ESC European Society of Cardiology

European Heart Journal - Case Reports (2023) **7**, 1–4
https://doi.org/10.1093/ehjcr/ytad343

CASE REPORT
Cardiovascular surgery

Composite haemangioendothelioma in the heart: a case report

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Received 12 August 2022; first decision 6 April 2023; accepted 19 July 2023; online publish-ahead-of-print 21 July 2023

Background Primary cardiac neoplasm is rare and generally benign. Epithelioid haemangioendothelioma, a potentially malignant tumour of vascular origin, has been occasionally described in the heart. Composite haemangioendothelioma, characterized by a heterogeneous architecture of vascular components and usually located in soft tissue of the extremities, has only been reported twice in the heart. We herein report another case of this extremely uncommon cardiac tumour.

Case summary Comprehensive cardiac examination of a 59-year-old female patient with palpitations and personal history of Hodgkin's lymphoma and chest radiation revealed a mass in the left atrium. After surgical resection, histopathological and immunohistochemical analysis identified a composite haemangioendothelioma. After two years, repeated imaging revealed neither signs of local relapse nor metastasis.

Conclusions Composite haemangioendothelioma, a very uncommon form of potentially malignant vascular tumour, can also be encountered in the heart. In this present case, the outcome was favourable two years after surgical resection without adjuvant therapy.

Keywords Case report • Primary cardiac neoplasm • Composite hemangioendothelioma • Hodgkin's lymphoma

ESC curriculum 6.8 Cardiac tumours • 7.5 Cardiac surgery

Learning points

- Malignant primary cardiac tumours are rare and can present with non-specific symptoms.
- Haemangioendothelioma, a potentially malignant tumour of vascular origin, has been occasionally described in the heart. Composite haemangioendothelioma, a very rare subtype characterized by a heterogeneous architectural pattern of vascular components and usually located in the soft tissue of extremities, can also be found in the heart.
- Non-metastatic, primary resectable cardiac tumour of vascular origin with potential malignity should be addressed surgically. The currently available data are insufficient to conclude on the benefit of adjuvant therapy.

Introduction

Primary cardiac neoplasm is rare and generally benign.[1] The clinical presentation is usually non-specific and unrelated to the histological type and dignity of the tumour.[2] Several malignant cardiac neoplasms of vascular origin have been described, but little is known about haemangioendothelioma and its different subtypes. We herein report an extremely rare case of composite haemangioendothelioma in the heart.

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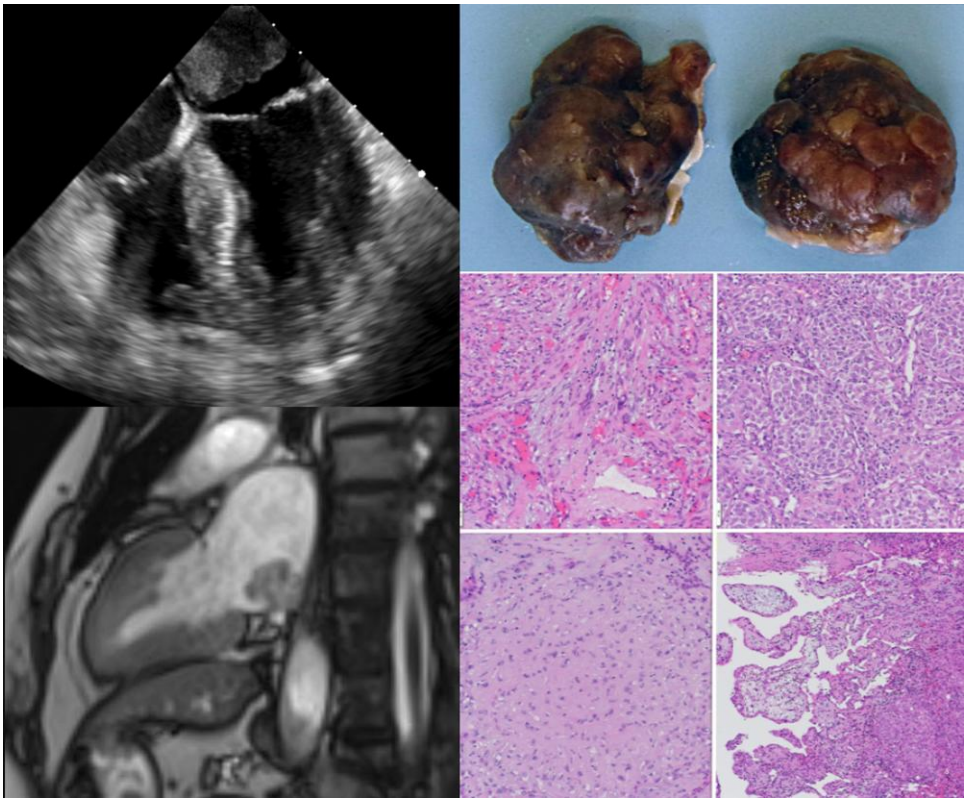
Handling Editor: Nikolaos Bonaros
Peer-reviewers: A Shaheer Ahmed; Katharine Kott
Compliance Editor: Emmanouil Mantzouranis

