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Case report

Cochlear implant function in a patient with Jervell and Lange-Nielsen syndrome after defibrillation by countershock

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ABSTRACT

Jervell and Lange-Nielsen syndrome (JLNS), a rare autosomal recessive congenital QT prolongation syndrome, is characterized by cardiac arrhythmias, syncopal episodes, and profound deafness. A cochlear implant (CI) for patients with JLNS is expected to result in hearing improvement. Sometimes, defibrillation is required if a patient experiences lethal arrhythmia. In this paper, we report a pediatric patient with JLNS who received defibrillation after CI surgery in his right ear at the age of 2 years. With intensive care, the post-operative course was uneventful, and the patient acquired satisfactory speech and hearing abilities. Five years after the surgery, he underwent defibrillation because of the incidence of syncopal attack. Thereafter, arrhythmic syncope recurred three times, which necessitated defibrillation therapy. To prevent recurrence of cardiac arrhythmia, he underwent ICD (implantable cardioverter-defibrillator) implantation at the age of 11 years. At present, CI works well and provides good hearing, while syncopal attack is prevented by ICD. From the experience of this case, electronic circuit of CI is thought to tolerate emergency countershock if the speech processor is removed.

1. Introduction

Jervell and Lange-Nielsen syndrome (JLNS), a rare autosomal recessive congenital QT prolongation syndrome, is characterized by cardiac arrhythmias, syncopal episodes, and profound deafness [1]. Patients with JLNS are expected to develop a good hearing ability with the placement of a cochlear implant (CI). Although these patients have a high probability of receiving defibrillation after CI surgery, only a little is known concerning the effects of countershock on cochlear implant [2,3]. In this paper, we report a case of pediatric JLNS patient who underwent CI surgery. He exhibited frequent syncopal attacks after the surgery which necessitated countershock for control. Even after defibrillation, CI continued to function properly, and the patient's hearing remains good.

2. Case

A 6-month-old male patient was referred to our department because of bilateral hearing loss. Auditory brainstem response was absent on both sides at 105 dB nHL. The patient was diagnosed through ultrasonography *in utero* as having a congenital arrhythmia, and electrocardiography (ECG) detected a prolonged QT interval (549 ms, Fig. 1). Immediately after birth, β-blocker therapy was initiated using oral atenolol. On the basis of the congenital sensorineural deafness and long QT interval, the patient was diagnosed with JLNS; this diagnosis was later confirmed by genetic testing, which reported variations in the KCNQ1 gene. The patient began wearing

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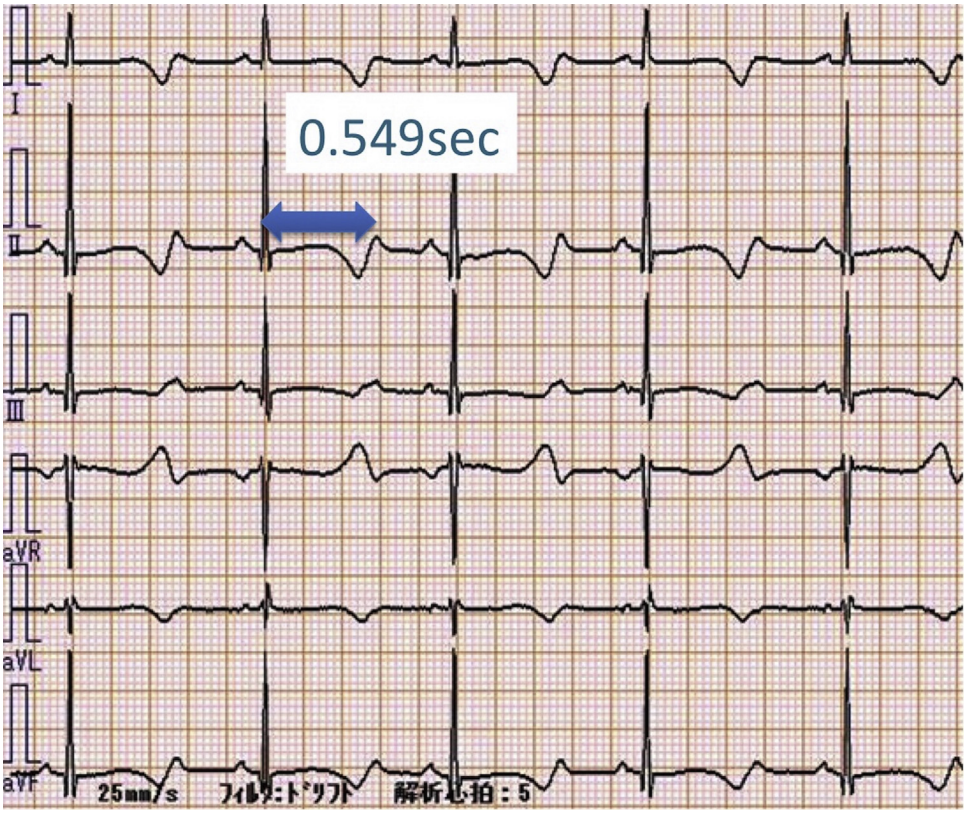

