ESC European Society of Cardiology

European Heart Journal - Case Reports (2019) **3**, 1–3
doi:10.1093/ehjcr/ytz027

Cardiac tamponade as first manifestation of atrial angiosarcoma

Sergio Tapia Concha[1], Miguel F. Carrascosa[2]*, Ivana Pulitani[3], and César Saiz-Pérez[4]

[1]Radiology Department, Hospital of Laredo, Avda. Derechos Humanos s/n, 39770 Laredo, Cantabria, Spain; [2]Internal Medicine Department, Hospital of Laredo, Avda. Derechos Humanos s/n, 39770 Laredo, Cantabria, Spain; [3]Cardiovascular Surgery Department, University Hospital Marqués de Valdecilla, Avda. Valdecilla 25, 39008 Santander, Cantabria, Spain; and [4]Emergency Department, Hospital of Laredo, Avda. Derechos Humanos s/n, 39770 Laredo, Cantabria, Spain

Received 24 July 2018; accepted 27 February 2019; online publish-ahead-of-print 19 March 2019

A 71-year-old man was referred to our hospital's emergency department because of acute non-exertional chest pain quickly followed by syncope. The temperature was 36°C, blood pressure 74/54 mmHg, heart rate 78 b.p.m., and respiratory rate 30 b.p.m.. On examination, patient's jugular veins were distended and heart sounds were muffled; pericardial friction rub was not heard. Blood tests showed creatinine 1.47 mg/dL (normal 0.6–1.18 mg/dL), troponin T 21 ng/L (normal <14), and D-dimer 11 150 ng/mL (normal <500 ng/mL). An electrocardiogram failed to display signs of myocardial infarction or acute pericarditis. Chest computed tomography revealed a pericardial effusion, a right atrial mass (*Figure 1*), and bilateral pulmonary nodules. A subsequent transoesophageal echocardiography performed in the operating room demonstrated cardiac tamponade, a swinging mass in the pericardial fluid that originated from the right atrial appendage (Supplementary material online, *Video*), and the likely presence of an atrio-pericardial fistula. At surgery (*Figure 2A*), since the atrial mass showed arterial bleeding into the pericardial space, direct closure of the defect was carried out; in addition, partial tumourectomy was performed (*Figure 2B*). Pathological testing of the mass was consistent with cardiac angiosarcoma (*Figure 2C and D*). Despite receiving paclitaxel-containing chemotherapy, he died of acute liver failure related to massive tumoural infiltration 3 months after surgery.

It is commonly accepted that metastatic tumours to the heart and pericardium are up to 100–1000 times more frequent than primary cardiac tumours.[1] Primary cardiac tumours are rare disorders with an autopsy incidence of ≤0.030%.[1] In adults almost 25% of primary cardiac tumours are malignant, and approximately a third of the malignant tumours are angiosarcomas. In primary cardiac angiosarcoma (PCA), males are affected more often than females, in a 2–3/1 ratio, and most patients are younger than 65 years.[2] The initial symptoms and signs of cardiac angiosarcoma are non-specific and include chest pain, dyspnoea, anaemia-related fatigue and malaise, and fever.[1,2] Nevertheless, patients are usually symptomless until the tumour has increased to a certain size or regional spread or metastases have developed.[2]

Primary cardiac angiosarcoma arises from the right atrium in nearly 90% of cases, usually as a multicentric mass.[2] In a report from a single centre, 33 cases of cardiac angiosarcoma were identified; of these, 17

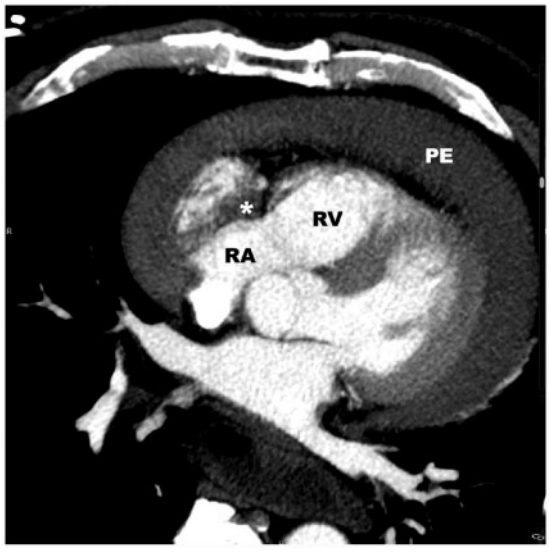

Figure 1 Cardiac angiosarcoma: a devastating condition. Computed tomography chest showing a massive pericardial effusion and an irregular mass (asterisk) seemingly located on the right atrium wall; contrast media extravasation out of the atrial cavity can also be appreciated. PE, pericardial effusion; RA, right atrium; RV, right ventricle.

