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Case Report

Complete AV block and cardiac syncope in a patient with Duchenne muscular dystrophy

Refik Emre Altekin (MD)*, Atakan Yanikoglu (MD), Mustafa Ucar (MD), Cengiz Ermis (MD, Phd)

Akdeniz University Faculty of Medicine, Cardiology Department, Dumlupinar Bouleward, Konyaaltı, Antalya, Turkey

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Summary Duchenne muscular dystrophy is an X linked hereditary progressive neuromuscular disease and it is characterized by development of weakness and atrophy in affected muscles. In late phases of disease with involvement of respiratory and cardiac muscles, patients die because of respiratory and cardiac failure. Cardiomyopathy is a common complication and various types of arrhythmia because of conduction system involvement can be seen. Herein we present a case with Duchenne muscular dsytrophy who was admitted to our clinic at an older age because of complete atrioventricular block without cardiomyopathy.
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Introduction

The reason for Duchenne muscular dystrophy (DMD) is a mutation of the dystrophin gene localized in Xp21 segment. Clinical manifestations begin in childhood and muscle weakness and atrophy develop progressively [1]. Cardiomyopathy and cardiac failure because of cardiac involvement is one cause of mortality in these patients [2]. Besides this, various types of arrhythmias can be seen with involvement of the conduction system. Sinus tachycardia is the most common of these and atrioventricular (AV) block can be seen rarely [3]. In our case, a patient with DMD and advanced AV block who was admitted to the emergency department with cardiac syncope is discussed.

Case report

A 38-year-old male patient with a diagnosis of DMD for 23 years, without any known cardiac disease was admitted to the emergency department with an episode of loss of consciousness. He had a total of 5 syncope episodes in the previous month, and was brought to the emergency department because of the long duration of the last episode. The patient was diagnosed with DMD when he was 15, and this was confirmed by muscle biopsies and genetic screening. The patients' ability of movement was progressively restricted in the following years and he was in a wheelchair for the previous 4 years. There was no history of using any cardiac medications. On physical examination, his mental status was in a state of confusion. Blood pressure was 70/40 mmHg,

* Corresponding author at: Akdeniz University Faculty of Medicine Hospital, Cardiology Department, Dumlupinar Bouleward, Zip Code:07100, Antalya, Turkey.
Tel.: +90 242 2676804; fax: +90 242 2274490.
E-mail address: dremre29@yahoo.com (R.E. Altekin).

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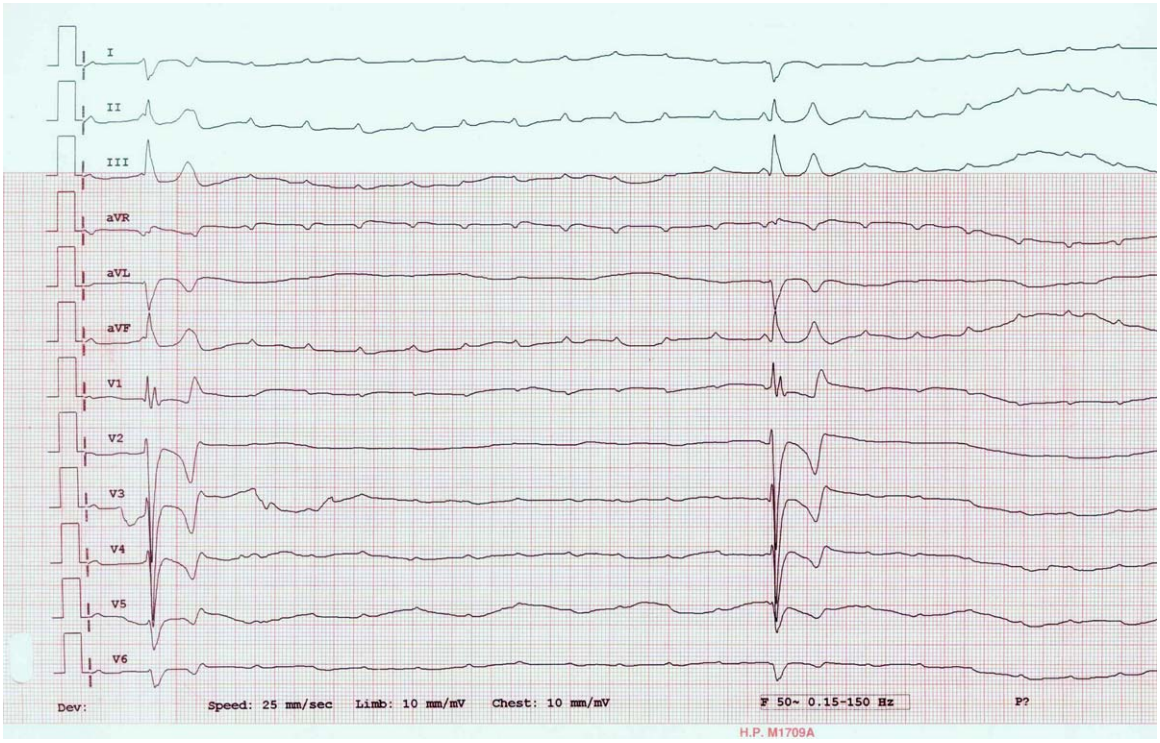


