

## Clinical Case Report

## A cardiac mass beyond symptomatic palpitations

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## 1. Case report

A 33-year-old Caucasian smoker female with no relevant past medical history was referred to Cardiology consultation because of persistent palpitations onset, mainly triggered by moderate daily activity. She also complained of light fatigue for usual sport activity. Physical examination was unremarkable. Twelve-lead ECG was normal, and 24-h Holter monitoring revealed the presence of frequent supraventricular arrhythmia with periods of type 2 Mobitz I atrioventricular block.

Chest radiograph (Fig. 1A) showed an abnormal prominence on the right heart border. Transthoracic echocardiogram (Fig. 1B, clip S1) revealed the presence of well-delimited, oval-shaped, apparently intrapericardial mass adjacent to the free wall of right atrium and the superior vena cava. Cardiac magnetic resonance (CMR) (Fig. 2) confirmed the presence of a huge paracardiac mass. Sequences for tissue characterization suggested the presence of homogeneous fluid content with high protein content (T1 isointensity and T2 hyperintensity). There was neither pleural nor pericardial effusion. This mass was not perfused, and there was no delayed enhancement, except for its borders, suggesting the presence of a fibrotic capsule.

Owing to possible related symptoms and evidence for a complex pericardial mass/cyst, the patient was proposed for surgical resection.

*Abbreviations:* RA, right atrial; CT, computed tomography; ECG, electrocardiography; CMR, cardiac magnetic resonance.

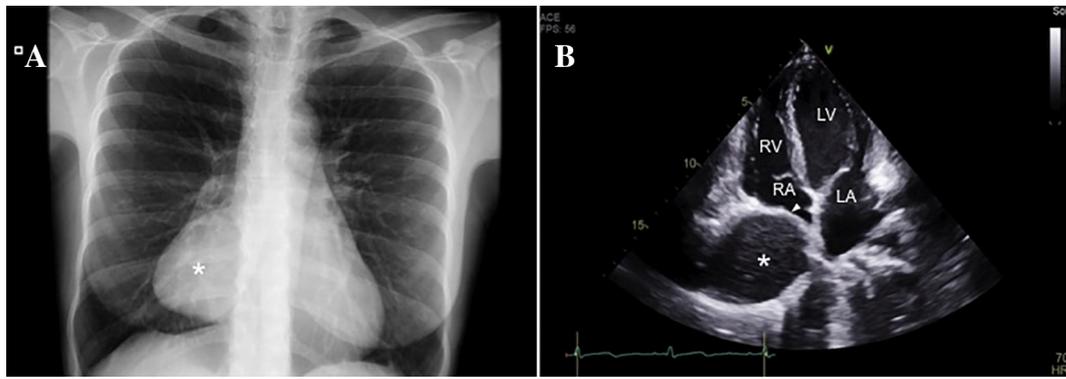
\* Corresponding author.

Cardiac computed tomography (CT) (Fig. 3) excluded coronary artery disease, and complete surgical excision was accomplished through video-assisted mediastinoscopy under general anaesthesia and alternate selected single-lung ventilation. The mass was easily detached from adjacent structures, being completely capsulated with no vascular invasion.

The specimen was sent for anatomopathologic study. Macroscopy showed a 7×6-cm cystic nodule, with a thin elastic-firm wall, an irregular inner surface, and a content of yellow-whitish granular material with scarce hairs (Fig. 4A and B). Microscopic examination of formalin-fixed, paraffin-embedded, and hematoxylin–eosin-stained samples complemented with histochemistry [Masson's Trichrome, for fibrous tissue/collagen] and immunohistochemistry [cytokeratin (MNF116), for epithelial tissue; chromogranin A, for neuroendocrine cell; calretinin, for mesothelial cells—using Roche–Ventana Benchmark Ultra apparatus and protocols] revealed an inner layer of stratified keratinized squamous epithelium with cutaneous adnexae (ectoderm origin), exocrine and endocrine pancreatic tissue (endoderm origin) in the cystic wall, and a pericardial layer on the outer surface (Fig. 5A–H). These findings led to the diagnosis of benign, mature, cystic, bigeminal pericardial teratoma. The patient had an uneventful recovery, with hospital discharge at the third postoperative day. Clinical follow-up at the first year was unremarkable, without any symptoms.

