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IMAGING VIGNETTE

BEGINNER

CLINICAL VIGNETTE

Acute Systemic Lupus Erythematous and Antiphospholipid Syndrome With Cardiac Complications

Manish Motwani, MBChB, PhD,[a,b] Matthew Luckie, MBChB, MD,[a] Ben Parker, MBChB, PhD[c,d]

ABSTRACT

This case illustrates the potential for multiple cardiovascular complications in acute systemic lupus erythematous with antiphospholipid syndrome. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2019;1:240–2)
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A 53-year-old Vietnamese woman presented with acute confusion, myalgia, pyrexia, and pancytopenia. On the basis of her systemic symptoms and specialist bloodwork, including positive antinuclear antibodies, she received a diagnosis of acute-onset systemic lupus erythematous (SLE) with macrophage activation syndrome. She was additionally found to have a triple-positive antiphospholipid (aPL) antibody profile (positive for lupus anticoagulant, anticardiolipin, and beta$_2$-glycoprotein antibodies) indicating a particularly high risk for thrombosis and neuropsychiatric manifestations (1,2). The latter was thought to account for the prominent encephalopathy (postulated mechanisms in aPL syndrome include aPL antibody-related microvascular thrombosis or a direct effect of aPL antibodies on brain tissue) (3). Brain magnetic resonance imaging revealed only chronic white matter change. She went on to develop acute kidney injury confirmed as lupus nephritis on biopsy, and her cardiac status also abruptly declined. Transthoracic and transesophageal echocardiography revealed a mildly dilated left ventricle with severe global systolic impairment, severe aortic regurgitation but no vegetations, and a dilated ascending aorta. Her troponin T level was moderately raised, but she had no reported chest pain or specific electrocardiographic changes. She was managed with low-molecular-weight heparin, high-dose steroids, 5 cycles of plasma exchange, and concurrent intravenous cyclophosphamide, as well as standard heart failure therapies. She responded well, making a remarkable multisystem recovery over the next 2 months, and was discharged home with specialist rheumatology, nephrology, and cardiology outpatient follow-up. The multiple cardiac sequelae of her acute lupus presentation are well visualized on follow-up cardiac magnetic resonance (CMR) scan at 12 months (**Figures 1A to 1D**, Video 1) that show

From the [a]Manchester Heart Centre, Manchester Royal Infirmary, Manchester University NHS Foundation Trust, Manchester, United Kingdom; [b]Institute of Cardiovascular Sciences, University of Manchester, Manchester, United Kingdom; [c]Kellgren Centre for Rheumatology, Manchester Biomedical Research Centre, Manchester University NHS Foundation Trust, Manchester, United Kingdom; and the [d]Centre for Musculoskeletal Research, University of Manchester, Manchester, United Kingdom. The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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prior aortic valvulitis with poor coaptation and moderate residual aortic regurgitation (Figures 1A and 1B, blue arrows), significant aortopathy involving a mildly dilated root and ascending aorta (Figure 1C, green arrow), and midwall fibrosis in the midinferior septum consistent with prior myocarditis (Figure 1D, orange arrow). The CMR scan also confirmed left ventricular recovery to normal size and systolic function (ejection fraction, 61%; end-diastolic volume, 132 ml). This case illustrates the potential of SLE and aPL syndrome to cause multiple cardiovascular complications and highlights the broad multiparametric imaging capabilities of CMR to visualize them all.

