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CASE REPORT

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CMR detects extensive intracavitary thrombi as solitary clinical presentation of Antiphospholipid Syndrome: A case report

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Abstract

Intracavitary thrombi are an important differential diagnosis of cardiac masses. Cardiac magnetic resonance imaging (CMR) allows their non-invasive characterization. This case highlights extensive cardiac thrombi detected by CMR as solitary presentation of antiphospholipid syndrome.

KEYWORDS

antiphospholipid syndrome, cardiac thrombi, CMR

1 | HISTORY OF PRESENTATION

A 51-year-old female patient was admitted to our hospital due to acute onset atrial fibrillation (AF) with a heart rate up to 150 beats per minute. The patient had woken up early in the morning with palpitations and dyspnea and called for emergency help. The initial ECG documented AF. Cardioversion was planned as the patient was highly symptomatic and blood pressure relatively low. Transesophageal echocardiography (TOE) revealed three unclear LV masses, one of them adjacent to the posterior mitral leaflet, a tiny one on the anterior leaflet and a 30×20 mm measuring intracavity mass attached to the basal septum (Figure 1). Therapeutic anticoagulation with heparin was started (with the transition to Vitamin K antagonists), and pharmaceutical frequency control pursued.

2 | PAST MEDICAL HISTORY

The patient reported a weight loss of about 10 kg and night sweats during the last 5 months but was otherwise healthy with no underlying diseases except hypercholesterolemia and smoking history.

3 | DIFFERENTIAL DIAGNOSIS

Cardiac masses are rare entities most commonly detected incidentally on non-invasive cardiac imaging. Approximately half of these cases are benign masses such as thrombi, fibroma, or lipoma, whereas the other half are either of primary or secondary malignant origin (e.g., sarcoma, rhabdomyoma, lymphoma, and other metastatic

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diseases). Cardiac masses attached to the valvular structures are most frequently fibroelastomas, vegetations, or thrombi.[1,2]

4 | CMR

CMR was performed to further discriminate between a malignant and non-malignant process.[3] CINE imaging using a balanced turbo field echo sequence (BTFE) revealed apical LV hypertrophy consistent with apical HCM. The CINE images revealed three intra-cavitary masses, an extensively furrowed mass (longitudinal extend 50 mm, distal diameters 18 × 15 mm) (Figure 2) and two smaller masses adjacent to the mitral valve (9 × 6 mm and 3 × 6 mm) (Figure 3). There were no wall motion abnormalities detected, and the LV function was normal.

T2-weighted STIR images excluded myocardial edema. The intracavitary masses presented with a discrete hypointense aspect, and T2-weighted images without fat suppression excluded lipoid structures (Figure 4).

All three masses showed no vascular perfusion and no late enhancement in dedicated sequences (Figure 5). The late enhancement scans revealed no signs of a myocardial infiltration or scarring.

Based on the CMR findings, the cardiac masses were classified as benign T2-weighted images, further ruling out lipoma as a non-malignant differential diagnosis. Thus, despite a normal LV function, thrombi were the most likely diagnosis (Figure 6).

5 | FURTHER WORKUP

PET/CT scan excluded extra-cardiac lesions, and cardiac CT excluded coronary artery disease. AS CMR confirmed giant thrombus formations within the LV cavity and excluded both ischemic myocardial damage and impaired cardiac function, an underlying rheumatic disease such as Lupus erythematosus (SLE) was suspected.

Lupus anticoagulant was highly positive with a value of 106.2 s (range 25–35.6 s). Anti-cardiolipin IgM antibodies (53 MPL-u/ml; range 0–20) and anti-cardiolipin IgG antibodies (IgG 22.9 GPL-U/ml, range 0–20) were elevated, Anti-β_2 Glycoprotein I IgG antibodies were marginally increased but still within the reference range (Anti-β_2 Glycoprotein I IgG 18.7 U/mL, range 0–20). Additionally, the antinuclear antibody titer (ANA above 1:10240) and the anti-dsDNA antibodies were highly elevated (1:320).