*Corresponding author. Tel: 34 942638500, Fax: 34 942607876, Email: miguel.carrascosa@scsalud.es
Handling Editor: Timothy Tan

Peer-reviewers: Tamas Habon, Francesca Musella, Esther Cambronero-Cortinas and Christian Jons

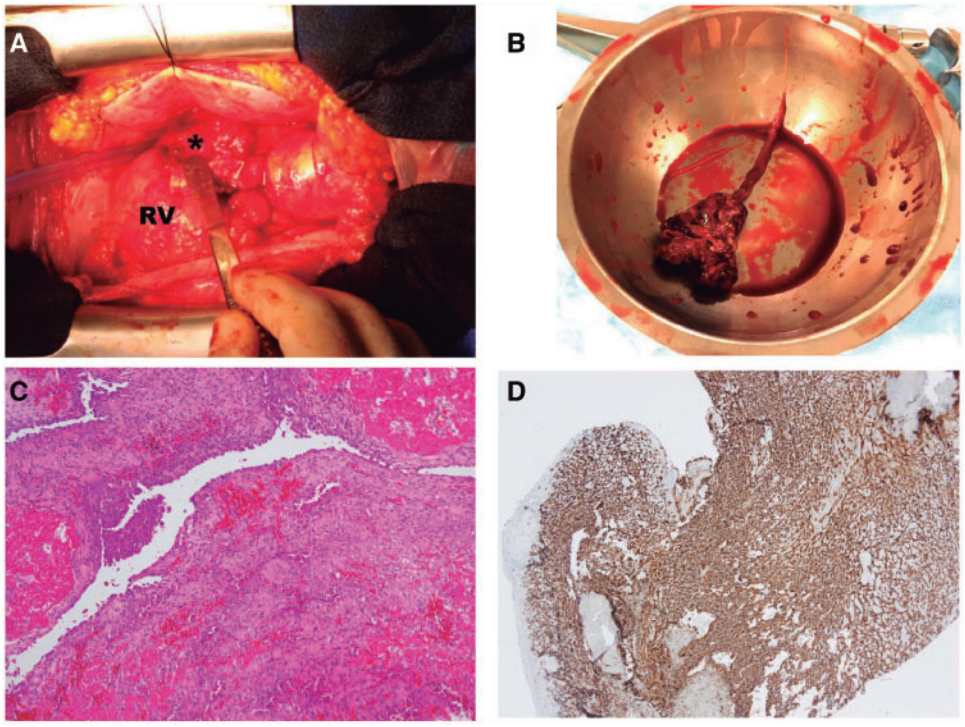

Figure 2 Cardiac angiosarcoma: a devastating condition. (A) Intraoperative appearance of the atrial mass (asterisk) (RV, right ventricle). (B) In gross examination, a 4 × 4 × 2-cm sample of the resected mass is seen to be infiltrated by a dark-brown tumour, which is marked by haemorrhage and necrosis. (C) Haematoxilin–eosin staining revealing an haemorrhagic tumour composed by solid sheets of epithelioid endothelial cells and irregular sinusoidal vascular channels lined by atypical endothelial cells (original ×40). (D) Immunohistochemical staining being strongly positive for CD34, which confirmed the vascular nature of the tumour (original ×4).

had transthoracic echocardiograms available.[3] Transthoracic echocardiogram as the initial diagnostic test had 75% sensitivity for visualizing PCA. Tumour extension into the pericardium was common and pericardial effusion was present in 15 patients (88%). Left ventricular ejection fraction was preserved in 16 patients (94%), and right ventricular function was mildly reduced in two patients at initial presentation. Tricuspid valve obstruction was found in three patients. The absence of a stalk was a universal finding that may help distinguish angiosarcoma from benign, primarily pedunculated tumours.

Appropriate evidence-based treatment guidelines have not been established due to the rarity of the PCA.[4] The preferred treatment of PCA is complete resection[5] and only patients who undergo complete excision have the possibility of cure.[1] However, owing to the frequently extensive nature of the tumour at the time of diagnosis, it is often impossible. It has been suggested that patients in whom complete tumour resection is not indicated or possible can undergo palliative tumour debulking for relevant or rapidly progressive symptoms.[1] Then again, the real benefit of adding adjunctive chemotherapy and/or radiation is still under investigation.[2]

The outcome for PCA is generally poor, with median survival being less than 1 year.[1] However, it has been reported that patients with cardiac angiosarcoma who undergo primary tumour resection have improved median overall survival compared with patients whose tumours remain in situ (17 vs. 5 months in one series[6]). Death usually results from myocardial infiltration, obstruction of flow, cardiac tamponade, and/or distant metastases.[4]

To sum up, we propose the following statements as learning points regarding PCA:

- Cardiac angiosarcoma is the most common primary malignant tumour of the heart. However, cardiac metastases are by far the most frequent cardiac neoplasms.
- PCA typically originates in the right atrium and cardiac tamponade may be one of its first manifestations.
- PCA prognosis is universally poor (median survival <1 year), since widespread metastases are present in most patients at the time of diagnosis.

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. Butany J, Nair V, Naseemuddin A, Nair GM, Catton C, Yau T. Cardiac tumours: diagnosis and management. *Lancet Oncol* 2005;**6**:219–228.
2. Patel SD, Peterson A, Bartczak A, Lee S, Chojnowski S, Gajewski P, Loukas M. Primary cardiac angiosarcoma—a review. *Med Sci Monit* 2014;**20**:103–109.
3. Kupsky DF, Newman DB, Kumar G, Maleszewski JJ, Edwards WD, Klarich KW. Echocardiographic features of cardiac angiosarcomas: the Mayo Clinic experience (1976-2013). *Echocardiography* 2016;**33**:186–192.
4. Bouma W, Lexis CP, Willems TP, Suurmeijer A, van der Horst I, Ebels T, Mariani MA. Successful surgical excision of primary right atrial angiosarcoma. *J Cardiothorac Surg* 2011;**6**:47.
5. Vander Salm TJ. Unusual primary tumors of the heart. *Semin Thorac Cardiovasc Surg* 2000;**12**:89–100.
6. Look Hong NJ, Pandalai PK, Hornick JL, Shekar PS, Harmon DC, Chen Y-L, Butrynski JE, Baldini EH, Raut CP. Cardiac angiosarcoma management and outcomes: 20-year single-institution experience. *Ann Surg Oncol* 2012;**19**:2707–2715.