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 30×20mm measuring intracavity mass attached to the basal septum (Figure 1). Therapeutic anticoagulation with heparin was started (with the transition to Vitamin K antagonists), and pharmacological 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
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-β2 Glycoprotein I IgG antibodies were marginally increased but still within the reference range (Anti-β2 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...
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. Accordingly, an immunomodulatory treatment with hydroxychloroquine was started. Therapeutic anticoagulation is intended to be continued indefinitely.

**DISCUSSION**

CMR allows the proper diagnosis of cardiac thrombi. 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.

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. As this case...
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. 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.

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.

8 | FOLLOW-UP

Repeat analysis of laboratory markers after 3 months revealed persistently elevated Lupus anticoagulant with a value of 93.4 s (range 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-β₂ 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-β₂ 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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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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REFERENCES
1. Tyebally S, Chen D, Bhattacharyya S, et al. Cardiac tumors. JACC CardioOncol. 2020;2(2):293-311.
2. Dujardin KS, Click RL, Oh JK. The role of intraoperative transesophageal echocardiography in patients undergoing cardiac mass removal. J Am Soc Echocardiogr. 2000;13(12):1080-1083.
3. Fussen S, De Boeck BW, Zellweger MJ, et al. Cardiovascular magnetic resonance imaging for diagnosis and clinical management of suspected cardiac masses and tumours. Eur Heart J. 2011;32(12):1551-1560.
4. Aringer M, Costenbader K, Daikh D, et al. 2019 European league against rheumatism/American college of rheumatology classification criteria for systemic lupus erythematosus. Arthritis Rheumatol. 2019;71(9):1400-1412.
5. Mousavi N, Cheezum MK, Aghayev A, et al. Assessment of cardiac masses by cardiac magnetic resonance imaging: histological correlation and clinical outcomes. J Am Heart Assoc. 2019;8(1):e007829.
6. Pazos-López P, Pozo E, Siqueira ME, et al. Value of CMR for the differential diagnosis of cardiac masses. JACC Cardiovasc Imaging. 2014;7(9):896-905.
7. Lemaître AI, Picard F, Maurin V, Faure M, Dos Santos P, Girerd N. Clinical profile and midterm prognosis of left ventricular thrombus in heart failure. ESC Heart Fail. 2021;8(2):1333-1341.
8. Miyakis S, Lockshin MD, Atsumi T, et al. International consensus statement on an update of the classification criteria for definite antiphospholipid syndrome (APS). J Thromb Haemost. 2006;4(2):295-306.
9. Hughes RK, Knott KD, Malcolmson J, et al. Apical hypertrophic cardiomyopathy: the variant less known. J Am Heart Assoc. 2020;9(5):e015294.
10. Espinosa G, Cervera R. Management of the antiphospholipid syndrome. Auto Immun Highlights. 2010;1(1):15-22.

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