

RECURRENT VENTRICULAR FIBRILLATION AND CARDIOGENIC SHOCK REVEALING CARDIAC INVOLVEMENT IN MYOTONIC DYSTROPHY: A CASE REPORT

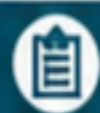
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BACKGROUND

Myotonic dystrophy is a multisystem genetic disorder associated with conduction abnormalities, ventricular arrhythmias, cardiomyopathy, and sudden cardiac death. Cardiac manifestations may remain clinically silent until a catastrophic event occurs. Early recognition and regular cardiac surveillance are essential to prevent life-threatening complications.



CASE PRESENTATION

- A male in his fourth decade of life collapsed suddenly during physical exertion at a family event. Immediate bystander cardiopulmonary resuscitation (CPR) was initiated.
- Upon arrival of emergency medical services, the initial documented rhythm was ventricular fibrillation (VF). Return of spontaneous circulation (ROSC) was achieved after eight defibrillation shocks. The patient was intubated and transferred to the emergency department.
- On admission, he was in profound cardiogenic shock. Venous blood gas analysis demonstrated severe mixed lactic acidosis. Laboratory testing revealed no significant anemia or electrolyte abnormalities. Electrocardiography showed no evidence of acute myocardial infarction.
- The patient subsequently suffered a second cardiac arrest. Initial rhythm was asystole, followed by VF during resuscitation. ROSC was achieved after approximately 15 minutes of advanced life support, including one additional defibrillation shock.
- Following the second resuscitation, the patient remained hemodynamically unstable and severely hypoxicemic.
- Bedside transthoracic echocardiography demonstrated global myocardial stunning with moderately-to-severely reduced left ventricular systolic function, without significant valvular abnormalities. Inferior vena cava dimensions were normal.
- Chest radiography revealed a right-sided pneumothorax, most likely traumatic secondary to CPR.
- Given the unclear etiology of recurrent cardiac arrest, urgent non-contrast head CT and CT pulmonary angiography were performed. No intracranial pathology or pulmonary embolism was identified. Imaging demonstrated a moderate-to-large right-sided pneumothorax, for which a chest tube was inserted with subsequent radiographic improvement.
- Review of available medical records revealed a previous diagnosis of **myotonic dystrophy**; however, the patient had not been followed regularly and the specific subtype could not be determined.
- The patient was admitted to the intensive cardiac care unit (ICCU).
- Formal transthoracic echocardiography demonstrated severe left ventricular systolic dysfunction with an ejection fraction (EF) of approximately 20%.
- Coronary CT angiography demonstrated normal coronary arteries without obstructive coronary artery disease.
- Cardiac magnetic resonance imaging demonstrated myocardial abnormalities consistent with cardiac involvement in myotonic dystrophy.



MANAGEMENT

- Following multidisciplinary discussion involving cardiology, intensive care, and heart failure specialists, escalation to mechanical circulatory support (VA-ECMO or Impella) was deferred because of gradual hemodynamic improvement.
- The patient was treated with norepinephrine, vasopressin, and milrinone. Over the following days, vasopressor and inotropic support were successfully discontinued.
- Guideline-directed medical therapy for heart failure was initiated, and a cardiac resynchronization therapy defibrillator (CRT-D) was implanted for secondary prevention of sudden cardiac death.



OUTCOME

- The patient was successfully extubated.
- No neurological deficits were observed.
- After two weeks of hospitalization, he was discharged in satisfactory clinical condition.
- Repeat echocardiography at discharge showed significant improvement in left ventricular systolic function, with ejection fraction increasing from 20% to 40%.