## 2. Discussion

Primary neoplasms of the heart are extremely rare, with an estimated prevalence of 0.001% to 0.03% [1]. Teratomas are tumors of embryonic origin that contain structures derived from at least two of the three germinal layers [2]. Germ cell tumors are thought to originate from primordial germ cells that fail to complete their migration from the urogenital ridge. Only 3%–7% of all germ cell tumors are extragonadal, the mediastinum being the most frequent extragonadal site [2,3]. Cardiac teratomas account for less than 1% of cardiac tumors in adults, and there have been few reported cases in the literature [4–7]. Ninety percent of the cardiac teratomas have been found in the pericardium and the rest in the myocardium [8]. Usually, they are adjacent to the right atrium and can grow up to several centimeters at the time of diagnosis.

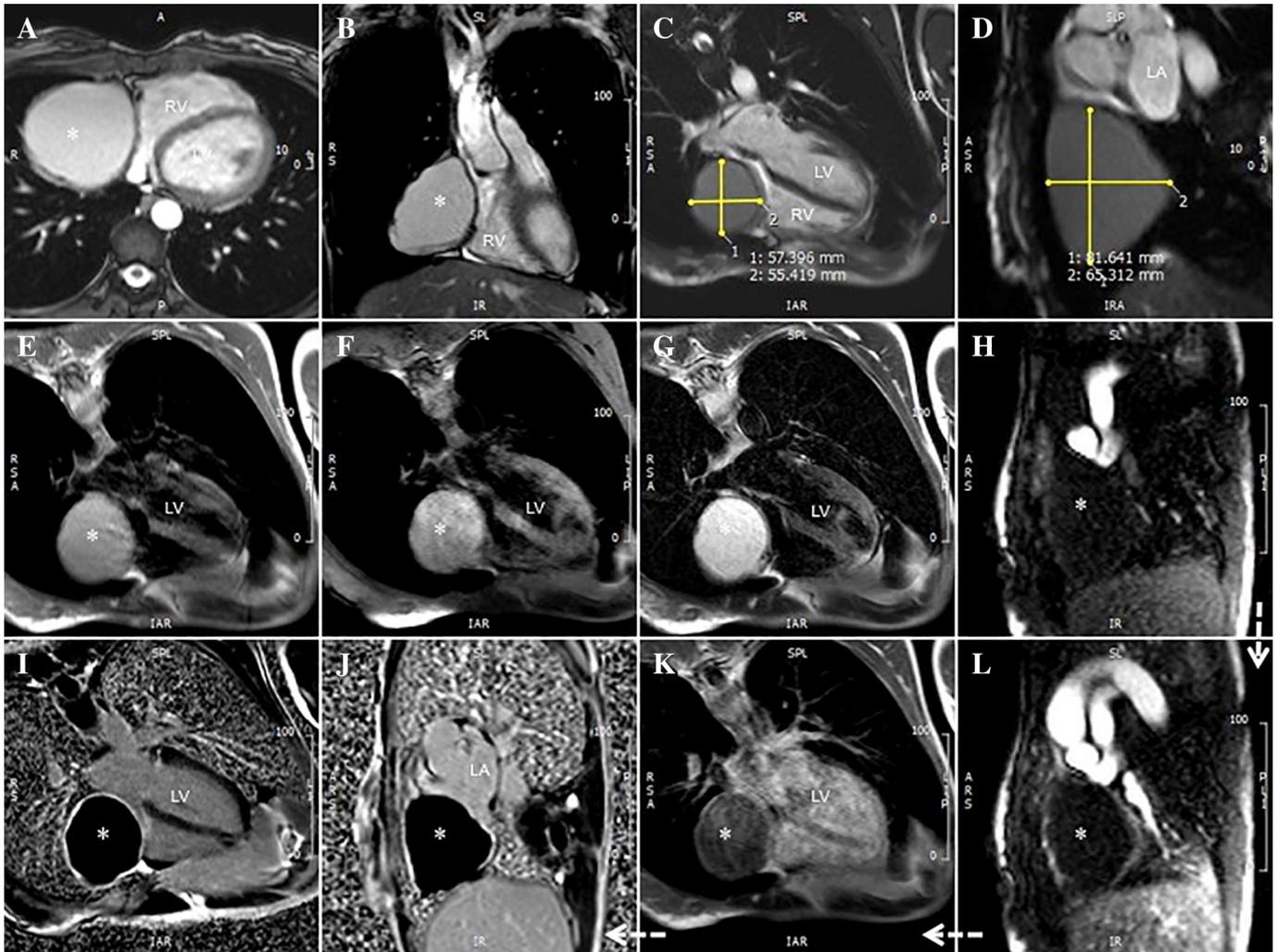


**Fig. 1.** An abnormal mass (\*) was detected in both chest X ray (A) and echocardiography (B). In panel B, there was a well-delimited, capsulated (arrowhead), homogeneous, hypoechoic mass shaping right atrial (RA) contour. LA, left atrium; LV, left ventricle; RV, right ventricle.