6 | MANAGEMENT

Due to highly positive ANA and Anti-dsDNA detection, a connection with systemic lupus erythematosus (SLE) was suspected. However, no other disease criteria for SLE

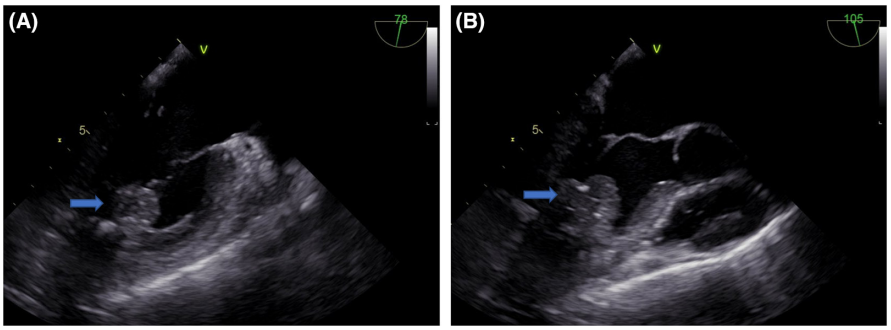

FIGURE 1 A and B: TEE findings: The blue arrow marks the largest of three intracavitary masses

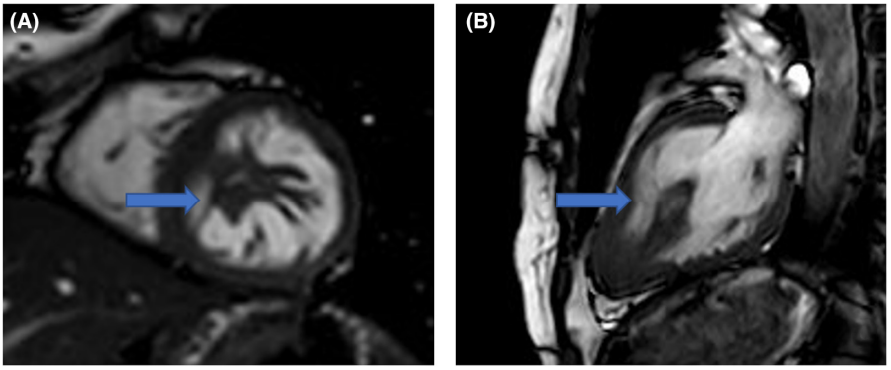

FIGURE 2 cMRI at baseline: CINE images of the extensively furrowed intracavitary mass, marked with a blue arrow in the (A) short axis and (B) 2Chamber view

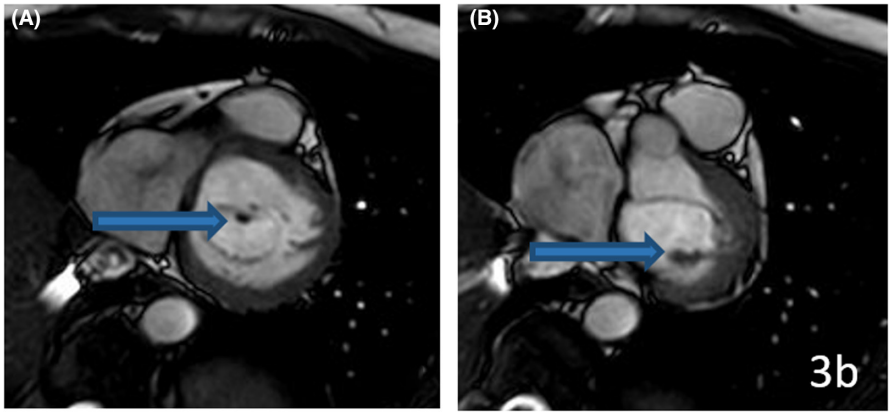

FIGURE 3 cMRI at baseline: The CINE short axis views show two smaller lesions attached to (A) the anterior and (B) the posterior mitral valve leaflet

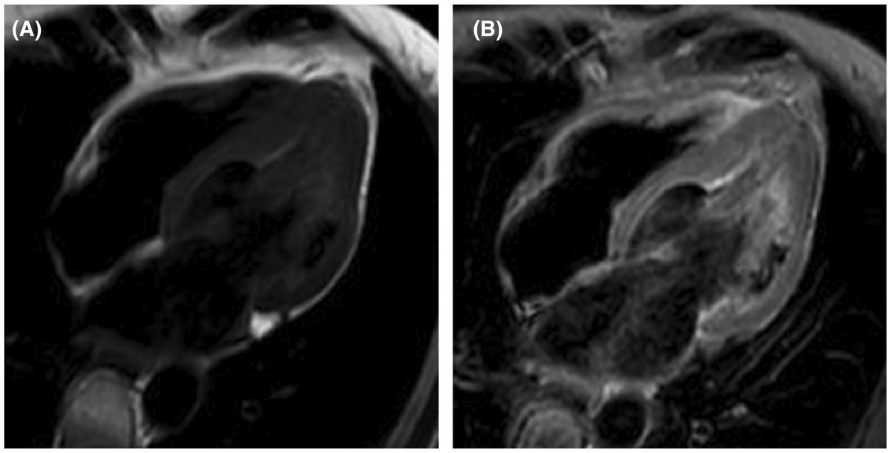

FIGURE 4 cMRI at baseline: T2-weighted four chamber views without fat suppression (A) excluded a lipomatous quality of the intracavitary masses and STIR (B) showed a rather hypointense aspect of the intracavitary mass

