

Inducible Ventricular Tachycardia Due to Dermatomyositis-Related Cardiomyopathy in the Era of Implantable Cardioverter-Defibrillator Therapy

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A 67-year-old man with a history of dermatomyositis, under treatment with prednisolone and methotrexate for the past 10 years, complained of palpitations and episodes of dizziness. During the last 3 years, the patient has been under treatment with an angiotensin II type 1 receptor blocker and amiodarone for hypertension and ventricular ectopic beats, respectively. He never experienced chest pain either on exercise or at rest. At the present evaluation, his ECG image revealed sinus rhythm with fragmented QRS complexes in the inferior leads, a sign indicative of scar and delayed conduction in the corresponding area¹ (Figure 1). The laboratory tests for troponin I, creatine phosphokinase and its cardiac isoform CK-MB, and transaminases were all within normal limits. The performed 24-hour Holter monitoring revealed 6 episodes of nonsustained ventricular tachycardia (VT) with maximum ventricular rate of 157 bpm. The following echocardiography demonstrated a slight increase in left ventricular dimension, hypokinesia of the inferior wall,

and a mildly reduced ejection fraction, estimated at 45%. On the basis of the echocardiographic data, the patient underwent coronary angiography that did not demonstrate significant coronary stenoses.

To further evaluate the symptomatic ectopic ventricular activity, an electrophysiological study was performed subsequently. With the use of a standard protocol, after ventricular pacing and stimulation with 3 extrasystoles, a sustained monomorphic VT with hemodynamic instability was induced and interrupted only after electric cardioversion (override with rapid ventricular pacing was insufficient to terminate VT) (Figure 2). After this intriguing finding in the electrophysiological study, and given that ischemia was not the trigger for this malignant tachyarrhythmia, cardiac magnetic resonance imaging was scheduled. Indeed, no scar in the distribution of coronary arteries was observed, whereas a late gadolinium enhancement in the middle myocardial layer of the inferior wall was revealed, which is a finding compatible

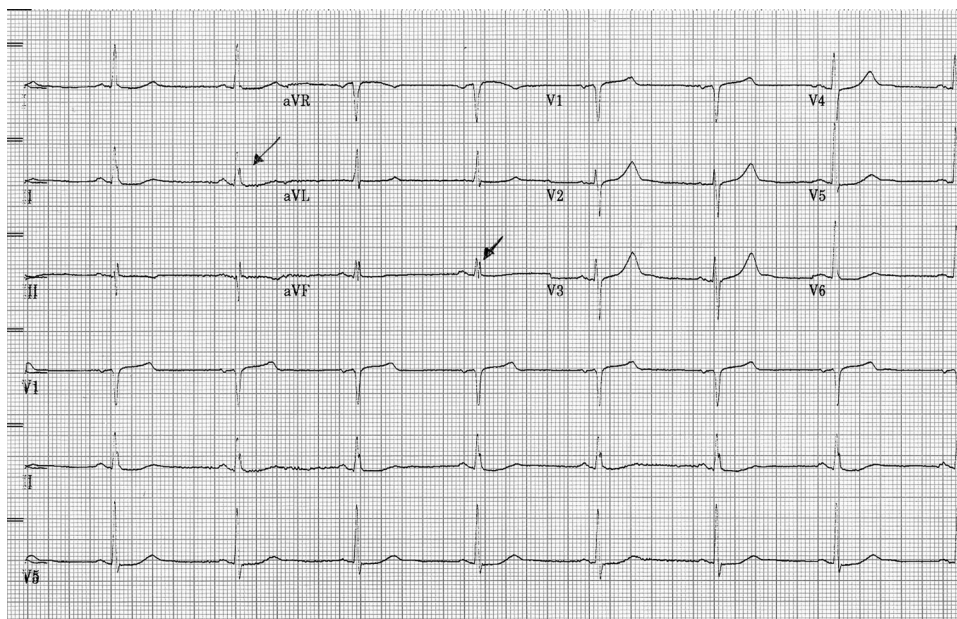


Figure 1. Resting ECG showing sinus rhythm with QRS fragmentation in the inferior leads (black arrows).

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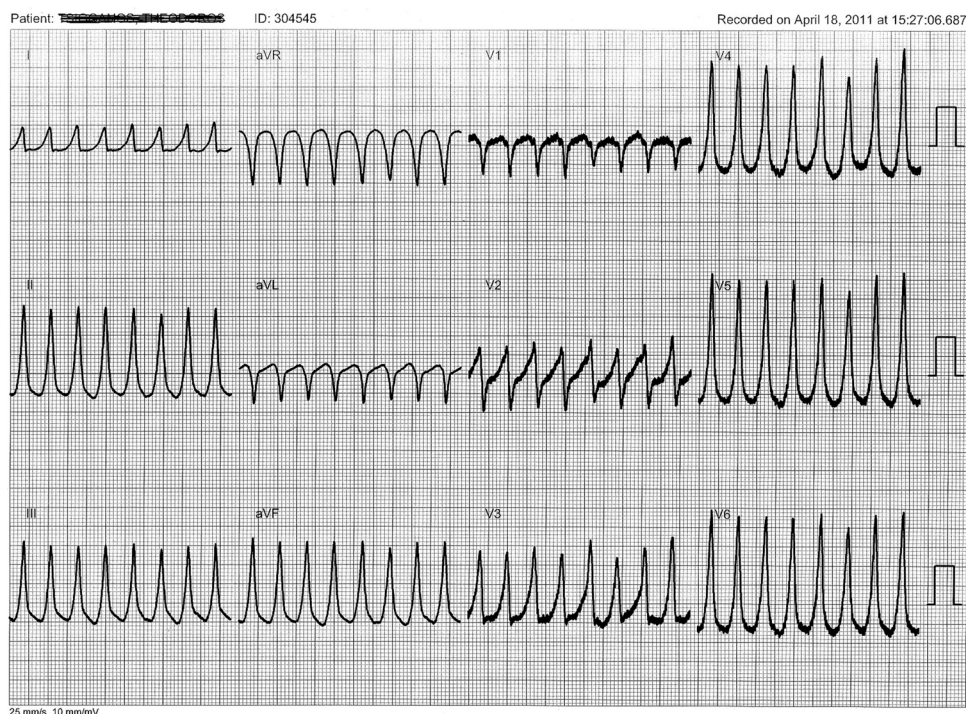