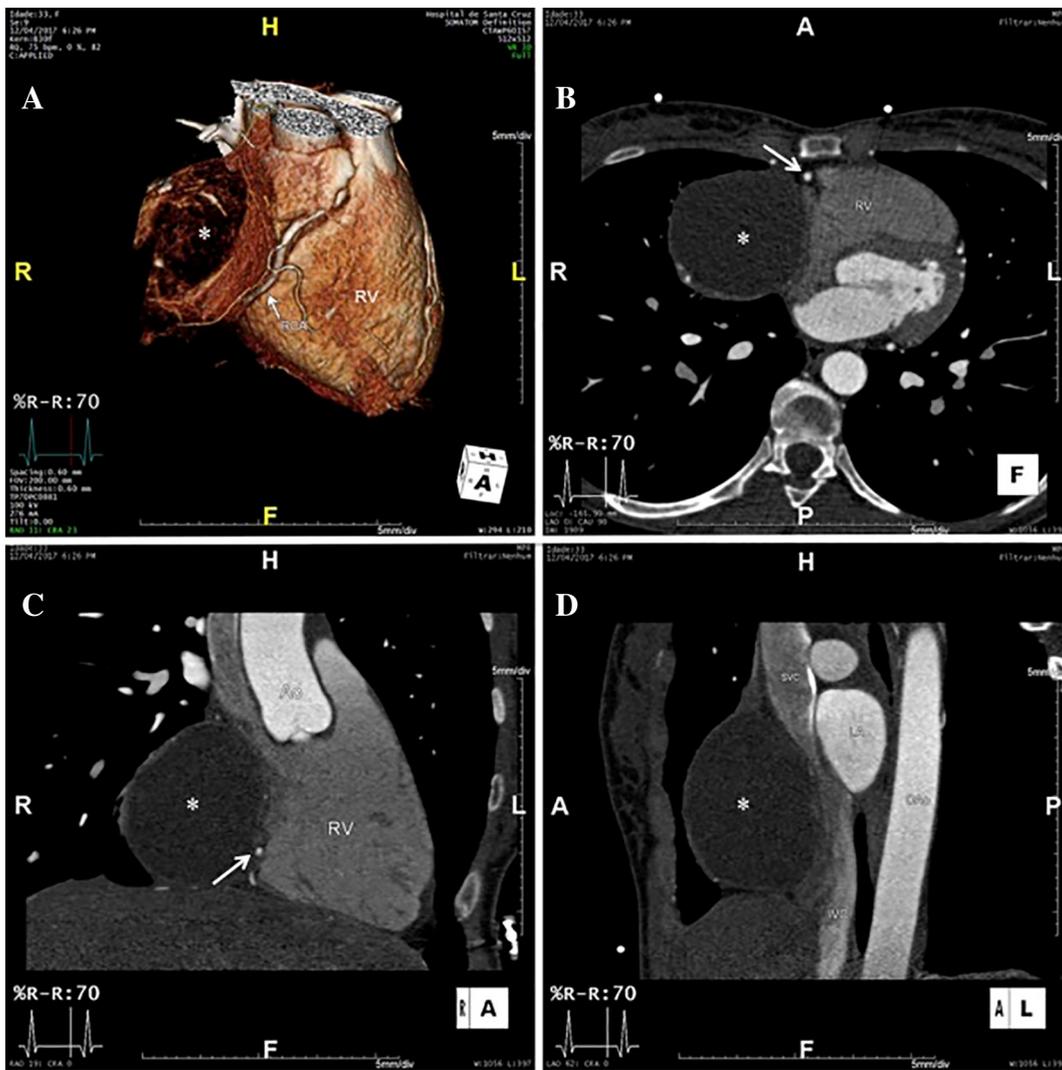
Intrapericardial teratomas are rare tumors in children, and their occurrence in adults is exceptional (<1%) [4,9]. The peak incidence at presentation in adults is in the second and third decades of life, with a slight female predominance (1.6:1) [10].

Typically, teratomas are diagnosed in early childhood due to early occurrence of symptoms. These are mainly related to mass location

and mechanical effects, including chest pain, cough, dyspnea, bronchial obstruction with recurrent respiratory infections, Horner syndrome, superior vena cava syndrome, and rarely palpitations. Erosion into an adjacent bronchus may rarely lead to trichoptysis (expectoration of hair). Palpitations, supraventricular arrhythmias, are generally due to compressive effects of large lesions on atrial walls [10].