Summary figure



Case presentation

A 59-year-old Caucasian woman underwent cardiological examination due to palpitations over the past two years. Her medical history was relevant for Hodgkin's lymphoma, treated with chest radiation and splenectomy at the age of 30. The patient was non-smoker and reported no illicit drug use. Apart from a slightly accelerated pulse, vital signs and physical examination were unremarkable. Electrocardiogram showed a sinus tachycardia. Echocardiography revealed a 2 × 3 cm mass in the inferior wall of the left atrium with no other cardiac abnormality (*Figure 1A–B*). Cardiac magnetic resonance imaging further described the mass as highly vascularized and infiltrating the inferior wall of the left atrium (*Figure 1C–D*). Cardiac magnetic resonance cine images best delineated the mass. In T1- and T2-weighted images, the mass appeared isointense and hyperintense to myocardium, respectively. Gadolinum infusion revealed a positive contrast uptake on first-pass perfusion and a strong, heterogeneous delayed hyperenhancement. Computed tomography angiogram of the thorax identified no metastasis. The patient was referred to our tertiary centre for surgical treatment. After sternotomy and initiation of cardiopulmonary bypass with bicaval cannulation, the tumour was exposed via left atriotomy and macroscopically excised. A subsequent partial resection of the posterior wall of the left atrium was reconstructed with bovine pericardium. The postoperative course was, apart from a single episode of paroxysmal atrial fibrillation, uneventful, and the patient could be promptly discharged.

Histopathological analysis of the tumour revealed heterogeneous pattern of epithelioid, stromal, vasogenic, and spindled cells, as well as hyaline matrix (*Figure 2*). The proliferation rate was moderate (1/10 high-power fields, Ki-67 proliferation index 20%). Cardiac myxoma and local recurrence of Hodgkin's disease were excluded with negative calretinin and specific immunohistochemistry (CD30, CD15, and Pax5), respectively. The partial expression of endothelial markers (CD31 and ERG) pointed to a vascular tumour. The WWTR1-CAMTA1 fusion, specific of the epithelioid type of hemangioendothelioma (HE), was negative. After external analysis for second opinion, the tumour was finally designated as composite haemangioendothelioma.

After 24 months, the patient was without cardiac or general symptoms. Repeated echocardiography and full body positron emission tomography–computed tomography revealed no signs of local relapse or metastasis.

Discussion

Epithelioid haemangioendothelioma (EHE) is a rare, potentially malignant neoplasia of vascular origin with an estimated prevalence of less than one in 1 million and a recently reported incidence of 0.03/10^6

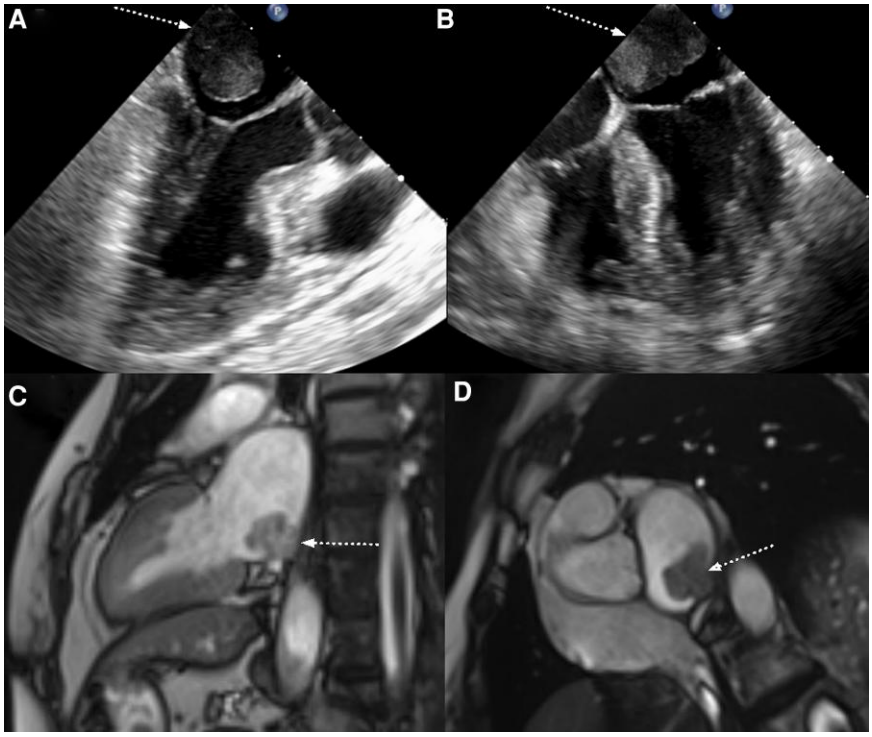


Figure 1 Transoesophageal echocardiography: (*A*) three-chamber view and (*B*) four-chamber view—voluminous mass (arrow) in the left atrium. Cardiac magnetic resonance imaging: (*C*) vertical long-axis and (*D*) short-axis view—mass (arrow) arising from the inferior wall of the left atrium.

