Idiopathic Ventricular Fibrillation Associated with Complete Right Bundle Branch Block?

Mamoru Manita¹, Tadashi Nakajima², Yoshiaki Kaneko² and Masahiko Kurabayashi²

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SCN5A 5432-5433insGAGT

Picture.

A 46-year-old man was referred to our hospital due to cardiac arrest caused by ventricular fibrillation (VF). An electrocardiograms (ECG) showed complete right bundle branch block (CRBBB) without ST elevation in the right precordial leads (Picture A). An ECG recorded 12 months earlier had also showed the same pattern (Picture B). Inter-

¹Department of Cardiology, Naha City Hospital, Japan and ²Department of Medicine and Biological Science, Gunma University Graduate School of Medicine, Japan

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Correspondence to Dr. Tadashi Nakajima, tnakajim@med.gunma-u.ac.jp

estingly, a pilsicainide provocation test induced J point elevation in the V1 lead without the typical coved-type ST elevation observed in Brugada syndrome (BrS) (Picture C). Echocardiography, cardiac magnetic resonance imaging and left cardiac catheterization revealed no structural heart disease or vasospastic angina. Programmed ventricular stimulation induced VF. The patient's father had died suddenly at 58 years of age.

Aizawa et al. showed that CRBBB is more prevalent in patients with idiopathic ventricular fibrillation (IVF) than in control subjects (1) and that CRBBB masks an ECG morphology characteristic of BrS (2). The present patient was thought to have IVF associated with CRBBB (IVF-CRBBB)

rather than BrS. We identified a novel SCN5A mutation, 5432-5433insGAGT (Picture D), illustrating a genetic background of IVF-CRBBB.

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References

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