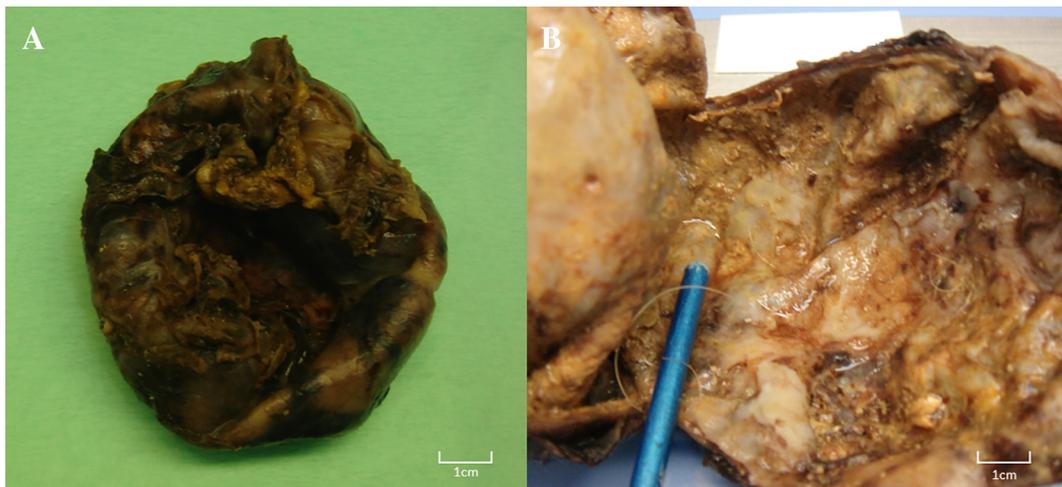
**Fig. 2.** Cardiac magnetic resonance. Panels A to L depict comprehensive protocol sequences addressing cardiac mass (\*) study. (A–B) Axial and coronal views with similar topographic relations as assessed by echocardiography. (C–D) Cine SSFP sequences with a homogenous hypointense mass. (E) T1-weighted sequence. (F) Fat-suppressed T1-weighted sequence. (G) T2-weighted sequence. Arrows connecting panels H, L, K, and J show the sequential protocol from mass perfusion (H, L), postcontrast T1 weighing (K), and delayed enhancement (I, J). Even considering tissue characteristics, this single mass is homogeneous, has a fibrotic capsule, is devoid of perfusion, and has no associated pleural or pericardial effusion. Taken together, these features support a benign behavior. LA, left atrium; LV, left ventricle; RV, right ventricle.



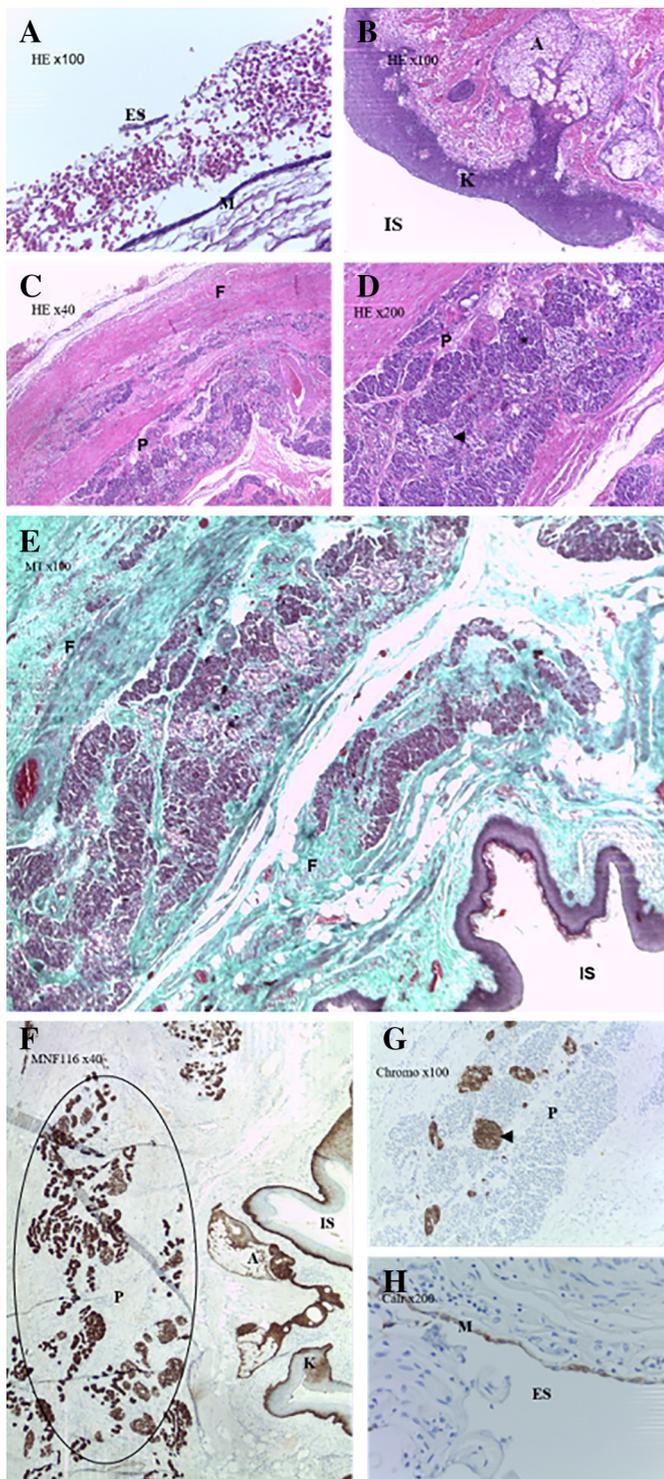
**Fig. 3.** Cardiac CT (A: volume rendering reconstruction; B, C, D: axial, coronal, and sagittal views). This was performed for the exclusion of coronary artery disease before mass excision. It revealed the presence of a homogenous well-circumscribed hypodense mass (\*), respecting right coronary artery path (white arrow) along the atrioventricular groove. Even so, both superior vena cava and inferior vena cava are shaped by this huge mass. Ao, ascending aorta; DAo, descending aorta; IVC, inferior vena cava; LA, left atrium; RV, right ventricle; SVC, superior vena cava.