Figure 1 Electrocardiogram showing complete atrioventricular block.

pulse was 15 beats/min and pulse oximetry saturation was 95%. Cardiac and respiratory system examinations revealed nothing with the exception of bradycardia. He has kyphoscoliosis and he had flexion type contractures and atrophy on both upper and lower extremities. There were advanced AV block and ventricular pause approximately lasting for 4.5 s on electrocardiography (ECG) (Fig. 1). There were not any metabolic or electrolyte abnormalities on laboratory examinations. Following transcuteanous pacing to 60 beats/min, transient subclavian venous pacing was performed in the emergency department and the patient was taken to the coronary care unit. The patient was in sinus rhythm spontaneously after 12 h of hospitalization and besides sinus tachycardia there was a trifascicular block which was composed of first-degree AV block, right bundle branch block, and left posterior hemiblock on ECG (Fig. 2). One day later,

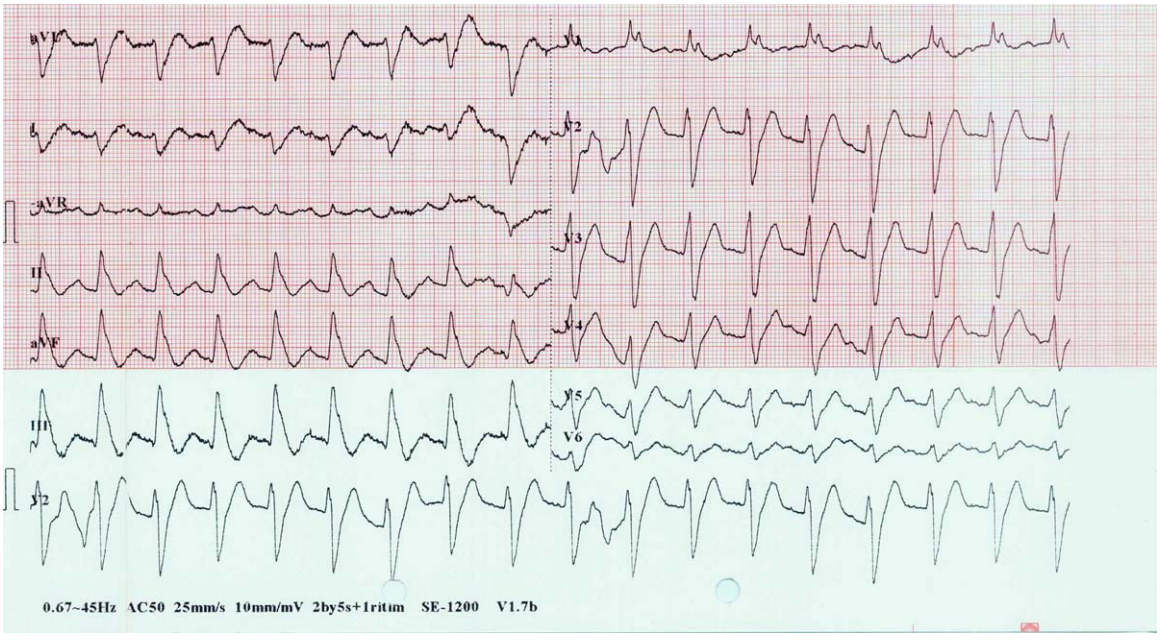


Figure 2 Electrocardiogram showing trifascicular block.

