

Rapid Progression of Heart Failure in a Patient with Idiopathic Inflammatory Myopathy

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Abstract Idiopathic inflammatory myopathy (IIM) is a rare autoimmune myopathy that includes polymyositis, dermatomyositis, inclusion body myositis and autoimmune necrotizing myositis. Cardiac involvement was considered a rare occurrence in IIM however, recent reports suggests that cardiac involvement is a common feature and portends poor prognosis as it is usually encountered in advanced disease. IIM leads to myocarditis with subsequent development of myocardial fibrosis, cardiac conduction system disease and cardiomyopathy resulting in both systolic and diastolic heart failure. Conduction abnormalities such as first, second and third degree atrioventricular blocks, right and left bundle branch blocks associated with IIM have been reported. We present a case of a 44-year-old woman with biopsy proven-IIM whose left ventricular ejection fraction (LVEF) and electrocardiogram (ECG) were recorded as normal two years prior. On presentation to our hospital ECG revealed atrial tachycardia and 2D echocardiogram revealed heart failure with reduced ejection fraction (20-30%). Patient quickly progressed to complete heart block. A cardiac resynchronization therapy-defibrillator (CRT-D) insertion was planned but patient succumbed to sepsis.

Keywords: *idiopathic inflammatory myopathy, atrial tachycardia, heart failure with reduced ejection fraction*

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1. Introduction

Idiopathic inflammatory myopathy (IIM) is the term used for rare autoimmune myopathies [1,2]. Recent research suggests that cardiac involvement is common in IIM and is a cause of significant morbidity and mortality. Cardiac involvement in these patients occurs secondary to myocarditis and myocardial fibrosis. Myocarditis and myopathy results in cardiomyopathy and heart failure in these patients. Additionally, it may also involve the conduction system of the heart and thus may cause various arrhythmias (3).

2. Case Presentation

A 44-year-old female with a past medical history of obesity, diabetes, hypertension, biopsy proven idiopathic

inflammatory myopathy (IIM) (Image 1), non-ischemic cardiomyopathy related to IIM, heart failure with a reduced ejection fraction of 20-30% and recurrent hospital admissions for heart failure exacerbations presented with worsening dyspnea and lower limb swelling. Patient was ventilator dependent due to chronic respiratory failure from IIM and enteral nutrition was provided via percutaneous endoscopic gastrostomy tube. On presentation her vitals were 36.8°C, heart rate at 84 beats/min, respirations at 20 breaths/min, and blood pressure at 90/63 mmHg. Physical examination revealed bilateral lower extremity edema and diminished lung sounds bilaterally. The strength in all four extremities was 0/5 and she was unable to perform any purposeful movements. Laboratory data was as follows: hyponatremia at 124 mEq/L (133 - 145 mEq/L), blood urea nitrogen (BUN) at 80 mg/dL (7 - 17 mg/dL) and creatinine at 0.25 mg/dL (0.52 - 1.04 mg/dL) alanine aminotransaminase (ALT) at 69 U/L (9 - 52 U/L), aspartate aminotransferase (AST) at 75 U/L (14 - 36 U/L),

troponin I at 1.65 ng/mL (0 - 0.034 ng/mL), and pro B-type natriuretic peptide (PBNP) at 9,640 pg/mL (0 - 125 pg/mL). Chest X-ray showed cardiomegaly with mild congestion and small bilateral effusions. Electrocardiogram (EKG) two years before showed normal sinus rhythm (Image 2) but at presentation she was noted to have atrial tachycardia 2:1 with pseudo-Q wave pattern (Image 3) which then progressed to complete heart block within 7 days of presentation. Transthoracic echocardiogram performed two years prior showed a normal (50-55%) left ventricular systolic function by visual assessment and had

progressively worsened to 25-30%. She was admitted to the cardiac care unit for medical management of heart failure exacerbation. In the setting of complete heart block cardiac resynchronization therapy-defibrillator (CRT-D) was planned as QRS complex duration was greater than 120 ms. CRT-D was considered instead of a pacemaker, as pacemaker placement would have contributed to left ventricular dyssynchrony which would have further contributed to cardiac remodeling and worsening of cardiac systolic function. However, patient succumbed to sepsis.

