

# Diagnostic approach to a cardiac mass: a case report of misdiagnosed cardiac synovial sarcoma

Safia Ouarrak <sup>1\*</sup>, Zakaria El Ouali <sup>2</sup>, Asmaa Elkebir<sup>3</sup>, Kawthar Mounna <sup>3</sup>, Mehdi Karkouri <sup>3</sup>, Leila Azzouzi<sup>1</sup>, and Rachida Habbal<sup>1</sup>

<sup>1</sup>Department of Cardiology, University Hospital of Ibn Rochd, Casablanca, Morocco; <sup>2</sup>Department of Rheumatology, University Hospital of Ibn Rochd, Casablanca, Morocco; and <sup>3</sup>Department of Anatomical Pathology, University Hospital of Ibn Rochd, Casablanca, Morocco

Received 27 July 2020; first decision 2 September 2020; accepted 15 January 2021

## Background

Cardiac synovial sarcoma (CSS) is an extremely rare malignant tumour with a poor prognosis. We report the case of a 31-year-old woman who presented with a CSS in the right atrium and was initially misdiagnosed with a tuberculoma. The aim of this article is to focus on the importance of having broad differential diagnoses including rare entities.

## Case summary

A 31-year-old White woman, with a close contact with a relative having pulmonary tuberculosis, presented to the emergency unit with severe dyspnoea. Chest radiography and echocardiography showed a large pericardial effusion with a mass in the right atrium. Pericardiocentesis removed bloody exudative fluid with adenosine desaminase at 17 UI/L and no malignant cells in the cytological study. Cardiac magnetic resonance revealed a tuberculoma of the right atrium. Intraoperatively, the mass was only biopsied because of the local invasion. Histological study concluded to a CSS. The patient died 3 days later.

## Discussion

This case highlights the importance of having broad differential diagnoses including rare entities. Histology was the key investigation for the diagnosis of CSS which has no clinical nor laboratory or imaging pathognomonic signs.

## Keywords

Cardiac synovial sarcoma • Tuberculoma • Pericardial effusion • Cardiac mass • Case report

## Learning points

- The diagnostic strategy of cardiac masses must include a wide range of differential diagnoses including rare entities such as malignant cardiac tumours.
- Therefore, tumours of neoplastic origin cannot be definitely excluded in the absence of malignant cells in the pericardial fluid.
- Positive levels of adenosine desaminase are not pathognomonic of tuberculosis and are also found in cardiac sarcomas.

## Primary specialties involved other than cardiology

Anatomical pathology, Oncology

## Introduction

Cardiac synovial sarcoma (CSS) is a rare entity of cardiac masses, accounting for less than 5% of primary cardiac sarcomas.<sup>1</sup> It usually

\* Corresponding author. Tel: +212 671 139513, Email: [safia.ouarrak@gmail.com](mailto:safia.ouarrak@gmail.com)

Handling Editor: Parham Eshtehardi

Peer-reviewers: Alberto Bouzas-Mosquera and Nikolaos Bonaros

Compliance Editor: Edwina McNaughton

Supplementary Material Editor: Nida Ahmed

© The Author(s) 2021. 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](mailto:journals.permissions@oup.com)

affects children and young adults, with non-specific features, and is associated with a poor prognosis.<sup>2</sup> We present the case of a 31-year-old woman who presented with a CSS in the right atrium and was initially misdiagnosed with a tuberculoma. The aim of this article is to focus on the importance of having broad differential diagnosis including rare entities.

