

## Complete atrioventricular block as initial manifestation of systemic lupus erythematosus

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## ABSTRACT

*Only a few cases of complete atrioventricular block (AVB) in adult lupus patients have been previously described, but only one as the initial manifestation. A 19-year-old woman who presented with seizures and loss of consciousness, was diagnosed with complete AVB and underwent pacemaker placement. Over the next weeks she developed serositis, joint, cutaneous, and renal involvement; positive antinuclear antibodies and high anti-SSA/Ro titers. This is the second case with AVB as a feature of SLE at onset. A review of previous complete AVB cases of adult SLE patients is presented.*

## Introduction

Heart involvement in SLE has a wide array of expressions, some of them not clinically evident, which require adequate image or functional studies to be detected. SLE can affect all heart structures, including the pericardium, producing variable amount of effusion, the myocardium developing myocarditis, the conduction system with either reversible or permanent derangement, the coronary vessels yielding ischemic cardiomyopathy, and valves leading to Libman-Sacks endocardial lesions (1). Conduction system involvement is described as the less frequent form of cardiac disease; electrocardiographic data are sinus tachycardia, atrial or ventricular premature beats, atrial fibrillation, bundle branch block, and more seldom seen, diverse grades of atrioventricular block (AVB) (2). Only 12 cases of SLE patients with complete AVB have been previously described (3-5); although there is only one previous case in which this conduction abnormality was reported as the initial SLE symptom (4). We present here the case of a young woman who developed such abnormality at the onset of SLE.

## Case report

A 19-year-old single woman was referred to our hospital with a temporary pacemaker and the diagnosis of complete AVB. She was a high school student, had family history of diabetes mellitus and a first-degree relative with rheumatoid arthritis. No previous acute

or chronic illnesses were referred. Four months previous to her admission, while she was at school, generalized seizures were witnessed by her classmates, she was taken to a hospital nearby and an EKG disclosed bradycardia of 42 bpm with an image of high grade AVB (Fig. 1). A cardiologist placed a temporary pacemaker with recovery of her consciousness state, and once stabilized, she was sent to our hospital. On admission to the hospital's cardiology department she was alert, with no neurological deficit; after the placement of a permanent tricameral pacemaker, she reported fatigue, chills, low-grade fever, and hand arthralgia by the last days. Both, head CT scan and electroencephalogram were normal, and she was discharged with nimesulide according to symptoms.

During the next 10 weeks, she noticed an increase of fatigue, which interfered with her school activities, as well as mild hair loss, malar rash, fever of 38.5°C, arthritis of hands and knees, and at least 10% of weight loss since the beginning of the symptomatic period; by that time she was sent to our Department. On physical examination, malar rash, pericardial and pleural rubs, arthritis of wrists, MCP/PIP joints, knees and ankles were recognized. Chest and abdomen CT-scan revealed pericardial and pleural effusions. Echocardiogram showed an ejection fraction of 52% and 500 cc of pericardial effusion. Laboratory depicted hemoglobin of 11.1 g/L, leukocyte count of 6.07/10<sup>9</sup>/L, lymphocyte count 2.4/10<sup>9</sup>/L, platelet count 217/10<sup>9</sup>/L, serum creatinine 0.6 g/L; urinalysis with proteinuria 2+/4+; positive C-reactive protein, and negative lupus anticoagulant and rheumatoid factor. Antinuclear antibodies disclosed 1:640 speckled pattern, positive anti-DNA antibodies in *Crithidia lucilae* immunofluorescence, and negative anti-Sm, anti-RNP, anti-SSB/La antibodies, but high titers of anti-SSA/Ro antibodies by ELISA. The patient was started on prednisone 40 mg qd and azathioprine 100 mg qd, which improved all clinical data. Two years later, she remains in clinical remission, with a functional pacemaker, and on azathioprine alone.