although the patient was in sinus rhythm, a double chamber pacemaker with both sensing and pacing features of atrium and ventricle (DDD-R mode) with an adjusted rate of 70 beats per minute, was implanted to the patient because of trifasciular block and a previously documented complete AV block. Echocardiography was performed and ejection fraction was found to be 55% without any wall motion abnormality with a left ventricular end diastolic diameter 49 mm and an end systolic diameter 34 mm. The patient was discharged without any complication after 48 h of hospitalization.

Discussion

DMD is a muscular disorder which is caused by a defect of X linked inherited dystrophin gene and is seen in 1/3500 of male births. Dystrophin binds the actin to protein extracellular matrix. In DMD, dystrophin is absent. In the absence of dystrophin, muscle membrane disintegrates and cellular death results with the oxidative stress caused by excessive intracellular calcium influx. Fatty and fibrous tissue takes the place of dead myocytes. Diagnosis can be made with the immunohistochemical staining of muscle biopsies and genetic screening for the defect [4].

Usually, symptoms begin earlier than 6 years of age but rarely presentation can be seen in late childhood. Muscles of the pelvis, shoulder, hip, and thigh are most commonly involved, but with time involvement of upper extremity, neck, and respiratory muscles can be seen. Lordosis and kyphoscoliosis because of muscle atrophy can be seen. Pseudohypertrophy is a common finding because of fatty and fibrous tissue deposition in muscle tissue besides the atrophy and weakness. Most of the patients die in the third decade, but there are rare cases living to 40 years of age [5].

Fatty and fibrous tissue takes the place of necrotic myocytes in case of cardiac involvement. Involvement starts with posterobasal segment. Septum, right ventricle, and all myocardium with the exception of the atria will be involved in time resulting in dilated cardiomyopathy. Cardiac failure is the result of death in 10% of cases [2]. Sinus tachycardia, wide R waves on precordial leads, increased R/S ratio, short PR, and a long QT interval because of conduction system involvement and Q waves in lateral derivations because of posterobasal segment involvement can be seen on ECG [5,6]. Because of the defect in dystrophin protein on membranes of Purkinje fibers, fatty and fibrous tissue replacement of the involved segment may rarely result in AV blocks [7]. Electrophysiologic studies revelaed conduction disturbances at His and infra His level in patients with various types of muscular dystrophy and in follow up permanent pacemakers were implanted because of AV block [8]. Because of symptomatic AV complete block development in the background of trifascicular block, a dual chamber permanent pacemaker was implanted in the patient. American Collge of Cardiology/American Heart Association guidelines for cardiac pacing and anti-arrhythmic devices suggest permanent pacemaker implantation as class I indication in patients with muscular dystrophies [9].

In two similar cases reported by Fayssoil et al., dual chamber pacemakers were implanted to DMD patients with cardiomyopathy because of AV complete block and sinus node dysfunction [5,10]. In our case there was no known prior cardiac disease and AV complete block is the first clinical presentation and there was no apparent left ventricle dilatation on transthoracic echocardiography. Usually, conduction system abnormalities are seen in late phases of disease in DMD patients with cardiomyopathy which had already developed.

Cardiac involvement in DMD is usually seen as cardiomyopathy at ages between 10 and 20 years. Conduction abnormalities develop with the involvement of the conduction system by fibrous changes which accompanies cardiomyopathy [2—10]. In our case, cardiac involvement developed in the late phase of disease and presented as AV complete block.

In muscular dystrophies, cardiac failure is common but also various rhythm problems can be seen because of involvement of the conduction system. Conduction problems without cardiac failure can be the first clinical signs of cardiac involvement. Because of this reason, conduction problems can be detected with ECG controls in asymptomatic patients with muscular dystrophy in the subclinical phase.

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