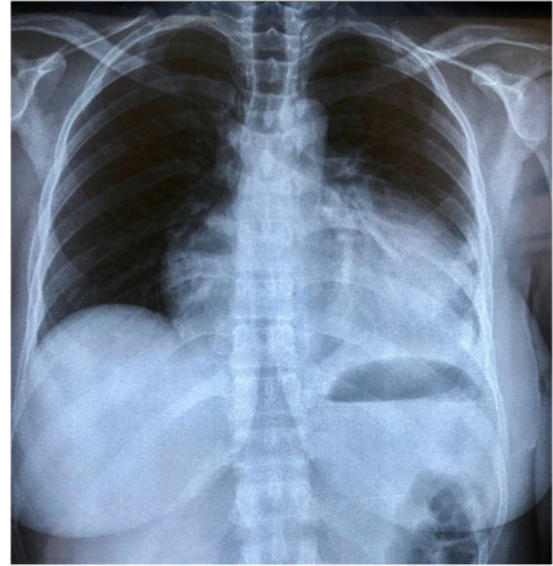
## Timeline

Time	Event
3 months prior to admission	First symptoms: asthenia, weight loss of 8 kg in 3 months, anorexia
10 days prior to admission	Onset of dyspnoea and general fatigue
Admission day (Day 1)	Clinical symptoms: severe dyspnoea (New York Heart Association class IV) Chest X-ray: striking cardiomegaly Electrocardiogram: sinus tachycardia and negative T waves diffusely Echocardiogram: abundant pericardial effusion with a mass in the right atrium Pericardiocentesis: removal of 1 L of bloody pericardial fluid
Day 3	Cardiac magnetic resonance: solid mass of the right atrium suggesting a tuberculoma
Day 4	Admission to the operating room: surgical biopsy
Day 7	Death of the patient

## Case presentation

A 31-year-old White woman, with no particular medical history, presented to the emergency unit with severe dyspnoea and general fatigue evolving for 2 weeks. She was living with her brother who was receiving treatment for pulmonary tuberculosis (TB). She was not receiving prophylaxis, in accordance with the national management protocol of TB contacts. She reported anorexia, asthenia, and weight loss of 8 kg in a period of 3 months prior to admission. On examination, blood pressure was 100/64 mmHg, heart rate was 130 b.p.m, and oxygen saturation was 94%. There were muffled heart sounds without murmur. Her jugular venous pressure was normal, her chest was clear, and there was no peripheral oedema.

Chest X-ray showed a striking cardiomegaly with a cardiothoracic ratio of 0.7 (Figure 1). Electrocardiogram showed a sinus tachycardia with negative T waves diffusely (Figure 2). Echocardiogram revealed a large pericardial effusion, with a mass in the right atrium measuring 27 × 35 mm. The left ventricular ejection fraction was normal (65%), and the inferior vena cava (IVC) was compliant and measuring



**Figure 1** Chest X-ray showing cardiomegaly.

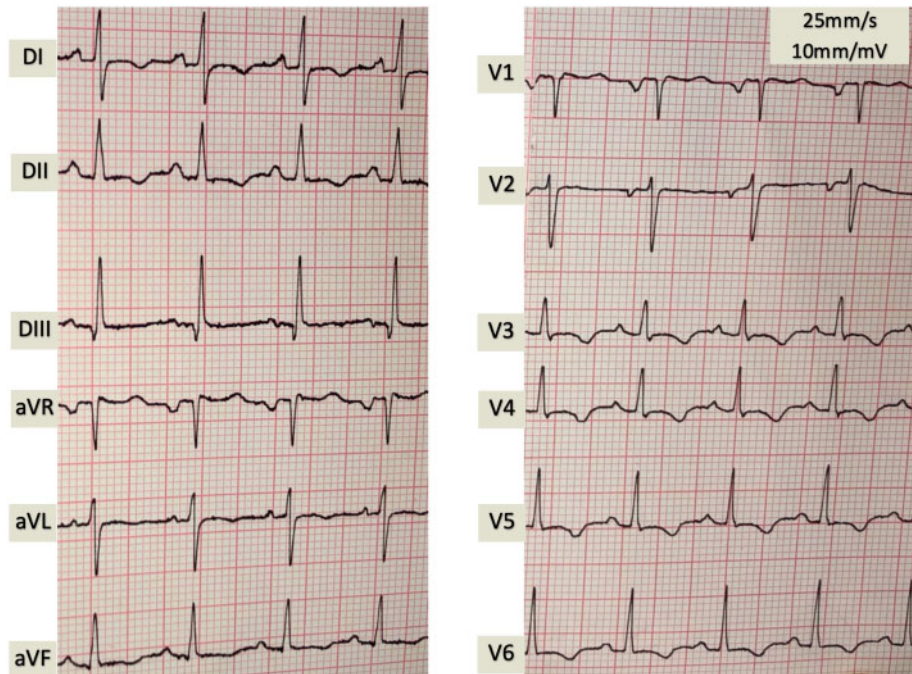
18 mm. The hemogram was normal, the C-reactive protein was elevated (24.5 mg/L, normal range < 5.0). The antinuclear antibodies and rheumatoid factor were negative.