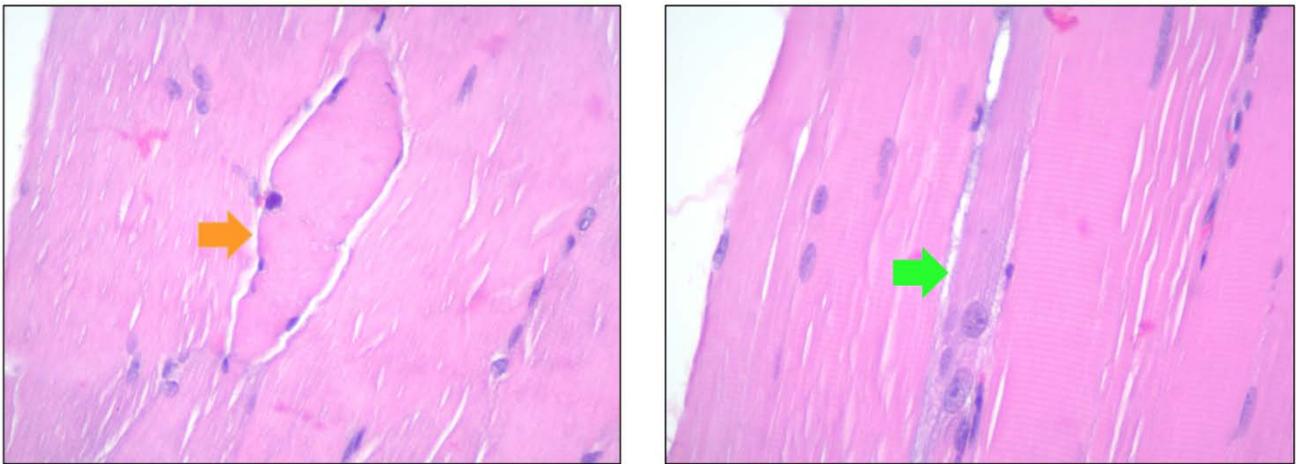


Image 1. Skeletal muscle biopsy of the patient showing a swollen, eosinophilic and vacuolated myofiber that lacks cross striations and shows several pyknotic nuclei (left, orange arrow) and a regenerating myofiber with basophilic cytoplasm and enlarged nuclei with conspicuous nucleoli (right, fluorescent arrow)

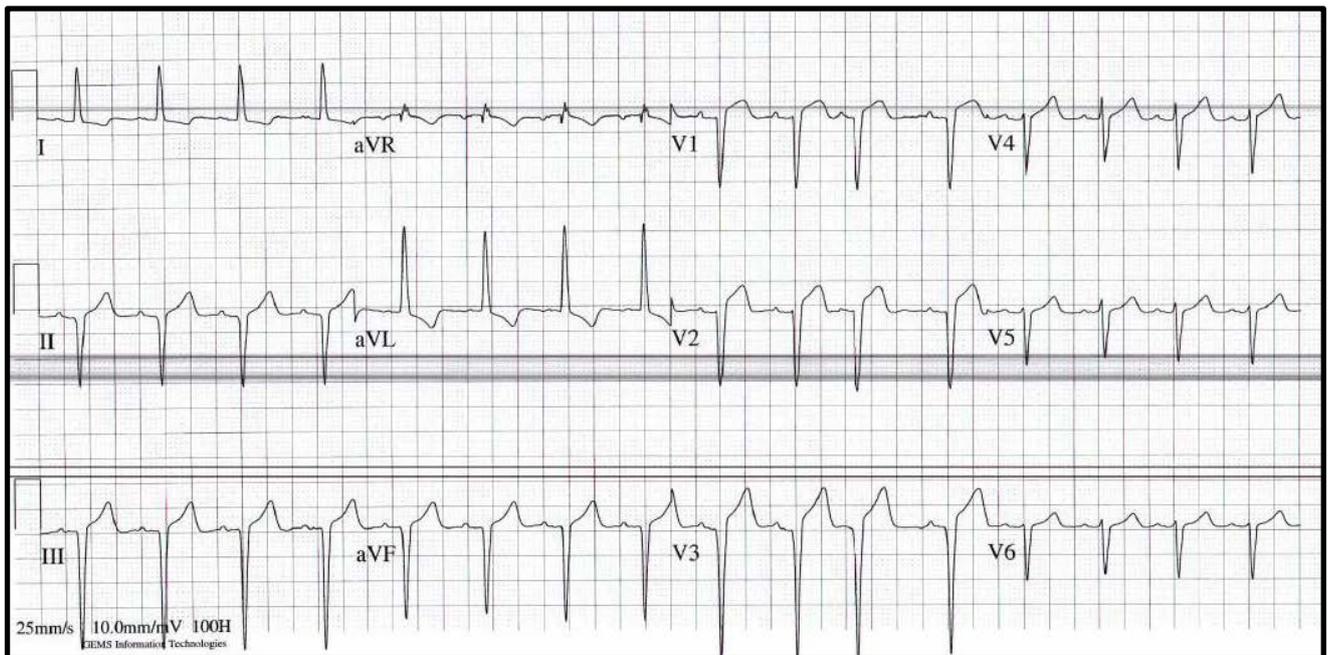


Image 2. EKG of the patient obtained 2 years ago showing shows NSR with narrow complexes and left ventricular hypertrophy

