ISSN: 2693-4965
DOI: 10.33552/OJCRR.2024.07.000673
Online Journal of Cardiology Research & Reports
Iris Publishers

Case Report

Myotonic Dystrophy Presenting as Atrial Flutter in Childhood: Case Report and Review of the Literature

Yousra Abbou-ou-cherif* and Alae Eddine El Bouanani

Cardiology department, Chartres hospital center, France

***Corresponding author:** Yousra Abbou-ou-cherif, Cardiology department, Chartres hospital center, France.

Received Date: July 15, 2024

Published Date: July 30, 2024

Abstract

Steinert's disease, or myotonic dystrophy type 1 (DM1), is an autosomal dominant genetic disorder that can affect both children and adults. It is characterized by myotonia and damage to various organs, including the heart. Cardiac damage is the most severe systemic impairment, significantly impacting the overall prognosis. It accounts for approximately one-third of the deaths in this population. The most common cardiac consequences are cardiomyopathy and arrhythmias, which often develop later in the course of the disease.

If cardiac arrhythmias are a cardinal prognostic marker in adults with DM1 [1-2], they are nevertheless considered rare in young people and have rarely been studied. We report the case of a 17-year-old boy suffering from Steinert's disease since the age of 11, with good follow-up that has revealed no cardiac damage so far. He experienced two episodes of palpitations and chest pain following physical exercise. In the emergency room, the electrocardiogram revealed a typical atrial flutter, indicating cardiac involvement.

Keywords: Steinert's disease, myotonic dystrophy type 1, Cardiac manifestations.

Case Report

A 17-year-old boy was taken to the emergency department after feeling sick following physical exercise, experiencing chest pain. He also reported two episodes of recurrent palpitations within the previous month. The initial electrocardiogram revealed atrial flutter, resulting in a heart rate of 160 bpm (Figure 1). Clinical examination showed no abnormalities, and echocardiography demonstrated moderately reduced left ventricular function with an ejection fraction of 45%, but no signs of a congenital heart defect.

Beta-blocking agents were able to sufficiently slow the heart rate, reducing it to 120 beats/min. Subsequently, catheter ablation of the arrhythmia was performed to prevent future cardiac decompensations and the development of rhythmic heart disease. The patient was discharged, and a cardiac MRI was requested, which did not reveal any signs of myocarditis, pericarditis, or any other cardiac damage associated with myotonic dystrophy. After 6 months of follow-up, no recurrence of the arrhythmia could be observed, and the echocardiogram showed a remarkable improvement in cardiac function.

Discussion

Steinert's disease or DM1 is the most common hereditary muscular dystrophy of adults, with a prevalence of 1 to 10 cases per 100,000 populations [3]. It is an autosomal dominant genetic disease characterized by myotonia and multisystem involvement: muscular, neurological, ocular, respiratory, digestive, endocrine,

and cardiac. Cardiac involvement is frequent, occurring in 80% of patients, and may precede muscular involvement. Although often asymptomatic, it conditions the vital prognosis and represents the second most frequent cause of death, after respiratory causes [4-5].

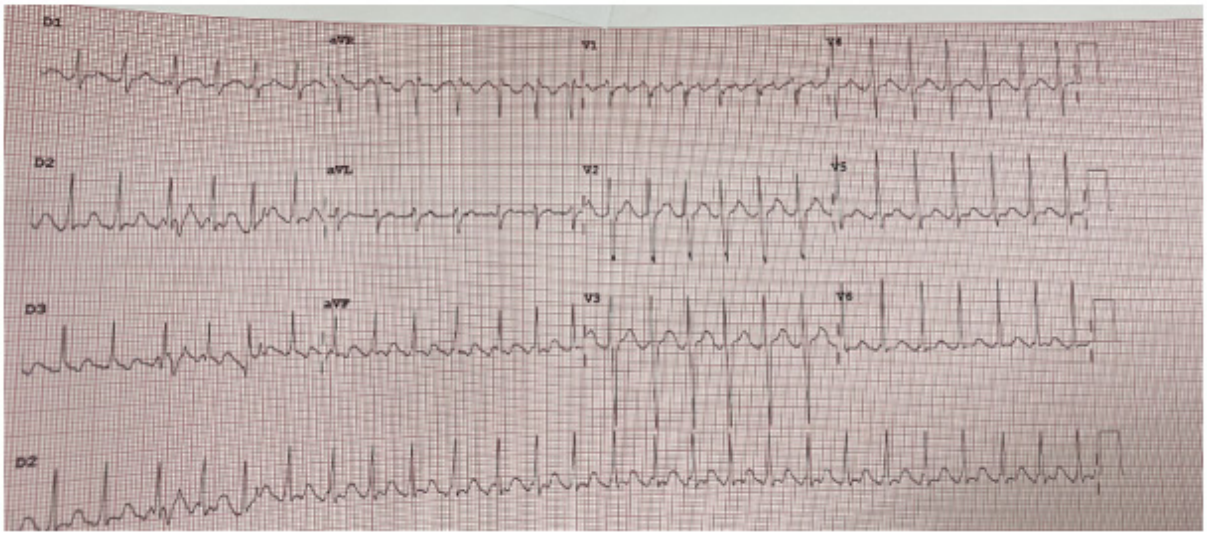

Figure 1: Electrocardiogram showed atrial flutter with 2:1 atrioventricular conduction giving a rate of 160 beats/min.

Endomyocardial biopsies in DM1 patients show specific changes such as perivascular interstitial fibrosis, fatty infiltration, myocardial hypertrophy, and sometimes focal myocarditis [6].