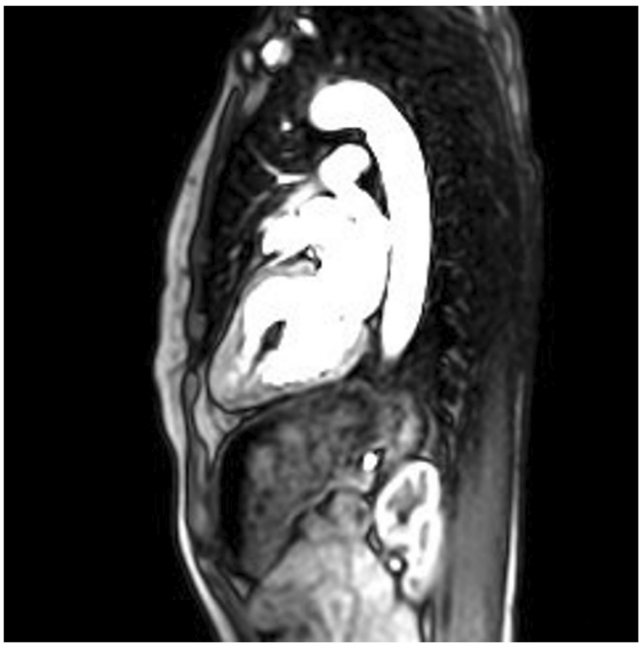

FIGURE 5 cMRI at baseline: First-pass perfusion of a 2Chamber view. The myocardium shows perfusion with a homogenous hyperintense aspect. In contrast, the central mass remains hypointense, excluding vascularity within the mass

according to the European League against Rheumatism (EULAR)/American College of Rheumatology (ACR) classification criteria were detected. Therefore, the patient was suspected of having an incomplete SLE.[4] Accordingly, an immunomodulatory treatment with hydroxychloroquine was started. Therapeutic anticoagulation is intended to be continued indefinitely.

7 | DISCUSSION

CMR allows the proper diagnosis of cardiac thrombi.[3] It is a diagnosis of exclusion, as most malignant entities such as metastases present with high intensity signaling on T2-weighted images, and contrast-enhanced images, whereas for example, fibromas present with hypointense signals on T2-weighted images and very high signal intensity on contrast-enhanced images in CMR. Thrombi show no contrast enhancement and either moderate hyper- or hypo-intense signals on the T2-weighted images.[5,6]

Cardiac intraventricular thrombi are often associated with cardiac dysfunction as seen, in patients with acute myocardial infarction, but also in any other type of chronic heart failure with reduced ejection fraction.[7] As this case

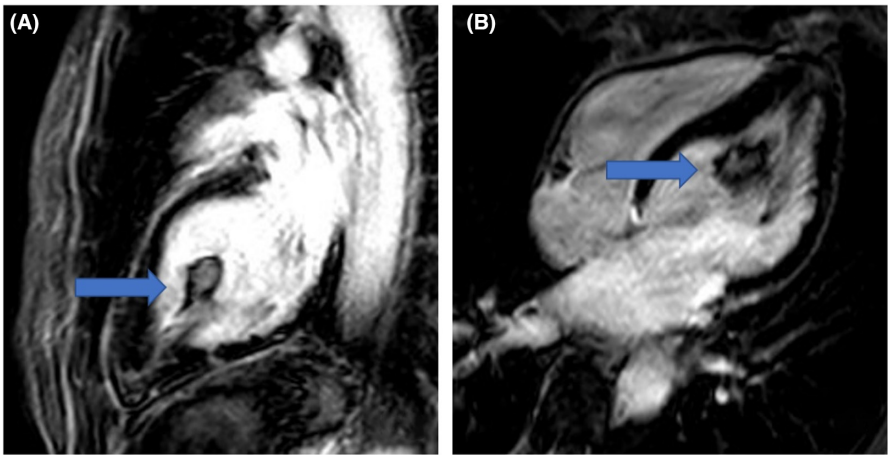

FIGURE 6 MRI at baseline: After administration of contrast agent, the intracavitary mass shows no hyperintense signal in (A) 2 Chamber view and (B) 4 Chamber view (blue arrows) on late enhancement images (TI 240 ms)