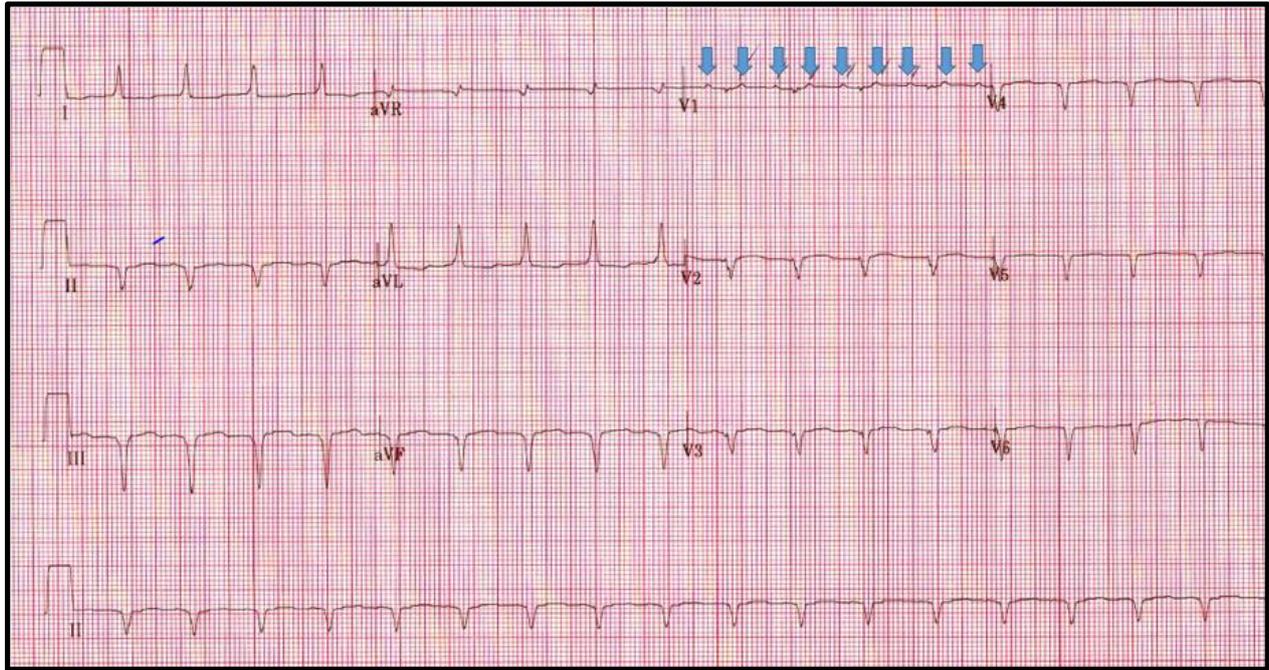


Image 3. EKG of the patient obtained during the present admission showing atrial tachycardia , pseudo-Q wave pattern in chest leads and low voltage QRS complexes in chest leads. P waves have been pointed with blue arrow

