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CASE REPORT

Chronic pericardial effusion in the setting of pericardial capillary haemangioma: a case report and review of the literature

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Introduction Cardiac haemangiomas are rare vascular tumours of the heart accounting for less than 5% of benign primary cardiac neoplasms. They are sometimes diagnosed incidentally, since patients can be asymptomatic. The clinical presentation in symptomatic patients, however, is variable, depending on size and exact localization of the tumour. Although cardiac haemangiomas have been reported everywhere in the heart, those localized in the pericardium are extremely rare.

Case presentation A 48-year-old female patient with a history of pericardial effusion and pneumonia was admitted to our hospital with progressive dyspnoea on exertion. Echocardiography demonstrated recurrence of pericardial effusion with 'swinging heart'. Further investigation by computed tomography, cardiac magnetic resonance imaging and coronary angiography revealed a hypervascular pericardial mass with typical 'tumour blush' after contrast injection. The tumour could be resected *in toto* by open heart surgery, and histological evaluation confirmed the diagnosis of a pericardial capillary haemangioma. There were no signs of recurrence of neither the pericardial effusion nor the tumour during follow-up.

Discussion We here report a very rare case of a pericardial haemangioma in the adult which was diagnosed by multi-modality workup of recurrent pericardial effusion. This case illustrates that in the setting of chronic pericardial effusion non-inflammatory and non-malignant causes should be taken into account.

Keywords Chronic pericardial effusion • Capillary haemangioma • Cardiac tumour • Case report

Learning points

- Pericardial haemangiomas are very rare and can cause variable symptoms depending on size and localization.
- In the setting of chronic pericardial effusion non-inflammatory and non-malignant causes should be taken into account. Computed tomography and cardiac magnetic resonance imaging help identifying such underlying diseases.

Introduction

The prevalence of primary cardiac tumours is estimated between 0.0017% and 0.33% with 75% of them classified as benign. Among these, cardiac haemangiomas, which are characterized by excessive endothelial proliferation, account for less than 5%.[1] They have been reported in different localizations within the heart, but especially pericardial occurrence is extremely rare. Whilst cardiac haemangiomas are sometimes diagnosed incidentally or at autopsy, the clinical presentation of symptomatic patients is variable including chest pain, arrhythmia, syncope or pericardial effusion depending on size and localization of the tumour.[2]

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We hereby report a case of pericardial haemangioma in an adult, which is extremely rare and has thus been reported in only a few patients so far.[3–13]

Timeline

Time	Events
3 months earlier	Hospitalization due to pneumonia treated with antibiotics. First notice of pericardial effusion-treatment with NSAID initiated
1 month earlier	Constant pericardial effusion despite NSAID treatment. Steroids added
Day 1	Patient presents to the emergency room with progressive exertional dyspnoea. Echocardiography shows progressive pericardial effusion with 'swinging heart'.
Day 2	Computed tomography reveals pericardial tumour
Day 5	Further non-invasive tissue characterization by cardiac magnetic resonance imaging
Day 6	Angiogram performed: Feeding vessels of the tumour arise from the left anterior descending artery.
Day 9	Complete surgical resection of the tumour. Diagnosis: Capillary haemangioma
6 months later	Follow-up without signs of recurrence

NSAID, non-steroidal anti-inflammatory drug.

Case presentation

A 48-year-old white female patient was admitted for workup of chronic pericardial effusion, which was first detected during an episode of pneumonia 3 months earlier, but remained constant despite complete remission of pneumonia after antibiotic treatment. At the time of admission, the patient was in a stable haemodynamic condition, but suffered from progressive dyspnoea on exertion (New York Heart Association Class III).

Lung auscultation on admission revealed diminished breath sounds and dull percussion sounds of the basal right lung. The rest of the physical examination, especially cardiac auscultation was inconspicuous with a regular rate and rhythm, normal heart sounds, and no murmurs. Initial vital parameters and laboratory testing were also normal. Bedside echocardiography demonstrated a large pericardial effusion up to 35 mm with 'swinging heart' phenomenon (*Figure 1*). Left and right ventricular systolic function as well as valvular function was normal. End-diastolic collapse of the right atrium and increased respiratory variation of mitral and tricuspid valve flow velocities have been noticed as indicators of a beginning haemodynamic relevance of the pericardial effusion, but diastolic right ventricle relaxation and vital parameters were not compromised. Because of the chronic setting of the large symptomatic pericardial effusion, pericardiocentesis was performed demonstrating a serous pericardial effusion with low cell count and no evidence of purulence or malignity. Besides the known pericardial effusion, computed tomography (CT) revealed an unclear mass in the pericardial space (*Figure 2*). The mass was attached to the anterior wall of the left ventricle, measuring $20 \times 22 \times 8$ mm, and demonstrated pronounced peripheral uptake of contrast media. For additional non-invasive tissue characterization cardiac magnetic resonance imaging (CMR) was performed using a 1.5-T Magnetom Aera (Siemens Medical System). Cine MR images confirmed residual, partially organized pericardial effusion without haemodynamic relevance, as well as the previously described mass (*Figure 3* and Supplementary material online, *Files S1–S3*), demonstrating isointense signal on T1- and hyperintense signal on T2-weighted images (*Figure 4A*). Most common entities of tumours in this location are pericardial cysts and lipomas, which both could be