Pericardiocentesis removed 1 L of bloody exudative and lymphocyte-predominant fluid with a protein rate of 66 g/L. Adenosine deaminase (ADA) in the pericardial fluid was at 17 UI/L (normal range < 50 UI/L). No malignant cells were found on cytological study. Cardiac magnetic resonance (CMR) confirmed the presence of a solid mass, measuring 39 × 28.5 mm, with lobulated contours, immobile and fixed to the wall opposite the emergence of the IVC, with no signs of local infiltration or invasion of vascular structures (Figure 3A and B, and Video 1). We thought that the most probable diagnosis was a tuberculoma, considering the patient's exposure. The patient was admitted to the operating room. Intraoperatively, there were multiple masses and vesicles on the heart, pericardium, and large vessels. The frozen-examination confirmed the neoplastic origin, and no further surgery was attempted. Ultimately, histopathology findings were consistent with a monophasic CSS (Figure 4A–D). The patient couldn't have a translocation investigation because she died 3 days after surgery secondary to tamponade.

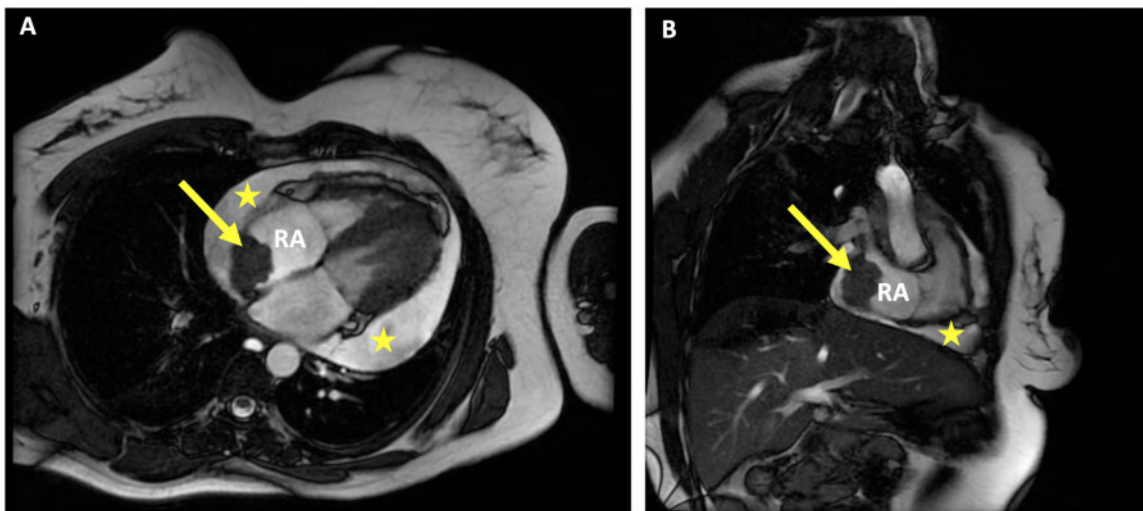
## Discussion

Cardiac masses represent a heterogeneous group of tumoural and non-tumoural masses. Usually, these masses represent thrombi or vegetations often occurring in a particular clinical context.<sup>3</sup> Based on the clinical presentation, laboratory and imaging findings, our case was a diagnostic challenge as she was initially misdiagnosed with tuberculoma.