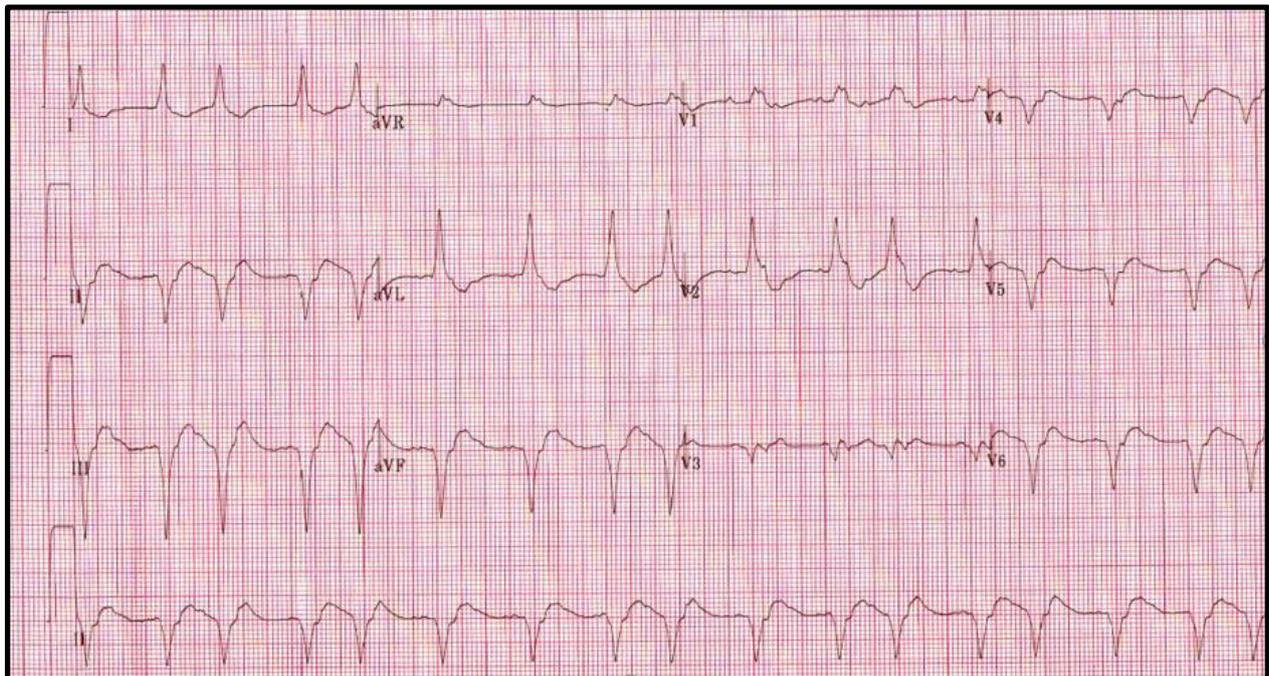


Image 4. EKG obtained subsequently revealing complete heart block(P waves and QRS complex are not related to each other in lead V1). Also right bundle branch block pattern and pseudo-Q waves may be noted in anterolateral leads

3. Discussion

IIM is a group of rare autoimmune disorder that includes polymyositis, dermatomyositis, inclusion body myositis and necrotizing myositis [1,2]. Cardiac involvement was thought to be a rare entity in IIM [4], however recent research suggest that cardiac involvement is the most common cause of morbidity and mortality in IIM [5]. Cardiac involvement is a poor prognostic factors in IIM [6]. Myocarditis is noted in upto 38% of the patients and

myocardial fibrosis in 22%. Dysrhythmias are reported in upto 32% of the cases [5]. Conduction abnormalities, angina, myocardial infarction and heart failure are the most common clinical presentation of cardiac involvement in IIM [4]. Arrhythmias are the most common cardiac presentation in patients with IIM. Up to 13.6% and 2.4% of IIM patients are noted to have arrhythmias as per prospective and retrospective cohorts respectively [5]. Arrhythmogenesis in IIM happens due to fibrosis of sino-atrial node, atrioventricular node or any other part of

the cardiac conduction system. Inflammation from the neighboring myocarditis can also lead to conduction abnormalities. Thus, IIM patient presenting with symptoms of arrhythmia or an arrhythmia noted in cardiac monitoring needs further evaluation and management [5]. A retrospective analysis reported heart failure in 11.7% of patients with IIM and accounts for 20% of cardiac deaths. Systolic and/or diastolic dysfunction is a result of recurrent myocarditis and myocardial fibrosis [5]. Myocardial infarction is noted in upto 6.8% of cases, but may be difficult to diagnose in IIM patients due to as CK-MB and Troponin T enzymes are also known to be elevated during skeletal muscle injury and recovery. Hence Troponin I levels must be obtained if myocardial ischemia is suspected in these patients [5]. Cardiac biopsy in IIM patients reveal revealed myonecrosis, edema, inflammatory infiltrate and patchy fibrosis [7]. IIM patients with cardiac involvement despite immunosuppressive therapy have a poor prognosis likely from advanced disease at the time of cardiac involvement [5]. This is the second case of atrial tachycardia reported in a patient with IIM [8]. Our patient had a normal EKG along with normal ejection fraction two years before but was noted to have atrial tachycardia on presentation and an ejection fraction of 25%. The patient then had a rapid progression of conduction disease and heart failure likely due to myocardial fibrosis and inflammation secondary to IIM. The patient's age group and cardiac involvement suggested poor prognosis. Early detection of arrhythmias and heart failure remains key in management of cardiac involvement in IIM. Physicians should be aware of the

rapid progression of conduction abnormalities and heart failure in IIM patients.

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