Figure 2. Inducible, monomorphic, sustained ventricular tachycardia during the electrophysiological study.

with a nonischemic, inflammatory process (Figure 3). Furthermore, the left ventricular ejection fraction, measured by magnetic resonance imaging, was 49%.

Our patient has a focal myocarditis in the inferior myocardial wall that most probably represents an extramuscular manifestation of dermatomyositis. In the literature, another case with undiagnosed dermatomyositis and syncope due to nonsustained VT has been reported, but the patient was not under treatment with corticosteroids or immunosuppressive agents, and no magnetic resonance imaging or electrophysiological study was performed to confirm the presence and the extent of the myocardial injury.²

Dermatomyositis belongs to idiopathic, inflammatory myopathies, an entity that also includes polymyositis and

inclusion-body myositis. Cardiovascular manifestations constitute a major cause of death in patients with myositis. The most frequent cardiac manifestations include left ventricular dysfunction due to myocarditis and arrhythmias, especially conduction abnormalities that may lead to third-degree atrioventricular block and syncope.³ Ventricular ectopic beats are frequent, but VT has been documented rarely.^{2,3} Although cardiac manifestations could be potentially lethal in patients with dermatomyositis, the prevalence of cardiac disorders and their therapeutic approach are not well established. From sporadic reports, it has been demonstrated that in patients with untreated dermatomyositis presenting with myocarditis, treatment with corticosteroids and immunosuppressive agents reduces the size of myocardial damage and improves wall

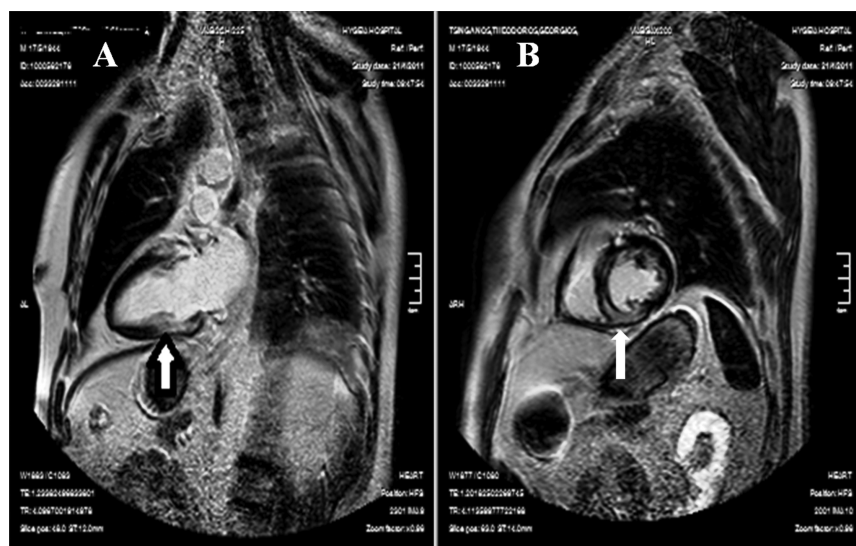


Figure 3. Cardiac magnetic resonance imaging, 2-chamber (A) and short-axis views (B), showing late gadolinium enhancement (LGE) in the middle myocardial layer of the inferior wall (arrows), compatible with a nonischemic inflammatory process.

hypokinesia.⁴ Nevertheless, no data exist regarding therapeutic interventions for patients who are already under treatment and in whom the disease is in remission, as was the case for our patient. With all the information taken into consideration and given that administration of antiarrhythmic agents probably would not add any benefit because our patient was already on treatment with amiodarone, we decided to proceed to implantable cardioverter-defibrillator therapy. Thus far, the follow-up period, 3 months after implantable cardioverter-defibrillator implantation, was uneventful.

In conclusion, inflammatory myopathies may insult the myocardium, leading to severe cardiac abnormalities that may constitute a major cause of death. Moreover, myocarditis in these patients may be silent and therefore misdiagnosed, especially when patients seem to “respond” to treatment with improvement of muscle strength and normalization of muscle enzymes. As the incidence of polymyositis/dermatomyositis

increases over the years, more careful evaluation of cardiac performance should be considered. In patients with ventricular ectopic beats and established myocardial damage, an electrophysiological study may provide a significant tool for risk stratification and appropriate management.

Disclosures

None.

References

1. Chatterjee S, Changawala N. Fragmented QRS complex: a novel marker of cardiovascular disease. *Clin Cardiol*. 2010;33:68–71.
2. Adler M, Banerjee S, Stratton R. Ventricular tachycardia as a presenting feature of dermatomyositis. *Heart*. 2002;88:441–443.
3. Lundberg IE. The heart in dermatomyositis and polymyositis. *Rheumatology*. 2006;45(suppl 4):iv18–iv21.
4. Allanore Y, Vignaux O, Arnaud L, Puéchal X, Pavy S, Duboc D, Legmann P, Kahan A. Effects of corticosteroids and immunosuppressors on idiopathic inflammatory myopathy related myocarditis evaluated by magnetic resonance imaging. *Ann Rheum Dis*. 2006;65:249–252.

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