On the one hand, most cases of cardiac tuberculomas are clinically asymptomatic, or may less commonly present with non-specific signs



**Figure 2** Electrocardiogram showing sinus tachycardia with negative T waves diffusely.



**Figure 3** (A) Cardiac magnetic resonance imaging in axial T2 FIESTA showing an intra-cavitary mass of the right atrium (yellow arrow) associated with a pericardial effusion (yellow stars). (B) Cardiac magnetic resonance imaging in sagittal T2 FIESTA with the same findings. RA, right atrium.

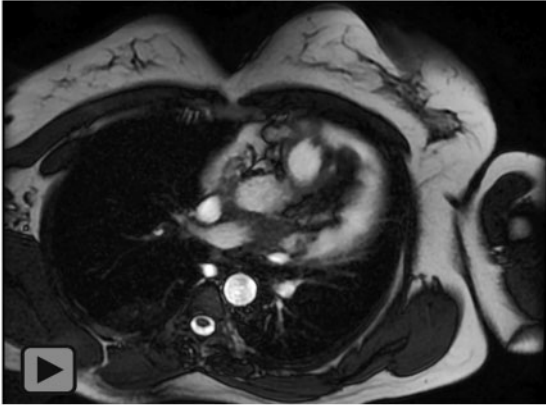
such as congestive heart failure or arrhythmia. The diagnosis is supported by the presence of exposure or history of TB.<sup>4</sup> This was the main argument that made us suspect TB at first.

On the other hand, cardiac tumours are extremely rare conditions, with a frequency ranging from 0.001% to 0.030% in which malignant tumours account only for 20%.<sup>5</sup> CSS are even more

uncommon, with about only 60 cases reported in the literature.<sup>6</sup> They represent less than 5% of all primary cardiac sarcomas, involving either the pericardium or cardiac cavities.<sup>1</sup> They usually affect males in the first decades<sup>2,7</sup> and are generally located in the pericardium.<sup>1</sup> Our case was a 31-year-old female with a CSS located in the right atrium. Clinical signs are non-specific: dyspnoea (68%), chest pain

(26%), cough (22%), fatigue (12%), and weight loss (6%).<sup>6,8</sup> Our patient presented dyspnoea, fatigue, and weight loss 3 months prior to admission.

According to a recent study, the most common aetiologies of pericardial effusion were malignancy (39%) and TB (27%).<sup>9</sup> The