EF
20%

On admission



EF
40%

At discharge



KEY IMAGING

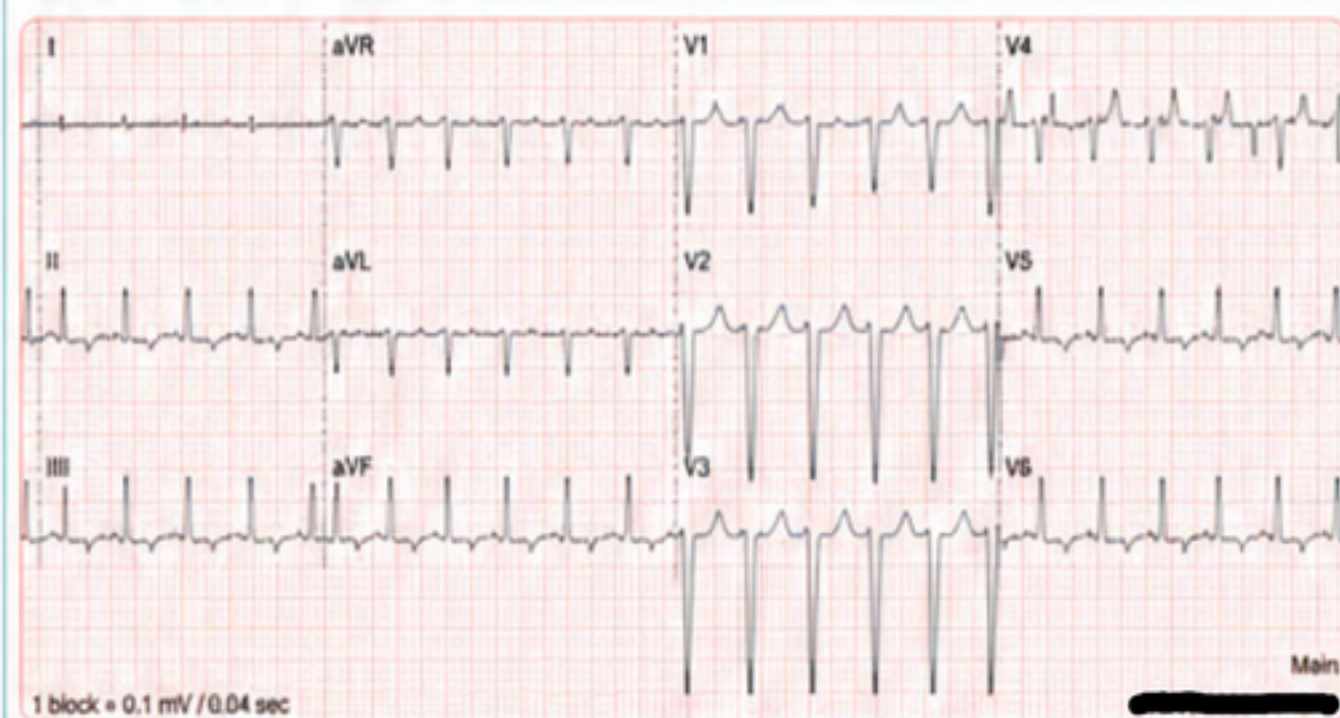


Figure 1. Admission 12-lead electrocardiogram demonstrating intraventricular conduction abnormalities without evidence of acute ST-segment elevation myocardial infarction.

