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Atrioventricular Heart Block and Syncope Coincident With Diagnosis of Systemic Lupus Erythematosus

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Abstract

We describe a 59-year-old woman with cardiac conduction abnormalities caused by lupus-induced myocardial damage. She had a history of arthralgias and antinuclear antibodies but no clinical history of systemic lupus erythematosus. She presented with syncope and Mobitz type II second-degree atrioventricular block. Anti-double-stranded DNA antibodies developed coincident with the identification of heart block. Cardiac magnetic resonance imaging showed late enhancing foci of gadolinium uptake that anatomically correlated with her conduction abnormalities. We conclude that her conduction disease represents an early and structural cardiac manifestation of systemic lupus erythematosus that is unusual in its presentation at the time of initial diagnosis.

Cardiac involvement with systemic lupus erythematosus (SLE) usually occurs in patients with an established diagnosis and typically involves the pericardium, myocardium, valves, and coronary vessels[1] with conduction abnormalities being rare. Previous reports of lupus-related heart block have occurred in patients with long-standing clinical disease or other possible aetiologies.[2,3] We describe the case of a 59-year-old Caucasian woman with no medical history presenting with symptomatic Mobitz type II atrioventricular block concomitant with an initial diagnosis of SLE. The etiology of her heart block was focal myocardial damage, likely lupus-induced, detected using late gadolinium enhancement (LGE) on cardiac magnetic resonance (CMR) imaging.

Case Description

The patient presented to the emergency department after a witnessed syncopal event. She had no prodromal chest pain, dyspnea, palpitations, or presyncope, and no previous syncopal episodes. She denied angina, dyspnea on exertion, paroxysmal nocturnal dyspnea, orthopnea, edema, fever, rash, and pleurisy.

The patient had no other medical history and no family history of cardiac disease. She was taking no medications. Her physical examination was unremarkable.

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Disclosures
The authors have no conflicts of interest to disclose.

On review of the medical records, the patient had a history of arthralgias with elevated serum antinuclear antibodies (ANAs) (Fig. 2). However, before her syncopal event, tests for anti-double-stranded (ds) DNA antibodies had been negative. Previous assessments by a rheumatologist found no evidence of connective tissue disease, and the elevated ANA had been followed expectantly.

Her admission electrocardiogram revealed a markedly prolonged PR interval (378 ms) and a right bundle branch block. Telemetry monitoring showed intermittent episodes of Mobitz type II atrioventricular block during which the patient experienced presyncope (Fig. 1A).

Admission lab tests were remarkable for newly positive anti-dsDNA titers, a 1:2560 titer of ANAs with a homogenous speckled immunofluoroescence pattern, and weakly positive IgG cardiolipin antibodies. A complete blood count, basic metabolic panel, cardiac-specific troponin, creatine phosphokinase, thyroid stimulating hormone, serum and urine protein electrophoresis, angiotensin-converting enzyme level, and complement levels were within normal limits. Rheumatoid factor, SS-A and SS-B antibodies, anti-Smith antibody, anti-cyclic citrillunated protein, SCL-70 antibody, JO-1 antibody, Lyme disease antibodies, and rapid plasma reagin were negative.

CMR imaging showed a normal left ventricular ejection fraction (63%), and LGE of the basal ventricular septum near the atrioventricular node and the anterolateral papillary muscle (Fig. 1C). Coronary angiography was deferred, because the affected regions of inflamed myocardium did not correspond to a typical coronary vascular territory.

Discussion

The differential diagnosis for premature cardiac conduction disease includes myocardial ischemia, infections, inflammatory disorders, rheumatologic disease, amyloidosis, and idiopathic disease. Our patient had no historical, physical, or laboratory evidence for any alternative diagnosis except SLE. Moreover, her history of nonerosive oligoarthritis, positive ANA, an abnormal titer of anti-dsDNA, and inflammatory cardiac lesion suggested a new diagnosis of SLE complicated by cardiac conduction disease.

The patient underwent an electrophysiology study demonstrating a prolonged His-V interval (Fig. 1B) with variable infra-His conduction delay, and easily inducible sustained polymorphic ventricular tachycardia degenerating into ventricular fibrillation. Cardiac rhythm disturbances might manifest as both conduction defects and/or tachyarrhythmias in autoimmune connective tissue diseases.[4] Therefore, a ventricular arrhythmia rather than heart block as the cause of her initial syncopal event could not be excluded, and the patient underwent defibrillator placement.

At 6-month follow-up, she had no further syncope or cardiac or rheumatologic symptoms. Her implantable cardioverter defibrillator recorded no malignant ventricular arrhythmias, but her conduction disease had progressed to complete heart block with complete pacemaker dependence.