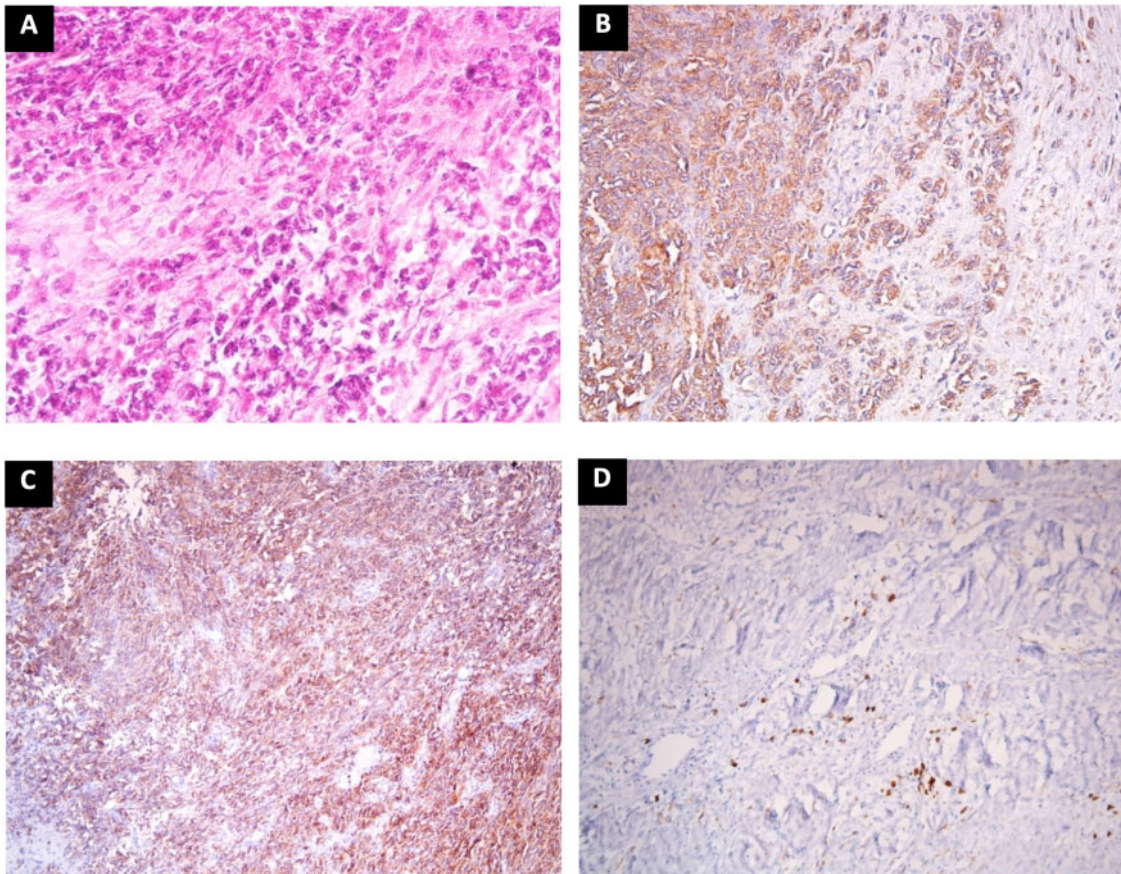
**Video 1** Dynamic cardiac MRI showing the mass in the right atrium associated with a pericardial effusion.

pericardiocentesis removed bloody exudative fluid with no malignant cells, supporting the hypothesis of TB. However, cancer cells might be absent in 25% of malignant pericardial effusions.<sup>9</sup> Consequently, a neoplastic origin cannot be definitely excluded in case of malignant cells absence in the pericardial fluid.

Moreover, ADA test in the pericardial fluid is useful in the diagnosis of TB. In a case-control study of patients with pericardial TB effusion, the cut-off value for the diagnosis of TB was 40 UI/L, with a specificity of 72% and sensitivity of 89%.<sup>10</sup> In our case, ADA level was at 17 UI/L. However, ADA can also be elevated in malignant conditions: in the study of Ma *et al.*,<sup>9</sup> there was no significant difference in the levels of ADA between the TB group ( $35.96 \pm 27.05$ ) and the malignant group ( $41.61 \pm 37.52$ ). Therefore, while malignancies can be associated with a positive ADA, negative ADA has been reported in patients with TB.<sup>11</sup>

Imaging investigations are also non-specific in differentiating between a tuberculoma and a CSS.<sup>6</sup> X-rays generally show a cardiomegaly, echocardiography detects the mass, and finally, CMR assesses its extension with better resolution.<sup>6,8</sup>

The key investigations in the diagnosis and subtype of CSS are the histology, immunochemistry, and genetic analysis.<sup>6</sup> Despite the name of the tumour, origin cells are not synovial cells, and the histogenesis is still debated.<sup>12</sup> Histologically, the tumour is occurring in the



**Figure 4** Histological sections,  $\times 40$  magnification: (A) haematoxylin and eosin stain showing spindle cells proliferation. Immunohistochemistry showing the diffuse expression of CD99 (B) and Bcl-2 (C), and focal expression of the epithelial membrane antigen (D).

monophasic or biphasic pattern, consisting of epithelial and/or spindle cells components.<sup>8</sup> Our patient was diagnosed with a monophasic CSS.