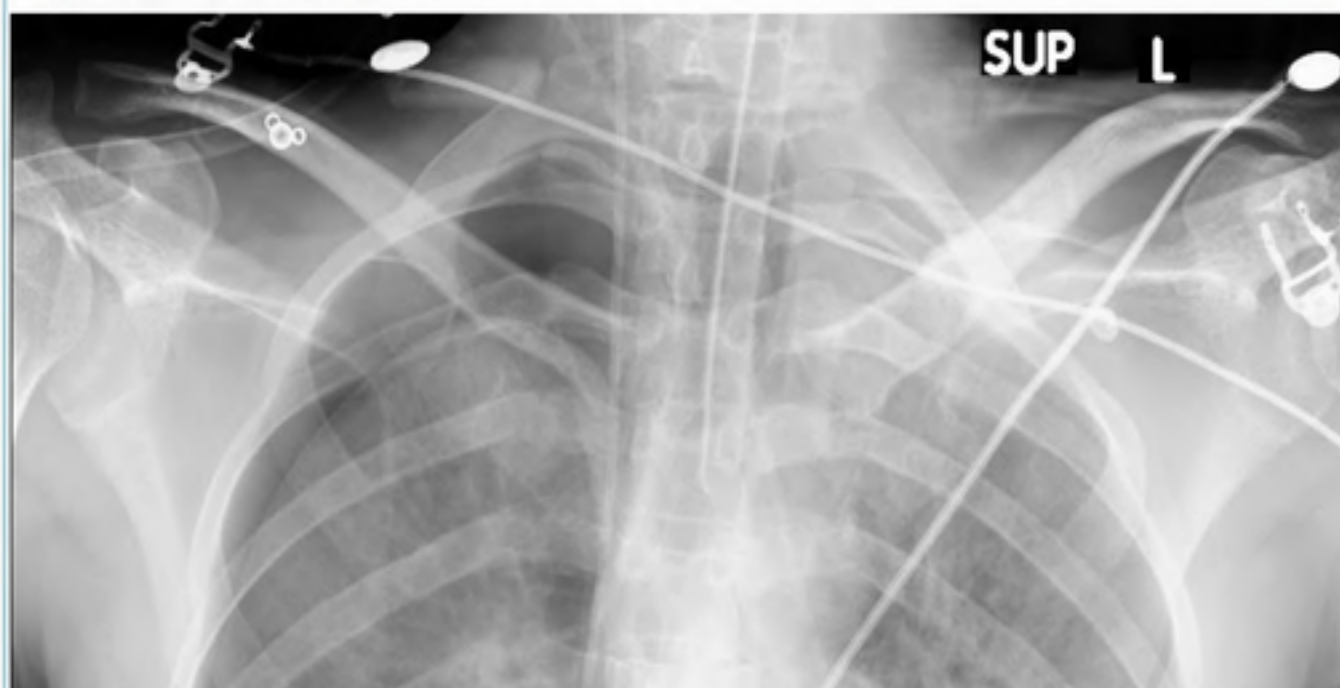


Figure 2. Portable chest radiograph demonstrating a right-sided pneumothorax.

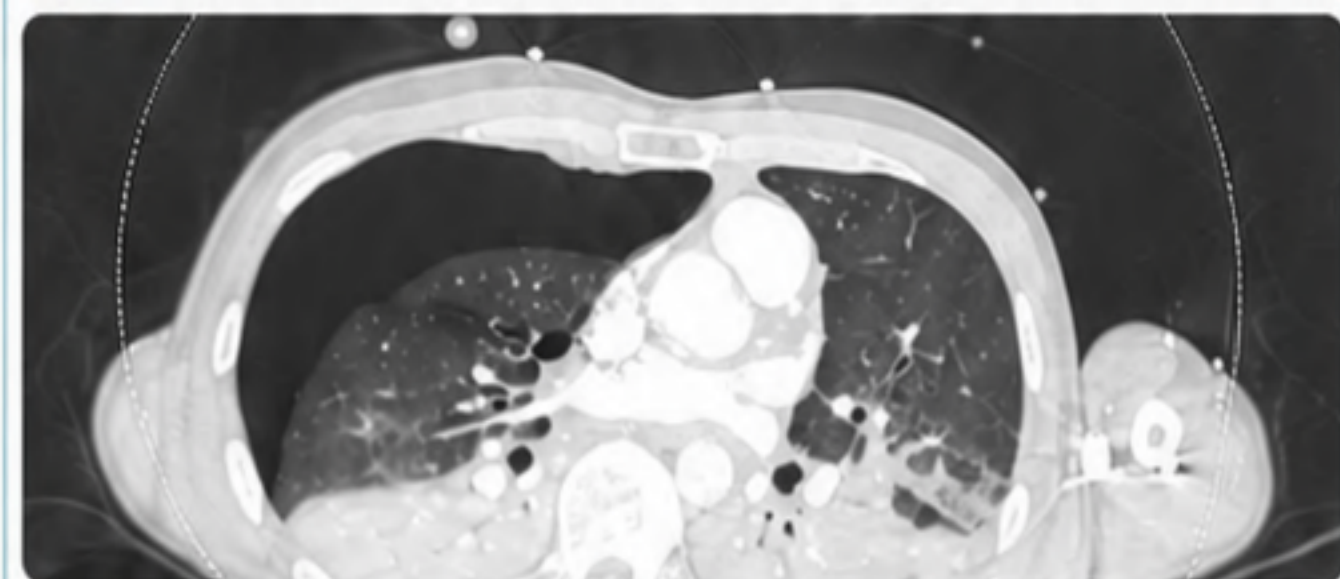


Figure 3. CT pulmonary angiography demonstrating a moderate-to-large right-sided pneumothorax, most likely traumatic secondary to CPR, without evidence of pulmonary embolism. Bilateral pulmonary consolidations most consistent with aspiration are also present.



LEARNING POINTS

- Sudden cardiac arrest may be the first manifestation of cardiac involvement in myotonic dystrophy.
- Ventricular fibrillation can occur in the absence of obstructive coronary artery disease.
- Early multidisciplinary management may result in favorable neurological and cardiac recovery despite profound cardiogenic shock.
- Routine cardiac surveillance should be considered in all patients with myotonic dystrophy.