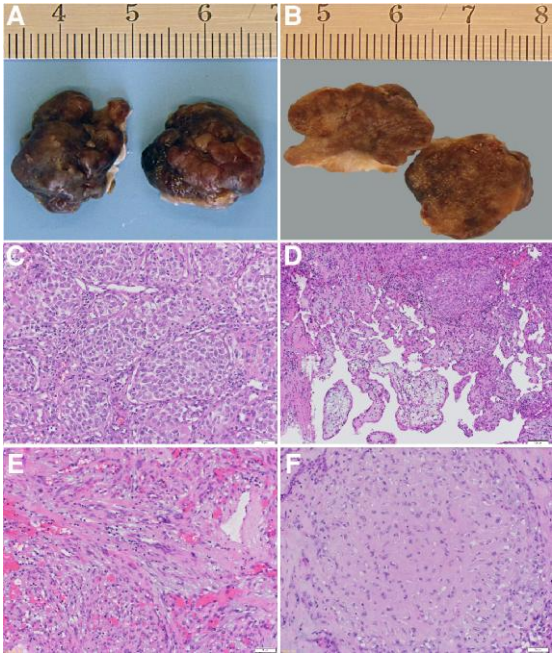


Figure 2 Excised tumour in two fragments: (*A*) external view and (*B*) internal view, with centimeter scale. High-power histological view (H&E stain): heterogeneous pattern of (C) epithelioid, (D) vasogenic, (E) spindled cells, and (F) hyaline matrix.

pat./year.[3–5] Epithelioid haemangioendothelioma is seldom localized in the heart. In the reported cases, outcomes were mostly favourable after complete surgical resection with or without radiochemotherapy.[6–10] Moulai *et al.*[11] reported on a case of cardiac EHE successfully treated with cardiac transplantation after neo-adjuvant chemotherapy. Composite haemangioendothelioma (CHE) differs from EHE by its heterogeneous architectural pattern, variable expression of vascular endothelial markers, and variable degree of malignancy.[12] Until recently, this extremely uncommon tumour was described almost exclusively in soft tissue of the extremities.[13,14] In 2020, Langguth *et al.*[15] reported on a case of CHE arising from the pericardium. In 2022, two cases of cardiac CHE were reported.[16,17] Details on these cases and comparison with the present case are listed in *Table 1*.

Of interest in the present case, the tumour was found in a patient 29 years after chest radiation for Hodgkin's lymphoma. Survivors of Hodgkin's lymphoma are at risk for developing radiation-induced malignancies, but literature is scarce about specific secondary cardiac neoplasms.[18] Angiosarcoma, one of the most common primary cardiac malignancies, has been reported with higher incidence after radiation, regardless of the location and disease treated.[19]

In term of prognosis, more than a 40% recurrence has been reported with CHE arising in the extremities.[14] The currently available data are insufficient to conclude on the benefit of adjuvant therapy. In both first two cases, short-term outcome was favourable after surgical resection. In the case of Zhou *et al.*, the patient was referred to the oncology department after surgery but an adjuvant treatment was not mentioned. In our case, the patient was free of recurrence two years after surgical resection alone. Close and prolonged monitoring is anticipated.

Table 1 Characteristics of cardiac composite haemangioendothelioma cases reported to date

Case	Sex/age (years)	Localization	Size (cm)	Markers	Ki-67 index	Adjuvant therapy	Follow-up (months)
Zhang *et al.*[16]	M/46	RV	2.0 × 1.2 × 0.5	CD-34 ERG	Slightly elevated	No	6
Zhou *et al.*[17]	F/50	RA	5.0 × 4.5 × 3.0	CD-31 ERG FLI-1 D2-40	*NA*	*NA*	1
Schaeffer *et al.*	W/59	LA	2.9 × 3.0 × 2.2	CD-34 ERG GLUT-1 WT-1	20%	No	24

RV, right ventricle; RA, right atrium; LA, left atrium; NA, not available

Lead author biography

Dr T. Schaeffer was a senior resident in the Department of Cardiac Surgery of the University Hospital of Basel, Switzerland, until 2022. He is currently working as a fellow in congenital heart surgery at the German Heart Center Munich, Germany.

Acknowledgements

We acknowledge Prof. Dr med. Thomas Mentzel for contributing to the diagnosis.

Consent: Appropriate informed consent was obtained from the patient for publication in accordance with the Committee on Publication Ethics (COPE) guidelines.

Conflict of interest: None declared.

Funding: None.

Data availability

The data underlying this article will be shared on reasonable request to the corresponding author.

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