Guidelines for the treatment strategy are not established yet, but the gold standard is the complete surgical resection.<sup>7</sup> Our patient's mass could not be resected because of the local invasion. The poor prognosis is a well-known feature of this disease, with a median overall survival of 27 months, influenced by age and chemotherapy.<sup>6,8</sup>

There were deficiencies in diagnostic approach and management of this patient. The initial diagnosis was misled by the patient's exposure to TB. Therefore, complete diagnostic workup for an intracardiac mass was not done. Some imaging procedures could have been useful to provide more detailed information about the degree of vascularization of the mass, such as coronary angiography or cardiac computed tomography with contrast. Transoesophageal echocardiography may have given some clues on the connection to the pericardium, the epicardium or the endocardium. Also, the performance of a body positron emission tomography would have been useful to detect additional masses or suspicious lymph nodes.

## Conclusion

This case of CSS, misdiagnosed with a tuberculoma, highlights the importance of having broad differential diagnoses including rare entities. Histology was the key investigation for the diagnosis of CSS, which has no clinical nor laboratory or imaging pathognomonic signs.

## Lead author biography



Safia Ouarrak was born in Morocco. She graduated from the Faculty of Medicine and Pharmacy of Casablanca. She is a Cardiology Resident in the University Hospital of Ibn Rochd, Casablanca. Her areas of interest are echocardiography and cardiovascular imaging.

## Supplementary material

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

**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

**Consent:** The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** none declared.

**Funding:** none declared.

## References

- Coli A, Cassano A, Novello M, Ranelletti FO, Lauriola L. Primary cardiac synovial sarcoma: a review correlating outcomes with surgery and adjuvant therapy. *J Card Surg* 2019;**34**:1321–1327.
- Fletcher CDM, Bridge JA, Hogendoorn PCW, Mertens F. *WHO Classification of Tumours of Soft Tissue and Bone*. 4th edn. World Health Organization, p213–215.
- Bruce CJ. Cardiac tumours: diagnosis and management. *Heart* 2011;**97**:151–160.
- Jagia P, Gulati GS, Sharma S, Goyal NK, Gaikwad S, Saxena A. MRI features of tuberculoma of the right atrial myocardium. *Pediatr Radiol* 2004;**34**:904–907.
- Centofanti P, Di Rosa E, Deorsola L, Actis Dato GM, Patanè F, La Torre M et al. Primary cardiac tumors: early and late results of surgical treatment in 91 patients. *Ann Thorac Surg* 1999;**68**:1236–1241.
- Wang J-G, Li N-N. Primary cardiac synovial sarcoma. *Ann Thorac Surg* 2013;**95**:2202–2209.
- Coli A, Chiariello GA, Novello M, Colizzi C, Massetti M. Treatment of cardiac synovial sarcoma: experience of two cases. *J Cardiothorac Surg* 2018;**13**:84.
- Duran-Moreno J, Kampoli K, Kapetanakis EI, Mademli M, Koufopoulos N, Foukas PG et al. Pericardial synovial sarcoma: case report, literature review and pooled analysis. *In Vivo* 2019;**33**:1531.
- Ma W, Liu J, Zeng Y, Chen S, Zheng Y, Ye S et al. Causes of moderate to large pericardial effusion requiring pericardiocentesis in 140 Han Chinese patients. *Herz* 2012;**37**:183–187.
- Tuon FF, Silva VD, Almeida GD, Antonangelo L, Ho YL. The usefulness of adenosine deaminase in the diagnosis of tuberculous pericarditis. *Rev Inst Med Trop São Paulo* 2007;**49**:165–170.
- Reuter H, Burgess L, van Vuuren W, Doubell A. Diagnosing tuberculous pericarditis. *QJM* 2006;**99**:827–839.
- Synovial sarcoma [Internet]. Available in: <http://www.pathologyoutlines.com/topic/softtissuesynovialsarc.html> (22 July 2020).