamber view showing a cluster of cystic lesions compressing the right atrium. b. Subcostal view showing a cluster of cystic lesions compressing the inferior vena cava and right atrium.

pulmonary migration. In progressive disease this may be further complicated by cyst rupture and hydatid fluid emboli, as illustrated in this report. The emphasis is therefore on multidisciplinary team management and early surgical resection of pericardial hydatid disease, even if clinically silent, followed by long-term medical therapy to avoid these life-threatening sequelae.

Author details

[1]Department of Medicine, Wexham Park Hospital, Frimley Health NHS Foundation Trust, Slough, UK

[2]Department of Medicine, University College Hospital London, London, UK

[3]Department of Cardiothoracic Surgery, St Bartholomew's Hospital, London, UK

[4]University of Nicosia Medical School, Cyprus

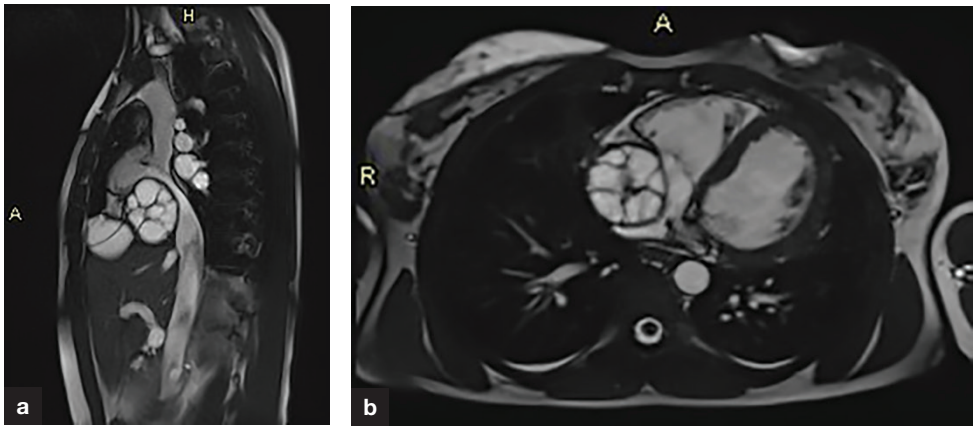

Figure 2. Cardiac magnetic resonance in 2021. a. Sagittal view showing a cluster of cysts compressing the inferior vena cava. b. Transverse view showing a cluster of cysts compressing the right atrium.

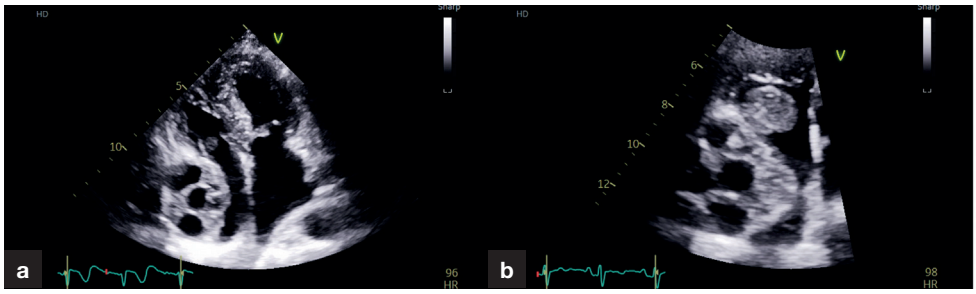

Figure 3. Transthoracic echocardiography in 2022. a. Four-chamber view showing a polycystic mass compressing the right atrium. b. Zoomed four-chamber view showing the polycystic mass compressing the right atrium and a solid intra-atrial mass highly suspicious of thrombus.

Video 1. Transthoracic echocardiography in 2022 demonstrating a polycystic pericardial mass compressing the right atrium and a hypermobile solid mass in the right atrium cavity suspicious of thrombus.

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Learning points

- The compressive and haemodynamic effects of progressive pericardial hydatid disease may culminate in intra- and extracardiac thrombus formation through venous stasis, leading to pulmonary artery thrombosis.
- Hydatid cyst rupture may be complicated by hydatid fluid emboli.
- In hydatid disease with pericardial involvement, a multidisciplinary approach with early surgical resection alongside adjunctive medical therapy is emphasised.