Cardiac mortality usually occurs as a result of progressive left ventricular dysfunction, sudden death, ischemic heart disease, or, more rarely, pulmonary embolism [7-8]. Sudden cardiac death can be caused by ventricular asystole, degeneration of ventricular tachycardia (VT), ventricular fibrillation (VF), or electromechanical dissociation [9-10]. Ventricular arrhythmias, although less frequent, are naturally of greater concern because of their lethal potential.

Atrial arrhythmias, in particular, atrial fibrillation, atrial flutter, and atrial tachycardia are the most common, affecting around a quarter of patients. Conduction disorders are also very common in myotonic dystrophy, affecting around 30-75% of cases [9]. Other cardiac disorders include structural heart disease in around 20% of patients, manifested by signs of heart failure, mitral valve prolapse in 13 to 40% of cases, and relaxation disorders [11-13].

Cardiac involvement can sometimes be the first manifestation of the disease, even in the absence of overt neuromuscular involvement. It is estimated that 65% of DM1 patients have an abnormal ECG [14]. Every cardiologist needs to be aware of this diagnosis.

In the pediatric population, the prevalence of cardiac manifestations of the disease has not been estimated in large populations. On the other hand, several case reports have been published on the occurrence of rhythm disorders, exclusively reported after the age of 10 and predominantly triggered by exercise. More often, these were atrial arrhythmias and, more rarely, sustained ventricular tachyarrhythmias, which can lead to sudden death in children [15].

This implies that early detection of cardiac damage should be considered after the age of 10 in all children with myotonic dystrophy, especially if the child takes part in sporting activities. A 12-lead ECG and echocardiography should retain the same indications as in adults.

Conclusion

While it is evident that the risk of arrhythmia, including sudden death, is heightened in Steinert's disease, a clear consensus on management has not been established. Although debated, it has been suggested that an annual surface ECG and echocardiography should be conducted. Good collaboration between neurologists and cardiologists is also crucial for effective patient management.

Acknowledgement

None.

Conflict of Interest

No Conflict of Interest.

References

1. Duboc D, Eymard B, Damian MS (2004) Cardiac management of myotonic dystrophy. In: Harper PS, Van Engelen B, Eymard B, Wilcox D [eds]. Myotonic dystrophy: present management, future therapy. Oxford: Oxford University Press 85-93.

2. Lazarus A, Varin J, Ounnoughene Z, Radvanyi H, Junien C, et al. (1999) Relationships among electrophysiological findings and clinical status, heart function, and extent of DNA mutation in myotonic dystrophy. Circulation 99(8): 1041-1046.

3. Bouhour F, Bost M, Vial C (2007) Maladie de Steinert. Presse Med (36): 965-971.

4. Mankodi A, Thorton C (2002) Myotonic syndrome. Curr Opin Neurol 15: 545-552.

5. D'Andrea A, Salerno G, Scarafile R, Riegler L, Gravino R, et al. (2009) Right ventricular myocardial function in patients with either idiopathic or ischemic dilated cardiomyopathy without clinical sign of right heart failure: effects of cardiac resynchronization therapy. Pacing Clin Electrophysiol 32(8): 1017-1029.

6. Phillips MF, Harper PS (1997) Cardiac disease in myotonic dystrophy. Cardiovasc Res33: 13-22.

7. Magrì D, Piccirillo G, Bucci E, Pignatelli G, Maria Cauti F, et al. (2012) Increased temporal dispersion of myocardial repolarization in myotonic dystrophy Type 1: Beyond the cardiac conduction system. Int J Cardiol 156(3): 259-264.

8. Nigro G, Russo V, Rago A, Antonio Papa A, Della Cioppa N, et al. (2012) Heterogeneity of ventricular repolarization in newborns with severe aortic coarctation. Pediatr Cardiol 33(2): 302-306.

9. Groh WJ, Groh MR, Chandan S, Kincaid JC, Simmons Z, et al. (2008) Electrocardiographic abnormalities and sudden death in myotonic dystrophy type 1. N Engl J Med 358(25): 2688-2697.

10. Nigro G, Nigro G, Politano L, et al. (2002) Is the value of QT dispersion a valid method to foresee the risk of sudden death? A study in Becker patients. Heart 87: 156-157.

11. Melacini P, Villanova C, Menegazzo E, Novelli G, Danieli G, et al. (1995) Correlation between cardiac involvement and CTG trinucleotide repeat length in myotonic dystrophy. J Am Coll Cardiol 25(1):239-245.

12. Wahbi K, Meune C, Becane HM, Laforet P, Bassez G, et al. (2009) Left ventricular dysfunction and cardiac arrhythmias are frequent in type 2 myotonic dystrophy: a case control study. Neuromuscul Disord 19(7): 468-472.

13. Bhakta D, Lowe MR, Groh WJ (2004) Prevalence of structural cardiac abnormalities in patients with myotonic dystrophy type I. Am Heart J 147(2): 224-227.

14. Nguyen HH, Wolfe JT, 3rd, Holmes DR, Jr, Edwards WD (1988) Pathology of the cardiac conduction system in myotonic dystrophy: a study of 12 cases. J Am Coll Cardiol 11(3): 662-671.

15. Bassez G, Lazarus A, Desguerre I, Varin J, Laforêt P, et al. (2004) Severe cardiac arrhythmias in young patients with myotonic dystrophy type 1. Neurology 63(10): 1939-1941.