



A Curious Case of Multiple Intracardiac Masses: Antiphospholipid Syndrome Manifesting as Multiple Intracardiac Thrombi

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Abstract

Antiphospholipid syndrome (APS) is a multisystem autoimmune disease characterized by acquired hypercoagulability, recurrent pregnancy loss, and elevated levels of antiphospholipid antibodies. The common cardiovascular manifestations include valvulopathy, coronary artery disease (CAD), myocardial dysfunction, cardiac thrombi, pulmonary thromboembolism, and pulmonary hypertension. Herein we present a case who presented with stroke with incidentally detected multiple cardiac lesions on echocardiography suspicious for mass. Cardiac magnetic resonance (CMR) was able to accurately characterize these lesions as cardiac thrombi, which were subsequently confirmed by endomyocardial biopsy. In this article, the case we discussed, highlights the importance of CMR in accurately characterizing the suspected mass lesion in echocardiography, thus arriving at an accurate diagnosis that changed patient management altogether.

Keywords

- ▶ antiphospholipid antibody syndrome
- ▶ cardiac MRI
- ▶ cardiac thrombus

Introduction

Antiphospholipid syndrome (APS) is a multisystem autoimmune disease that presents with arterial and venous thrombosis and recurrent fetal loss. Cardiovascular involvement is an often underrecognized manifestation of APS, which can contribute to significant morbidity and mortality. Imaging, especially cardiac magnetic resonance (CMR), plays a crucial role in the detection and diagnosis of cardiac involvement. Prompt diagnosis with the commencement of specific therapy is crucial in preventing disease progression and complications. We report a case of a middle-aged gentleman who developed a stroke and pulmonary embolism secondary to intracardiac thrombi. The presence of multiple cardiac masses, characterized by CMR as thrombi, was central in

arriving at a diagnosis of APS. Multiple intracardiac thrombi is a rare cardiac manifestation of APS,¹ and the occurrence of multisystem thromboembolic phenomenon from intracardiac thrombus has been sparsely reported.^{2–5}

Case Report

A 51-year-old male patient, with previous two episodes of lower limb deep vein thrombosis, presented with acute onset of left hemiparesis. MRI brain revealed an acute infarct in the right frontal lobe (→ **Fig. 1A**) with occlusion of the right middle cerebral artery. An echocardiogram revealed multiple intracardiac lesions, with cardioembolic stroke suspected to be the probable cause of the right frontal lobe infarct. CMR done for a detailed evaluation of these cardiac lesions (→ **Figs. 1,2,3**)

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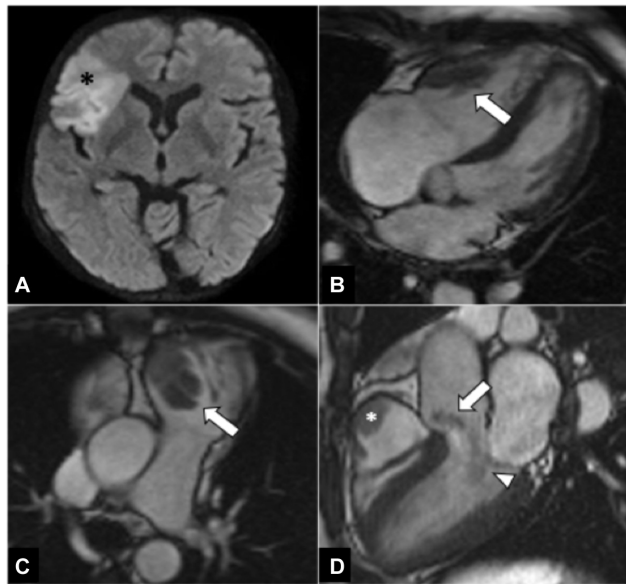


Fig. 1 MRI diffusion-weighted image (A) showing acute infarct (*) in the right frontal lobe. Four chamber (B), axial (C), and three-chamber (D) SSFP images showing lesions in the RV, RV outflow tract, and aortic valve leaflet (arrows in B, C, and D, respectively), with associated aortic regurgitation (arrowhead in D). The lesion in the RV outflow tract (asterisk in D) is also seen in the three-chamber (D) SSFP image.

showed multiple intracardiac lesions in the right atrium (RA), and right ventricle (RV), and a small nodular lesion was seen attached to the free edge of the right aortic valve leaflet on the aortic surface (►Fig. 1B–D). The lesions were hyperintense on T1 weighted image (►Fig. 2A), nonenhancing at perfusion imaging (►Fig. 2B), and showed no enhancement except for peripheral enhancement at the attachment sites with cardiac chambers on late gadolinium enhancement (LGE) images.