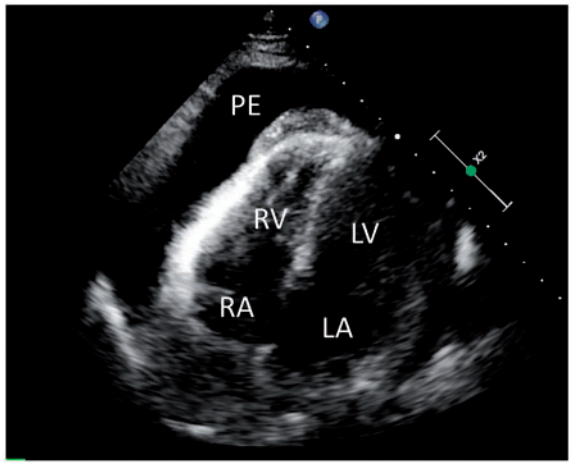

Figure 1 Bedside echocardiography showing large pericardial effusion. LA, left atrium; LV, left ventricle; PE, pericardial effusion; RA, right atrium; RV, right ventricle.

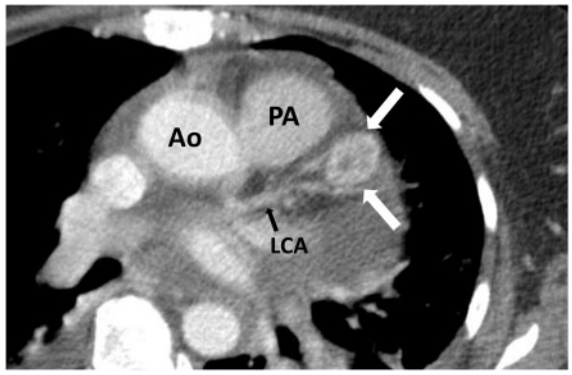

Figure 2 Computed tomography showing circumscribed mass (arrows) measuring $20 \times 22 \times 8$ mm attached to the anterior wall of the left ventricle in proximity to the left anterior descending artery. Ao, ascending aorta; LCA, left coronary artery; PA, pulmonary artery.

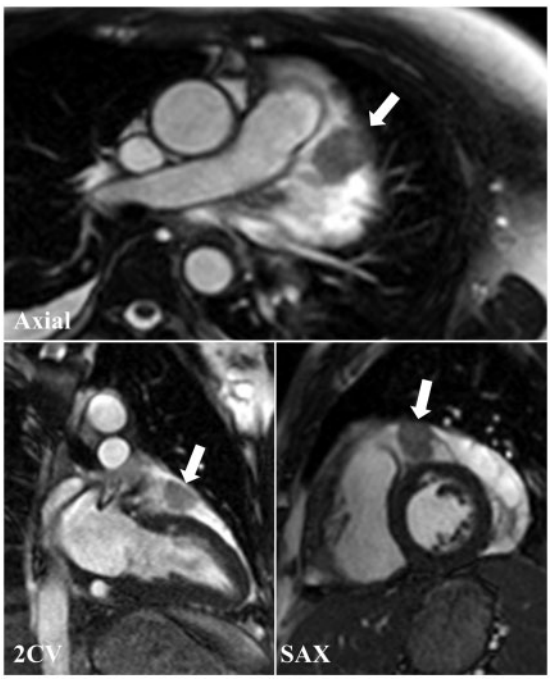

Figure 3 Steady state free-precession cine cardiac magnetic resonance imaging confirming suspicious mass. See also Supplementary material online, *Files S1–S3*. SAX, short axis; 2CV, 2-chamber view.

ruled out by T1- and T2-weighted MR images, fat saturation, and perfusion studies (*Figure 4*). After administration of gadolinium contrast, CMR first-pass perfusion depicted peripheral perfusion of the mass, suggesting a hypervascular tumour (*Figure 4B* and Supplementary material online, *File S4*). Late gadolinium enhancement showed intense inhomogeneous signal after 5 min indicating slow blood flow (*Figure 4C*). In regards to the marked arterial perfusion of the mass as well as its proximity to the left anterior descending artery (LAD) and its diagonal branches, coronary angiography was performed. Contrast injection led to a 'tumour blush', emphasizing the vascular characteristic of the tumour, and revealed feeding arteries from the LAD (*Figure 5*).

