

July 2021

Nonbacterial Thrombotic Mitral Valve Endocarditis Presenting as Embolic Stroke in a Young Patient with Lupus and Anti-phospholipid Syndrome

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Recommended Citation

McCuen A, Saad M, Terpstra EJ, Stys AT. Nonbacterial Thrombotic Mitral Valve Endocarditis Presenting as Embolic Stroke in a Young Patient with Lupus and Anti-phospholipid Syndrome. *Aesculapius*. 2021 Jul 20; 2(1):Article 5. Available from: <https://red.library.usd.edu/aesculapius/vol2/iss1/5>. Free full text article.

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DESCRIPTION

A 37-year-old man, on systemic immunosuppression for clinically and biochemically quiescent lupus nephritis, presented with left hemiparesis. Right frontal, parietal and occipital ischemic infarcts on brain MRI were concerning for embolic stroke. There was no evidence of atrial fibrillation on EKG/telemetry or deep venous thrombosis on duplex ultrasound of the lower extremities. The patient did not have signs or symptoms of heart failure.

Following an unrevealing transthoracic echocardiogram, transesophageal echocardiogram (TEE) showed sub-centimetre (7 mm) fixed mitral valve leaflet echodensities and moderate mitral regurgitation (Figure 1). No thrombus was seen in the left atrial appendage.

Small vegetations on atrial and ventricular sides of mitral valve leaflets (Figures 1 and 2) without independent motion, the absence of bacteremia and systemic signs of infection, and a history of lupus were suggestive of nonbacterial thrombotic endocarditis. Further workup for hypercoagulability was consistent with anti-phospholipid syndrome (previously undiagnosed, secondary to lupus).

Nonbacterial thrombotic endocarditis is known to be associated with hypercoagulable and/or autoimmune disorders. [1] Small vegetations on the ventricular side of mitral valve may help differentiate it from infective endocarditis, as the later typically involves the atrial side with larger, mobile vegetations. However, diagnosis of nonbacterial thrombotic endocarditis requires exclusion of infective endocarditis.

Nonbacterial thrombotic vegetations, a consequence of hypercoagulability, are composed of fibrin deposits on otherwise-healthy valves. This likely explains their predisposition for the ventricular side of the leaflets where there is relative stasis of blood. Mainstay of treatment is therapeutic anticoagulation. Warfarin was started before discharge in the above-described patient. Near-complete resolution of left hemiparesis was reported on 2-month follow up visit. Clinical and echocardiographic surveillance for moderate-severe mitral regurgitation will continue.

LEARNING POINTS

1. Diagnosis of nonbacterial thrombotic endocarditis requires high clinical suspicion because the usually sub-centimetre vegetations are rarely visualised on transthoracic echocardiogram.
2. Involvement of the ventricular side of mitral valve leaflets (Figures 1 &2), absence of bacteremia and systemic signs of infection, and a history of autoimmune/hypercoagulable disorder are suggestive of nonbacterial thrombotic endocarditis.

REFERENCES

1. Liu J, Frishman WH. Nonbacterial Thrombotic Endocarditis: Pathogenesis, Diagnosis, and Management. *Cardiology in Review*. 2016;24(5).

FIGURE/VIDEO CAPTIONS

1. TEE: mid-esophageal long axis view.
Left panel: small echodensities on atrial (orange arrow) and ventricular (yellow arrow) sides of mitral valve;
Right panel: colour Doppler suggesting at least moderate mitral regurgitation
2. TEE: mid-esophageal mitral valve commissural view.
Non-mobile echodensity on ventricular side of mitral valve (yellow arrow). There is no systolic prolapse into the left atrium.

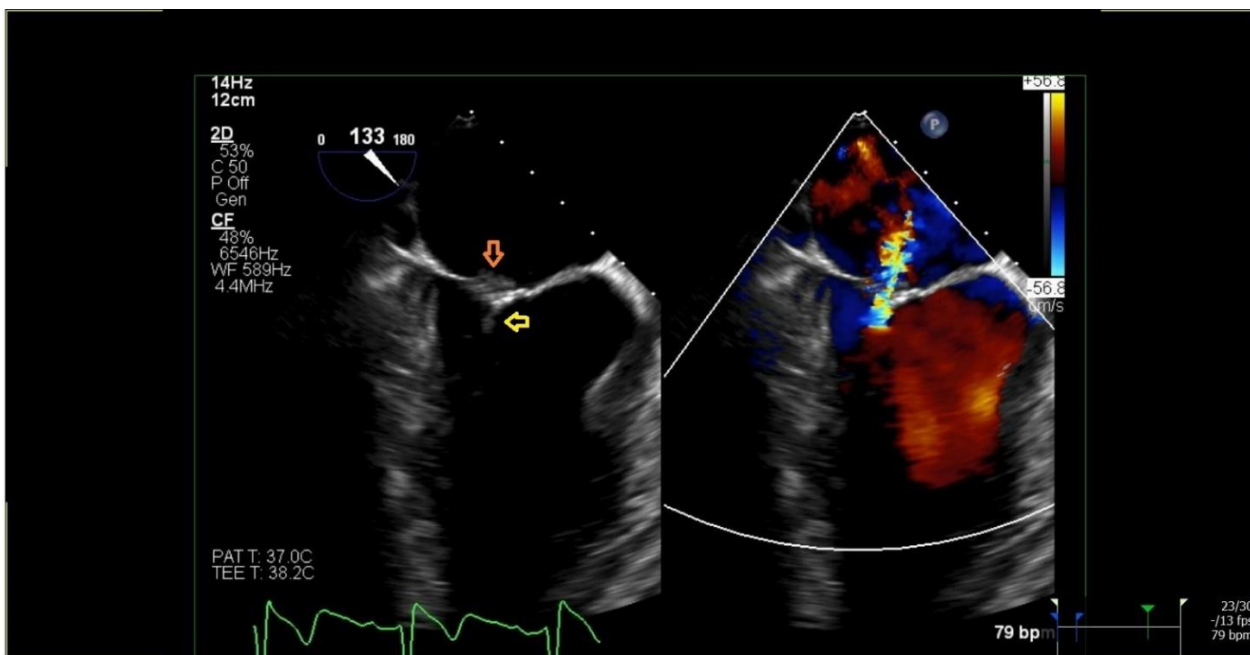


Figure 1

TEE mid-esophageal long axis view demonstrate small echodensities on atrial (orange arrow) and ventricular (yellow arrow) sides of mitral valve (panel A); Colour Doppler suggests at least moderate mitral regurgitation (panel B).



Figure 2

TEE mid-oesophageal mitral valve commissural view shows non-mobile echodensity on ventricular side of mitral valve (yellow arrow). There is no systolic prolapse into the left atrium.