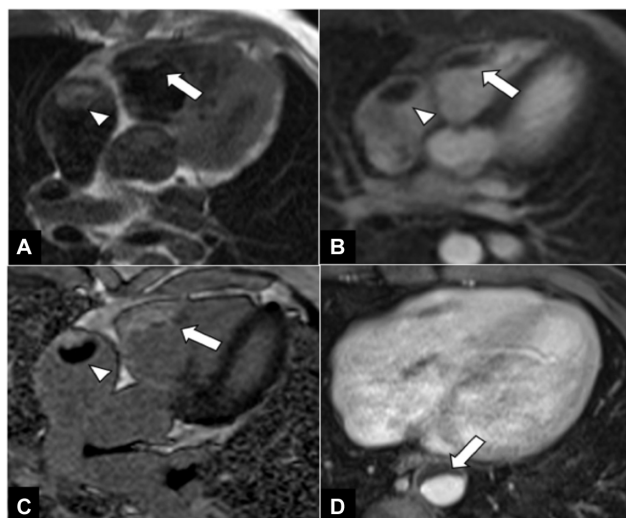


Fig. 2 Axial SSFP T1-weighted (A) image showing hyperintense lesion in the RA (arrowheads) and RV (arrows). Four-chamber perfusion image (B) shows the hypo-perfused lesion (arrows). Late gadolinium enhanced (LGE) with high inversion time (TI) of 600 ms (C) showing non-enhancing lesions with peripheral enhancement in the RA (arrowheads) and RV (arrows) at the attachment sites suggestive of chronic thrombus. Post-contrast T1-weighted VIBE axial image (D) showing eccentric intra-luminal, non-enhancing hypointense lesion (arrow) along the anterior wall of the mid-descending thoracic aorta.



Fig. 3 Follow-up CT pulmonary angiogram done 1 month after MRI showing an eccentric filling defect in the lower lobe segmental branch of the right pulmonary artery (arrow in A) and small subpleural pulmonary infarction in the postero-basal segment of the right lower lobe (arrow in B). Sagittal CT image (C) shows a reduction in the size of the lesions in the RV (arrowhead) and RV outflow tract (arrow) compared with the previous MRI. Axial CT image showing persisting thrombus (arrow in D) in the descending thoracic aorta.

Similar findings were also noted in LGE images with a high inversion time of 600 ms (►Fig. 2C), confirming the diagnosis of thrombus. A nonenhancing thrombus was also seen in the descending thoracic aorta (►Fig. 2D). An endomyocardial biopsy of the RV lesion was done and pathological examination showed fibrinous material suggestive of thrombus, consistent with CMR diagnosis.

Workup for hypercoagulable states revealed positive lupus anticoagulant and antinuclear antibody confirming the diagnosis of secondary APS. The patient was started on immunosuppressants, antiplatelets, and anticoagulants and remained asymptomatic. Follow-up contrast-enhanced CT done 3 months later revealed a reduction in the size of the thrombi in the RA and RV (►Fig. 3C) with chronic emboli in the segmental branches of the right lower lobe pulmonary artery (►Fig. 3A) and a small pulmonary infarct in the lower lobe of the right lung (►Fig. 3C), likely secondary to embolism of the thrombi in the RA and RV. The aortic thrombus had also mildly reduced in size (►Fig. 3D).

The patient was managed medically and the anticoagulant dose was optimized to achieve the target INR. The patient improved clinically with medical management and was kept on follow-up.

Discussion

APS is a multisystem autoimmune disease associated with recurrent arterial and venous thrombosis, placental insufficiency, fetal loss, and the presence of antiphospholipid antibodies. It can occur as a primary disorder (primary APS) or secondary to another autoimmune disease (secondary APS), most commonly systemic lupus erythematosus. The revised Sapporo classification criteria are widely used for the diagnosis of APS and require the presence of at least one

clinical and at least one laboratory criteria.⁶ The cardiac manifestations include valvulopathy, coronary artery disease (CAD), myocardial dysfunction, pulmonary hypertension, and intracardiac thrombus.⁷ These manifestations are either secondary to immune-mediated and/or thrombotic mechanisms. Valvular involvement is the most common cardiac manifestation of APS presenting as a valvular thickening and non-bacterial thrombotic endocarditis (Libman–Sacks endocarditis). The mitral valve and aortic valve are the most commonly involved valves. Aortic valve lesions confer an increased risk of stroke. APS is associated with premature accelerated atherosclerosis, coronary artery disease, and an increased risk of myocardial infarction (MI). MI may result from accelerated CAD, microvascular injury, or coronary artery embolism. Pulmonary thromboembolism with resultant chronic thromboembolic pulmonary hypertension (CTEPH) is a common manifestation of APS and can develop secondary to in situ thrombosis and embolism from lower limb deep vein thrombosis and right cardiac chambers thrombi.^{1,8,9}

Intracardiac thrombus is a rare cardiac manifestation of APS and is thought to be secondary to myocardial dysfunction and intra-cardiac thrombosis. CMR, owing to its superior tissue contrast, is central to the detection and characterization of thrombi, differentiating it from true cardiac neoplasms.¹⁰ Thrombophilia, vasculitis, and eosinophilic cardiac disease can also present with multifocal cardiac thrombi and should be included in the differential diagnosis in such situations. Antiplatelets, anticoagulants, and immunosuppressant agents are the cornerstone of APS treatment. Surgery may be indicated in patients with severe valvular dysfunction and with recurrent embolism despite anticoagulation.⁸

A diagnosis of hypercoagulable states, including APS, should be considered in patients having multiple cardiac thrombi. Imaging, especially CMR, plays a crucial role in the diagnosis of cardiac involvement.

Conflict of Interest

None declared.

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