With the diagnosis of chronic pericardial effusion, as well as an unclear hypervascular and hyperperfused tumour with peripheral contrast uptake on CT and CMR located within the pericardial space in close proximity to the LAD, a decision for open surgical exploration via anterolateral thoracotomy was made by our Heart Team. Intraoperative frozen section was indicative for a benign vascular tumour, most likely a haemangioma. Consequently, the tumour was removed completely and specimens were further evaluated. Histological workup revealed numerous, capillary-type small vessels within a fibro-oedematous background. Staining for CD31 and ETS-related gene (ERG) expression were positive, highlighting endothelial cells. Proliferation rate of less than 5% was assessed using Ki67-staining. Thus, the differential diagnosis of a malignant neoplasm including metastasis, which is much more common than primary pericardial tumours, could be excluded and the final diagnosis of a capillary-type

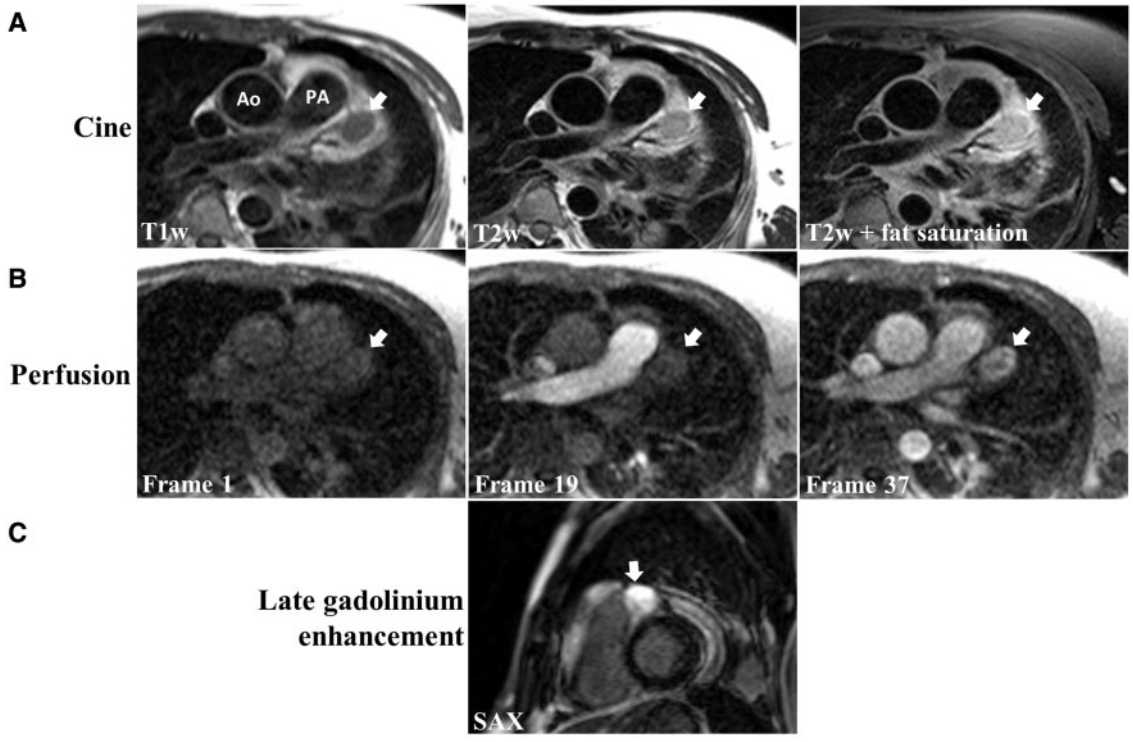

Figure 4 (*A*) Tumour (arrows) demonstrates isointense signal in T1-weighted (left) and hyperintense signal in T2-weighted (middle) images. Fatty tissue could be ruled out by fat saturation (right). Representative images of cardiac magnetic resonance imaging first-pass perfusion (*B*) depict early peripheral perfusion of the mass (see also Supplementary material online, *File S4*). Late gadolinium enhancement (*C*) sequence shows intense enhancement after 5 min. Ao, ascending aorta; PA, pulmonary artery; SAX, short axis; T1w, T1-weighted; T2w, T2-weighted.