Conclusion

This case demonstrates that Mobitz type II atrioventricular block can be an initial cardiac manifestation of SLE, offers a clear anatomical visualization of the lesion that caused the heart block, and highlights the asymptomatic period of serological positivity which often precedes clinical SLE. The patient’s pattern of increasing ANA culminating in the development of anti-dsDNA antibodies and overt disease is consistent with autoantibody crescendo, which often anticipates clinical SLE.[5] We speculate that cumulative lupus-

induced autoimmune effects led to a structural lesion involving the conduction system, resulting in high-grade atrioventricular block. The multiple LGE lesions seen onCMRimaging suggest a broader inflammatory process in the heart.

Previous descriptions of SLE-associated heart block have been poorly characterized and confounded by other possible aetiologies. Our patient lacked confounding causes of atrioventricular block and clearly had the onset of cardiac disease concomitant with anti-dsDNA antibody positivity. With the inflammatory myocardial changes seen on CMR imaging providing an anatomical explanation for her electrical abnormalities, her heart block clearly represents the first clinically identifiable cardiac manifestation of SLE.

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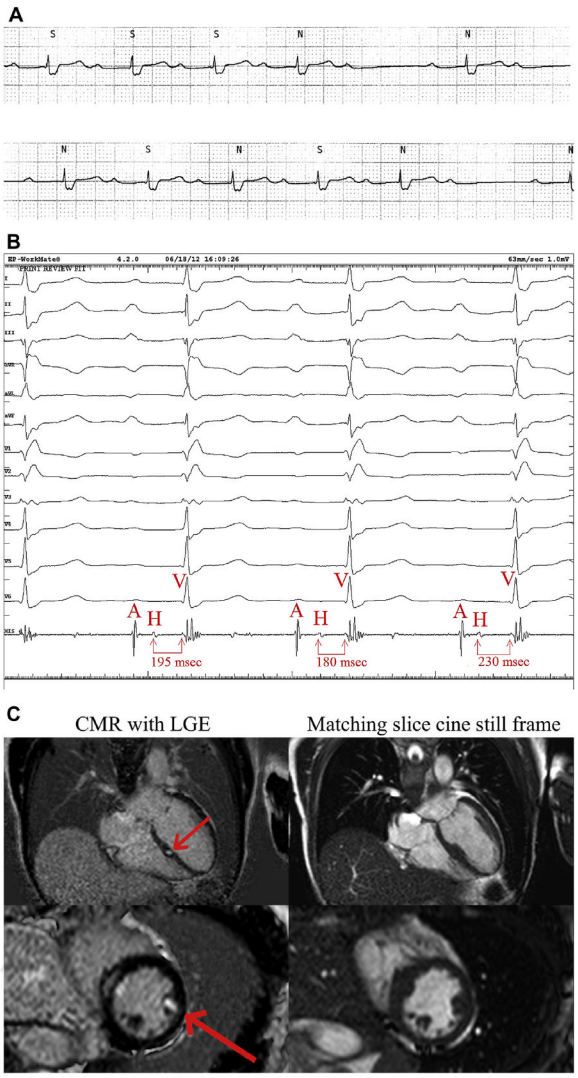

Figure 1.

Second-degree atrioventricular block and corresponding focus of septal LGE on CMR. (**A**) Rhythm strip during presyncope. (**B**) Intracardiac electrogram showing long His-V intervals. (**C**) CMR showing LGE in the septum and anterolateral papillary muscle (**red arrows**). CMR, cardiac magnetic resonance; LGE, late gadolinium enhancement.

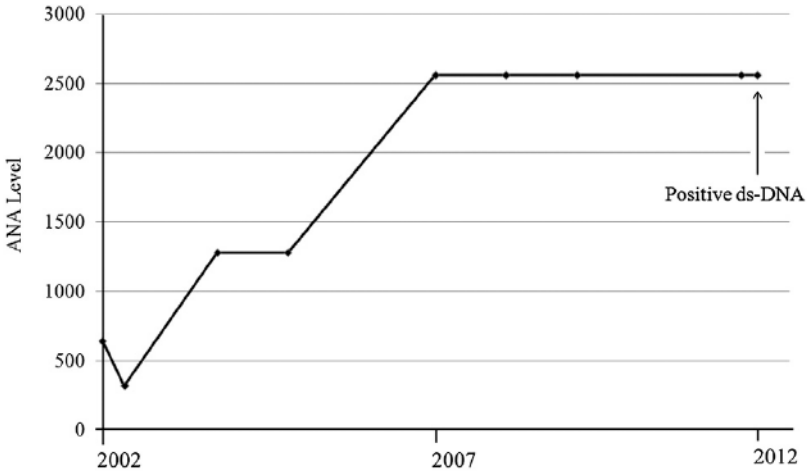

Figure 2.
Antinuclear antibody crescendo prior to the development of anti-dsDNA antibodies and overt cardiac disease. ANA, antinuclear antibody; ds, double-stranded.