Multimodality imaging is a routine procedure when assessing both mediastinal and pericardial masses. Conventional radiology is useful but may have limited sensitivity for small lesions, as CT, for the detection of

calcified material such as mature teeth and bone. Echocardiography is commonly one of the first imaging techniques, not only providing pericardial mass characterization but also and more importantly allowing the



**Fig. 4.** Macroscopic view of the cystic tumor. Closed (A). Opened, displaying the wall, inner surface, and scarce residual content, namely, keratin and hairs (B).



**Fig. 5.** Microscopic features, including additional histochemical and immunohistochemical characterization, are shown (A–H). The tumor external surface (ES) is covered by pericardial, calretinin-positive, mesothelial cells (M). The wall presents pancreatic tissue (P) embedded in fibrosis (F). The pancreatic elements—endocrine (◄) and exocrine (\*)—are cyokeratin (MNFI16) positive, and the endocrine ones—Langerhans Islands—are also positive for chromogranin. The inner surface (IS) is covered by MNFI16-positive stratified keratinized squamous epithelium (K) and cutaneous adnexae (A).

identification of possible hemodynamic interference due to specific topographic relations with great vessels. Both CT and CMR are important for additional diagnostic information, mainly related to the suspicion of

malignancy. CMR tissue characterization specifically enables the distinction from the commonest simple pericardial fluid cyst, which is frequently clinically silent, not needing surgical excision. CT is indicated for staging when in the presence of a malignant lesion.

The diagnosis of pericardial teratoma is made by pathologic evaluation. Gross pathology may allow definitive diagnosis from the presence of distinct mature tissue components. However, histology is crucial for maturity assessment, as mature teratomas (more than 50% of the tumor has well-differentiated elements) are typically benign. Despite this, both immature and mature types may show malignant transformation, resembling a non-germ-cell tumor somatic-type cancer. So, whenever detected, these lesions should always be excised owing to a theoretical risk of malignant transformation (<2% incidence of brain, lung, and bone metastasis) [11,12]. Additionally, as composed of mature elements, teratomas are relative insensitive to both chemo- and radiotherapy, limiting treatment to surgical excision. Moreover, symptoms such as those related to compression of adjacent structures, arrhythmias, or cyst infection should also lead to surgical excision.

The case reported is relevant not only for the late diagnosis, taking place in adulthood, but also due to the diverse findings in both imaging and pathology studies, confirming the diagnosis. Each imaging technique provided complementary and useful data, yet cardiac magnetic resonance was particularly important, raising the suspicion of a complex pericardial cyst. Immunohistochemistry was performed to highlight the structures present in the lesion, despite not being essential for the diagnosis according to diverse morphological expression coming from distinct germinal layers.

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.carpath.2018.10.007>.

#### Conflicts of interest

None.

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