Fig. 1. Patient's electrocardiogram, indicating a prolonged QT interval (549 ms).

hearing aids at the age of 1 year. However, its effect was limited. Therefore, he underwent a CI surgery in the right ear at the age of 2 years. Meticulous care was taken before, during, and after the surgery to avoid stress that could precipitate lethal arrhythmia. After the surgery, the patient acquired satisfactory speech and listening abilities. The pure tone average was 26.3 dB HL in a free-field condition, and the speech discrimination score was 90%. Five years after the surgery, the patient had four syncopal episodes. Defibrillation with a biphasic electrical waveform was delivered immediately after the first syncopal attack. Even after defibrillation, the CI functioned properly and the patient's hearing remained preserved. There were no increases in impedance on any electrodes (Fig. 2). No changes in CI mapping were needed. At 11 years of age, the patient underwent implantable cardioverter-defibrillator (ICD) surgery. The postoperative course was uneventful. The defibrillation threshold was set with a direct current of 20 J. Six months after the surgery, the ICD was activated because of ventricular fibrillation. After this event, the CI continued to work properly and the patient's hearing remained preserved.

3. Discussion

JLNS is caused by mutations in the KCNQ1 and KCNE1 genes [1]. The KCNQ1 gene encodes the alpha subunit and the KCNE1 gene the beta subunit of Kv7.1 [4]. Kv7.1 is a voltage-gated potassium channel that is found throughout the body [5]. In our case, genetic test revealed mutation in the KCNQ1 gene. In the inner ear, Kv7.1 channels are present in the membrane of the stria vascularis. Although mutation in the KCNQ1 gene causes degeneration of the spiral ganglion in mouse [6], implantation of CI provided good hearing in our patient as same in Siem et al's case [7].

CI surgery in JLNS patient owns two critical risks: one is the possibility to induce lethal arrhythmia during surgery and the other is the possibility to damage the electronic circuit of CI and/or the inner ear tissue when a defibrillator is used for treatment of arrhythmic syncope. First problem is at most important. Medical staffs should always be aware of and prepare for the occurrence of lethal arrhythmias during surgery. Administration of a β-blocker is the first-line treatment for lethal arrhythmias [8]. Although β-blocker therapy cannot always prevent lethal arrhythmias, it is effective to decrease mortality rate due to arrhythmic syncope. Up to 25% of patients still suffer from arrhythmia even after β-blocker treatment. In such cases, ICD implantation is suggested. In cases with refractory arrhythmias after ICD implantation, left cardiac sympathetic denervation is another treatment option. In our case, the patient was treated with a β-blocker at the time of CI surgery. During the surgery, arrhythmia occurred several times but was not lethal, and the CI device was successfully implanted. The second problem is that JLNS patients with CI have a high likelihood of receiving defibrillation after the CI surgery. We could find only two papers describing the effect of electrical defibrillation on the CI. Busse et al. reported a patient who received defibrillation. In their report, a 61-year-old male underwent electrical defibrillation (500 J). Despite the need for

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