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

Andreas Seitz1*, Peter Ong1, Maik Backes2, and Heiko Mahrholdt1

1Department of Cardiology, Robert Bosch Medical Center, Auerbachstr. 110, 70376 Stuttgart, Germany; and 2Department of Radiology, Robert Bosch Medical Center, Auerbachstr. 110, 70376 Stuttgart, Germany

Received 14 November 2017; accepted 30 January 2018; online publish-ahead-of-print 6 March 2018

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%. 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.

* Corresponding author. Tel: +49-711-8101-6048, Fax: +49-711-8101-3795, Email: andreas.seitz@rbk.de. This case report was reviewed by Brian Halliday.

© The Author(s) 2018. Published by Oxford University Press on behalf of the European Society of Cardiology. This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (http://creativecommons.org/licenses/by-nc/4.0/), which permits non-commercial re-use, distribution, and reproduction in any medium, provided the original work is properly cited. For commercial re-use, please contact journals.permissions@oup.com
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 3         | 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 (New York Heart Association Class III).

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 × 22 × 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 × 22 × 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.)
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...
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.

**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 Yousef 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 | ?                  |
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.

References

1. McAllister HA Jr, Primary tumors of the heart and pericardium. Pathol Annu 1979;14(Pt2):325–355.
2. Brizard C, Latremouille C, Jebara VA, Acar C, Fabiani JN, Deloche A, Carpenter AF. Cardiac hemangiomas. Ann Thorac Surg 1993;56:390–394.
3. Hicken WJ, Scherlis S. Angiomatosis of the pericardium. Report of a case and review of the literature. Ann Intern Med 1963;59:236–242.
4. Ramasubbu K, Wheeler TM, Reardon MJ, Dokainish H. Visceral pericardial hemangioma: unusual location for a rare cardiac tumor. J Am Soc Echocardiogr 2005;18:981.e7.
5. Zeina AR, Zaid G, Shafir D, Rosenschein U, Barmer E. Images in cardiovascular medicine. Huge pericardial hemangioma imaging. Circulation 2007;115:e315–e317.
6. Edie J, Lim PS, Addonizio VP, Kostacos E, Bell K, Litt H. Pericardial hemangioma taking origin from the posterior wall of the left atrium. Ann Thorac Surg 2009;87:e54–e56.
7. Liebetrau C, Szalay Z, von Gerlach S, Möllmann H, Nef H, Hamm C, Weber M. Pericardial hemangioma vascularized via left anterior descending. Clin Res Cardiol 2010;99:405–407.
8. Omura K, Onishi T, Yamawaki K, Salem Omar AM, Kanzawa M, Ishida T, Shite J, Kawai H, Itoh T, Okita Y, Hirata K-I. A rare case of pericardial hemangioma with bloody pericardial effusion. J Cardiol Cases 2010;2:e15–e19.
9. Gupta N. Intrapericardial hemangioma: a case report. J Clin Diagn Res 2013;7:169–170.
10. Ben Yousef A, Zairi S, Ouerghi S, Ayadi A, Bousmina M, Beraires F, Kilani T. Epicardial cavernous hemangiomas: a two-case report. Tunis Med 2014;92:268–271.
11. Sabeti S, Zahedifard S, Soleimantabar H, Zarghampour M, Toutkaboni M. Coexistence of pericardial and hepatic hemangiomas. Iran J Pathol 2015;10:167–172.
12. Vargas RS, Phansalkar M, Padhi S, Phansalkar D, Nair SR. Pericardial haemangioma: a common tumour in an unusual location: case report and review of literature. J Clin Diagn Res 2017;11:e15–e17.
13. Sbrana F, Mannucci F, Airo E, Aquaro GD, Prediletto R. Cardiac tamponade due to apixaban therapy in a patient with unknown pericardial hemangioma. Intern Emerg Med 2017;doi:10.1007/s11739-017-1736-3.
14. Palmer TE, Tresch DD, Banchek LJ. Spontaneous resolution of a large cavernous hemangioma of the heart. Am J Cardiol 1986;58:184–185.
15. Solum AM, Romero SC, Ledford S, Parker R, Madani MM, Coletta JM. Left atrial hemangioma presenting as cardiac tamponade. Tex Heart Inst J 2007;34:126–127.
16. Adler Y, Charron P, Inazio M, Badano L, Barón-Escuviñas G, Bogert J, Brucato A, Gueret P, Klingel K, Lions C, Masch B, Mayosi B, Pavie A, Ristic AD, Sabaté Tenas M, Seferovic P, Swedberg K, Tomkowski W, Achenbach S, Agewall S, Al-Attar N, Angel Ferrer J, Arad M, Asteggiaro R, Bueno H, Czifra ALP, Carenz S, Ceconi C, Evangelista A, Flachskampf F, Giannakoulis G, Gielen S, Habib G, Kohl P, Lambrinou E, Lancilotti P, Lazaros G, Linhart A, Meurin P, Nieman K, Piepoli MF, Price S, Roos-Hesselink J, Roubille F, Ruschitzka F, Sagristà Saureda J, Sousa-Uva M, Uwe Vogt J, Luis Zamorano J. 2015 ESC Guidelines for the diagnosis and management of pericardial diseases. Eur Heart J 2015;36:2921–2964.