

Paper-like bilateral sternocleidomastoid muscle atrophy may suggest the occurrence of sustained ventricular tachycardia

Keiji Matsunaga, MD, Yuichi Miyake, MD, PhD, Takahisa Noma, MD, PhD, Tetsuo Minamino, MD, PhD

From the Department of Cardiorenal and Cerebrovascular Medicine, Faculty of Medicine, Kagawa University, Kagawa, Japan.

Introduction

A paper-like bilateral sternocleidomastoid muscle atrophy on computed tomography images, which is characteristic of myotonic dystrophy, could help diagnose the cause of sustained ventricular tachycardia (VT), despite the absence of typical clinical characteristics.

Case report

A 43-year-old man with a sustained wide QRS complex tachycardia and a left ventricular ejection fraction of 40% was referred to our hospital (Figure 1). We diagnosed this arrhythmia as VT from the findings of atrioventricular dissociation and QRS morphology in V_6 (based on the Brugada criteria¹); electrical cardioversion was performed to treat VT. We believe the VT origin was the middle to the apex of the inferior region, according to the findings of the electrocardiogram and late gadolinium-enhanced cardiac magnetic resonance imaging (Supplemental Figure 1).

Unexpectedly, the chest computed tomography (CT) to evaluate pulmonary edema and pleural effusion revealed paper-like bilateral sternocleidomastoid muscle atrophy (Figure 2). Careful physical examination of the neck following the detection of paper-like bilateral sternocleidomastoid muscle atrophy on the CT images did not reveal clinical findings of bilateral atrophy, such as neck flexor weakness and rotation disorder of the head. Because these CT images are characteristic of myotonic dystrophy, we performed a genetic test and confirmed the expansion of a cytosine-thymine-guanine repeat of the myotonic dystrophy protein kinase gene. Initially, we administered amiodarone (150 mg once

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KEY TEACHING POINTS

- Myotonic dystrophy must always be considered as the potential cause of ventricular tachycardia (VT), as it is associated with the risk of sudden death caused by asystole following atrioventricular block or VT in patients with myotonic dystrophy.
- Bilateral sternocleidomastoid muscle atrophy is one of the characteristic findings of myotonic dystrophy, and the range of prevalence of myotonic dystrophy ranges from approximately 1:100,000 in some areas of Japan to approximately 1:10,000 in Iceland, with a European prevalence of 3–15 per 100,000.
- Close attention to a paper-like bilateral sternocleidomastoid muscle atrophy on computed tomography images could help diagnose VT with muscular dystrophy.

daily) and carvedilol (1.25 mg twice daily) for treating VT; however, significant bradycardia occurred. Furthermore, his signal-averaged electrocardiogram showed abnormal findings. Based on these findings, he underwent cardiac resynchronization therapy with a defibrillator (CRT-D). After CRT-D implantation, the occurrence of VT with the same morphology was observed, and we confirmed atrioventricular dissociation during VT, using intracardiac electrocardiograms again (Supplemental Figure 2). Since the exclusion of the junctional ectopic tachycardia with complete left bundle branch block was difficult only from the 12-lead electrocardiograms, we finally diagnosed this arrhythmia as VT from the poor QRS morphology correspondence rate between this arrhythmia and the sinus rhythm from intracardiac electrocardiograms (Supplemental Figure 2, white arrows).

Address reprint requests and correspondence: Dr Tetsuo Minamino, Department of Cardiorenal and Cerebrovascular Medicine, Faculty of Medicine, Kagawa University, 1750-1 Ikenobe, Mikicho, Kita-gun, Kagawa 761-0793, Japan. E-mail address: minamino@med.kagawa-u.ac.jp.



Figure 1 A: Electrocardiography on admission. White arrows show dissociated P waves. B: Electrocardiography after cardioversion.

Bilateral sternocleidomastoid muscle atrophy is one of the characteristic findings of myotonic dystrophy, and the range of prevalence of myotonic dystrophy is approximately 1:100,000 in some areas of Japan to approximately 1:10,000 in Iceland, with a European prevalence of 3–15 per 100,000.² Myotonic dystrophy, although rare, must always be considered as the potential cause of VT, as it is associated with the risk of sudden death caused by asystole following atrioventricular block or VT in patients with myotonic dystrophy.³ However, myotonic dystrophy is often underdiagnosed, because the muscle atrophy is compensated by slow disease progression.

Close attention to a paper-like bilateral sternocleidomastoid muscle atrophy on CT images could help diagnose VT with muscular dystrophy, despite the absence of typical clinical characteristics such as neck flexor weakness and rotation disorder of the head.



Figure 2 Computed tomography at the level of the larynx.

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Appendix Supplementary data

Supplementary data Supplementary data associated with this article can be found in the online version at https://doi.org/1 0.1016/j.hrcr.2019.10.017.

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