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ABBREVIATIONS AND ACRONYMS

ANA = antinuclear antibodies

aPL = antiphospholipid

AR = aortic regurgitation

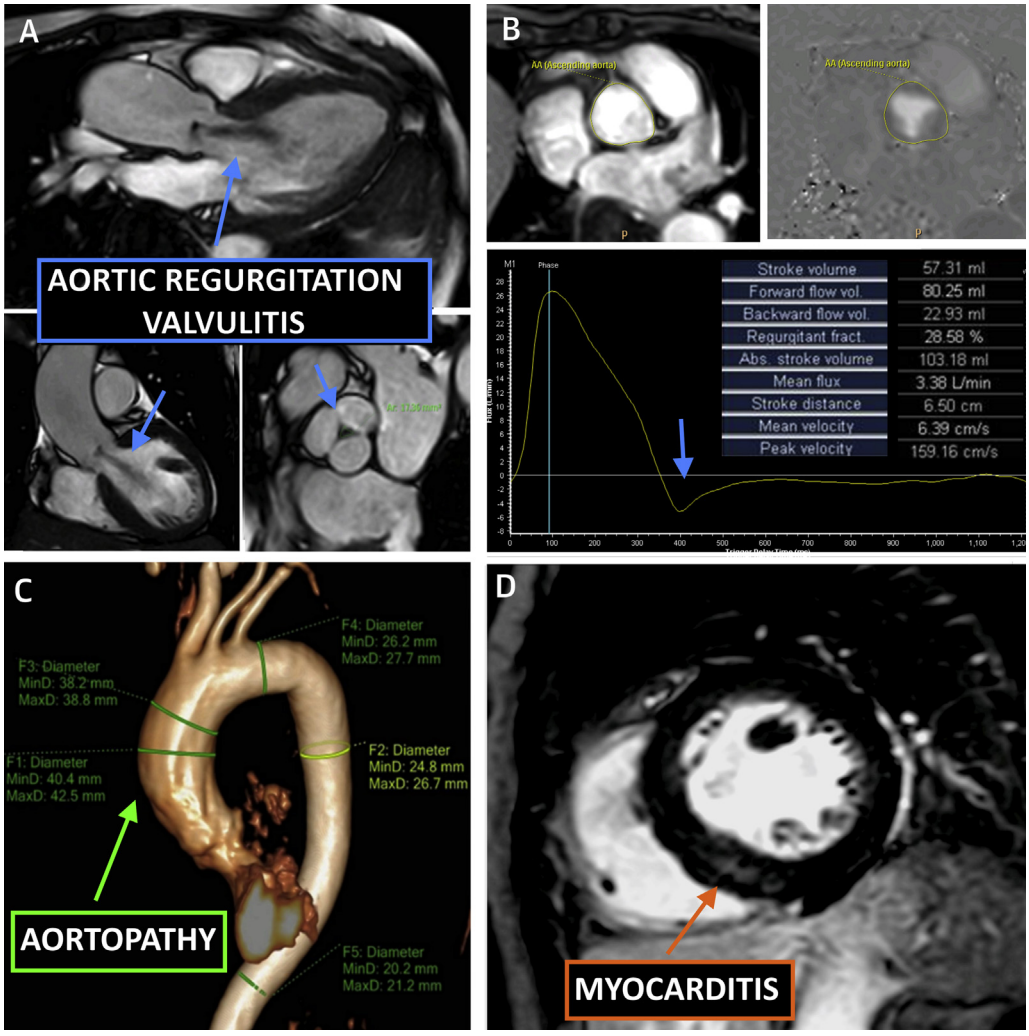
CMR = cardiovascular magnetic resonance

ECG = electrocardiogram

LV = left ventricle

SLE = systemic lupus erythematous

FIGURE 1 CMR in Acute SLE



Multiple cardiac sequelae following acute systemic lupus erythematous (SLE) are seen on follow-up cardiac magnetic resonance (CMR) imaging at 12 months. **(A)** Cine images show thickened aortic valve leaflets consistent with prior valvulitis and significant residual aortic regurgitation **(blue arrows)** (Video 1). **(B)** Phase-contrast images quantify residual aortic regurgitation as moderate (23 ml/beat; regurgitant fraction 29%) **(blue arrow)**. **(C)** Contrast-enhanced angiography with 3-dimensional volume-rendered reconstruction illustrates significant aortopathy involving a mildly dilated root and ascending aorta **(green arrow)**. **(D)** Late gadolinium enhancement images show midwall fibrosis in the midinferior septum consistent with prior myocarditis **(orange arrow)**. Abs = absolute; MaxD = maximum diameter; MinD = minimum diameter.

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ADDRESS FOR CORRESPONDENCE: Dr. Manish Motwani, Manchester Heart Centre, Oxford Road, Manchester University NHS Foundation Trust, Manchester, United Kingdom. E-mail: mani.motwani@mft.nhs.uk.

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KEY WORDS antiphospholipid syndrome, aortopathy, myocarditis, systemic lupus erythematosus, valvulitis

APPENDIX For a supplemental video, please see the online version of this paper.