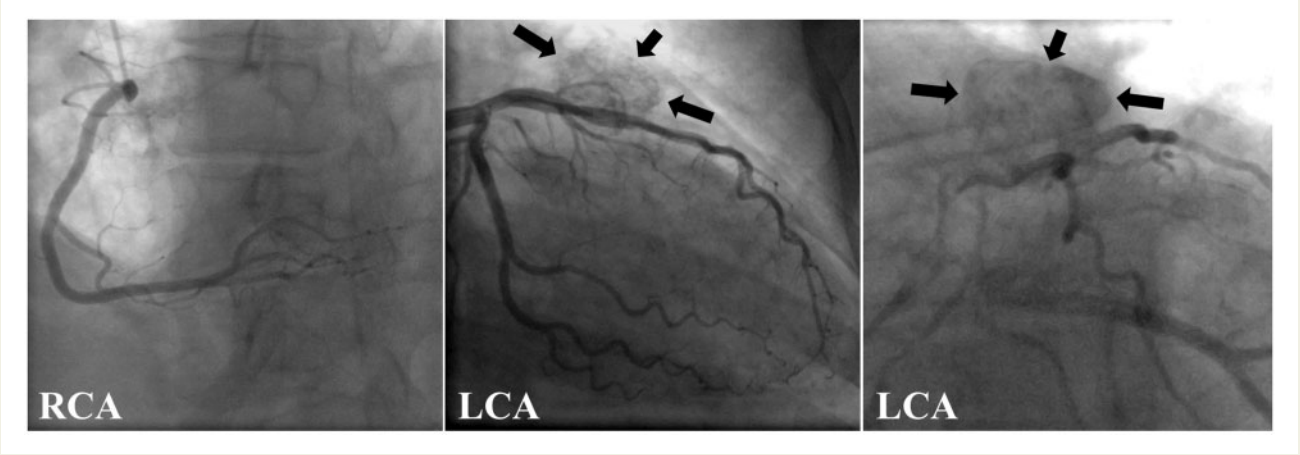

Figure 5 Coronary angiography reveals hypervascularization of the tumour (arrows) and shows characteristic 'tumour blush'. Feeding arteries of the tumour arise from the left coronary artery (middle, right). LCA, left coronary artery; RCA, right coronary artery.

Table 1 Previously reported cases of pericardial haemangioma in adult patients

Author	Year	Patient age	Clinical presentation	Complete resection?
Hicken *et al.*[3]	1963	36	Syncope	No
Ramasubbu *et al.*[4]	2004	44	Chest pain	?
Zeina *et al.*[5]	2007	37	Syncope, palpitation	No
Ediae *et al.*[6]	2009	75	Asymptomatic, incidental finding	Yes
Liebetrau *et al.*[7]	2010	58	Dyspnoea, pericardial effusion	Yes
Omura *et al.*[8]	2010	78	Dyspnoea, pericardial effusion	Yes
Gupta[9]	2013	40	Chest discomfort, palpitation	Yes
Ben Youssef *et al.*[10]	2014	24/79	Palpitations	Yes
Sabeti *et al.*[11]	2015	72	Asymptomatic, incidental finding	Yes
Vargis *et al.*[12]	2017	63	Dyspnoea	Yes
Sbrana *et al.*[13]	2017	78	Dyspnoea, haemorrhagic pericardial effusion	?

haemangioma of the pericardium was made. After surgery, the patient rapidly improved and at follow-up 6 months after the initial presentation neither the tumour nor the pericardial effusion recurred.

Discussion

Cardiac haemangiomas are rare vascular tumours of the heart accounting for less than 5% of benign primary cardiac neoplasms. The natural history of cardiac haemangiomas is variable, ranging from asymptomatic persistence to life-threatening complications.[14,15] Thus, surgical removal remains the treatment of choice, yielding an excellent long-term prognosis and low recurrence rate.

To our knowledge, since the 1960s only 12 cases of pericardial haemangioma in the adult have been reported in the literature, as summarized in detail in *Table 1*. Some of them were accompanied by pericardial effusion as in the actual case. Though, the underlying mechanism of pericardial effusion in the setting of pericardial haemangiomas is not fully understood yet. Pericardial friction at the site of the tumour as well as rupture of tumourous microvessels are considered possible explanations for the fact that both serous and haemorrhagic pericardial effusions have been observed. Pericardiocentesis has low value in diagnosing pericardial haemangioma. However, depending on the clinical setting and the extend of the pericardial effusion it may be indicated, i.e. for cardiac tamponade, large symptomatic effusions not responding to medical therapy, or if bacterial or neoplastic origin is suspected (Class I recommendation according to the European Society of Cardiology guidelines for management of pericardial disease).[16] Considering the possible fatal complications including arrhythmia and injury of cardiac, hepatic, or pulmonary structures, the procedure should be performed by an experienced operator.

This case does not only illustrate the multi-modality workup of an unclear cardiac tumour but also nicely underscores that in the setting of chronic pericardial effusion non-inflammatory and non-malignant causes should be taken into account.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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