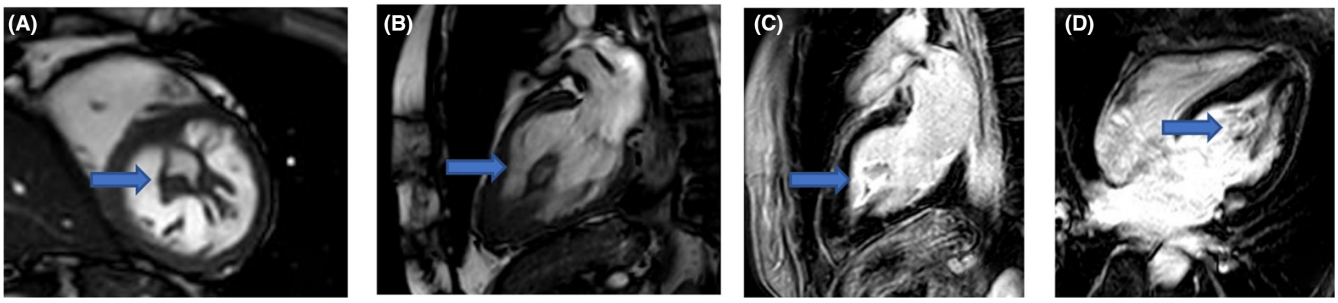

FIGURE 7 cMRI after 12 months. The corresponding views to Figure 2 are shown. The blue arrow marks the decreasing intracavitary thrombus in both CINE images (A, short axis view and B, two chamber view) and LGE images (C, two chamber view and D, four chamber view)

shows, other clinical conditions can also be associated with the development of thrombi. Antiphospholipid syndrome (APS) is a rare disease defined by both clinical signs and laboratory findings.[8] Patients may present with any kind and any severity degree of thrombotic events. Other frequent findings in APS are complications during pregnancy that may result in abortions or premature births. Clinically, APS is often linked with systemic lupus erythematosus. The diagnosis of the latter requires both immunological and clinical findings. Among these, affections of the mucocutaneous, serosal, musculoskeletal and renal systems, as well as neuropsychiatric and hematologic pathologies, are required.[4]

The CMR scans additionally revealed apical hypertrophic cardiomyopathy (HCM). There are a couple of cases reporting on patients with HCM and intraventricular thrombi, however, these cases were associated with apical aneurysms and midventricular obstruction that was both not present in our patient.[9]

8 | FOLLOW-UP

Repeat analysis of laboratory markers after 3 months revealed persistently elevated Lupus anticoagulant with a value of 93.4 s (rage 25–35.6 s), elevated Anti-Cardiolipin IgM and IgG (63.4 MPL-u/ml, Anti-Cardiolipin IgG 27.4 GPL-U/ml, range 0–20), and slightly positive Anti-β_2 Glycoprotein I IgG (18.7 U/ml, range 0–20). Thus, diagnosis of APS could be confirmed.

In repeat cardiac MRI after 12 months, the giant ventricular thrombus formation was still detectable, but much smaller with a maximum distal diameter of 6 mm and less dense in appearance. The two smaller thrombi had dissolved. Left ventricular functional parameters remained stable (Figure 7).

Both anticoagulation and immune-modulating therapy were continued.

9 | CONCLUSIONS

In a patient with unclear intra-cardiac tumors, CMR enabled diagnosis of ventricular thrombus formation as the first clinical sign of an APS associated with atypical SLE. Our case demonstrates that not only vascular but also cardiac thrombosis might be the sole presentation of APS. Of note, at least one specific antibody, either Lupus coagulant, Anti-Cardiolipin antibodies or Anti-β_2 Glycoprotein

I, has to be positive in two separate measurements within 3 months apart to confirm diagnosis of APS. Treatment in most cases consists of life-long anticoagulation.[10]

AUTHOR CONTRIBUTIONS

Theresa Reiter: Conceptualization; investigation; visualization; writing – original draft; writing – review and editing. **Senem Demirbas:** Investigation; writing – review and editing. **Marc Schmalzing:** Investigation; methodology; writing – review and editing. **Wolfram Voelker:** Methodology; writing – review and editing. **Wolfgang Rudolf Bauer:** Methodology; writing – review and editing. **Gülmisal Güder:** Conceptualization; supervision; visualization; writing – original draft; writing – review and editing.

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None.

CONFLICT OF INTEREST

The authors declare that they all have no conflict of interest.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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