*Competing interests: none declared.*

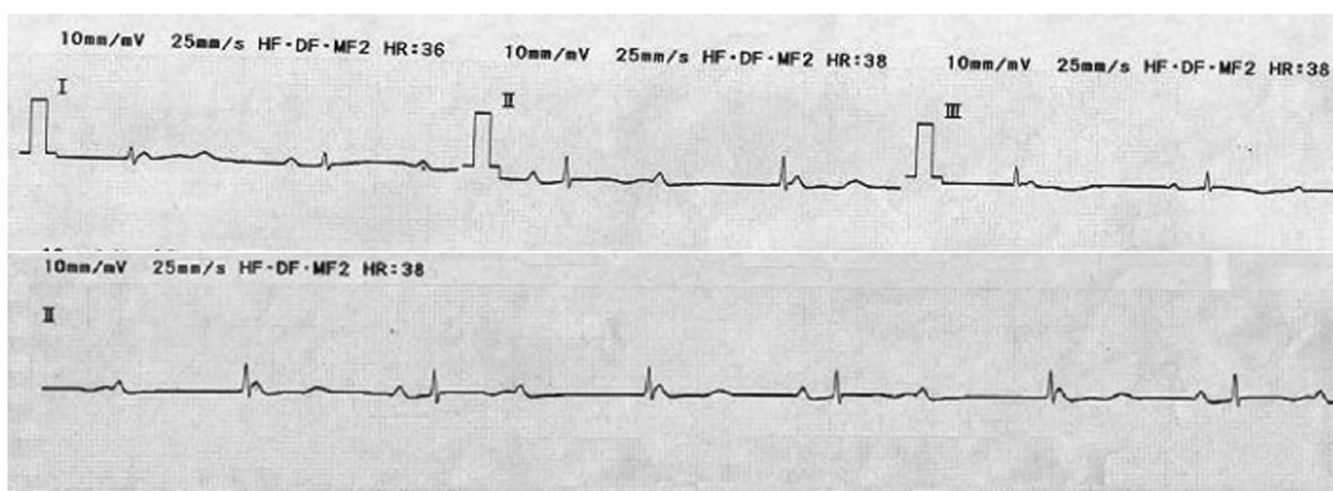


Fig. 1. Surface leads in the electrocardiogram showing the classical image of third degree atrioventricular block.

## Discussion

The heart is commonly affected in SLE patients; the most common clinical manifestation is pericarditis, which might often be clinically indolent and found only in post-mortem studies. Libman-Sacks endocarditis is also present in 13-74% of patients; and arterial coronary disease remains a leading cause of death in SLE patients, associated with atherosclerosis, vasculitis, and thrombosis related to anticardiolipin antibodies (1).

Arrhythmia in SLE patients may present as sinus tachycardia, which might be seen as a part of SLE activity (6) or even like a life-threatening problem; ischemic heart disease secondary to accelerated atherosclerosis (7) anti-cardiolipin antibodies or vasculitis, and direct damage of the conduction system have been associated with this condition. Even more, conduction system abnormalities in SLE are documented in two settings: the first, in babies born to women with SLE or primary Sjögren's syndrome, particularly when they have high anti-SSA/Ro antibody titers (8); and the second, in adult patients with cardiac rhythm abnormalities and different grades of AV block seen during SLE flares, and in some cases due to antimalarial cardiotoxicity. Lazzerini and coworkers have demonstrated that anti-Ro/SSA antibodies are closely related to QT interval prolongation and complex ventricular arrhythmia (9), suggesting a possible interference in the bioelectric myocardiocytes activity. Complete AVB in adult SLE patients

has been reported in 12 previous cases, almost all reviewed by Lim and Joshua (3), and another reported by Gómez-Barrado (5). They described patients with active disease; in 5 of them, anti-SSA/Ro antibodies were found, meanwhile other 4 cases had anti-Sm/U1RNP antibodies. AVB regressed to normal AV conduction with steroid administration in 4 cases, but other 6 required the use of a permanent pacemaker. Maier and coworkers, presented the case of a 27 year-old woman who developed a complete AVB that resolved spontaneously and required nothing but a temporary pacemaker, and six weeks later she developed full symptoms corresponding to SLE; also developing high anti-SSA/Ro antibodies titers (4).

In newborns, complete AVB is associated with transplacental passage of anti-SSA/Ro and anti-SSB/La antibodies. The pathogenesis of this disorder involves apoptosis of myocardiocytes, and secondary conduction system fibrosis (10). In adult patients with this disorder anti-SSA/Ro antibodies have been found as in the case described herein; but the pathogenic mechanism of complete heart block in patients with positive anti-SSA/Ro antibodies has not been completely established, although there have been proposed changes in the bioelectric cell properties. Anti-Sm/U1RNP antibodies have also been found in about 50% of previous cases with complete AVB; however, no explanation for this presentation has been proposed.

The permanent pacemaker has not been removed in our patient in accordance with some cases previously described, in whom pacemakers are used in spite of the apparent resolution of the conduction abnormality because of the possibility of recurrence. It should be emphasized that conduction cardiac abnormalities in SLE adult patients are a rare manifestation of disease and, as we report here, can be the initial symptom. This case represents the 13<sup>th</sup> adult SLE patient with complete AVB, and the second in whom this cardiac abnormality is recognized before the full presence of other features of disease. Relation between anti-SSA/Ro antibodies and conduction system derangement in adult patients is warranted.

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