Case Report

Successful radiofrequency catheter ablation of sinoatrial nodal reentrant tachycardia after a total cavo-pulmonary connection procedure with an extra-cardiac conduit

Naoaki Onishi (MD)*, Kazuaki Kaitani (MD), Yukiko Hayama (MD), Chisato Izumi (MD, FJCC), Yoshihisa Nakagawa (MD)
Division of Cardiology of Tenri Hospital, Nara, Japan

A R T I C L E   I N F O
Article history:
Received 19 January 2015
Received in revised form 15 May 2015
Accepted 22 May 2015

Keywords:
Total cavo-pulmonary connection procedure
Extra-cardiac conduit
Heterotaxy syndrome
Sinoatrial nodal reentrant tachycardia
Radiofrequency catheter ablation

A B S T R A C T
We report a tachyarrhythmia case of a 32-year-old female with a single ventricle and heterotaxy syndrome. She had surgery involving a total cavo-pulmonary connection procedure using an extra-cardiac conduit (EC) at the age of 17 years. A tachycardia was repetitively induced with single atrial extrastimuli. An activation map was created revealing a centrifugal propagation pattern from the high atrial wall adjacent to the EC. At that site, a structure resembling the crista terminalis was recognized with intracardiac echocardiography. Therefore, high output energy was required to eliminate the tachycardia. It was thought to be a sinoatrial nodal reentrant tachycardia.

Learning objective: Radiofrequency catheter ablation (RFCA) of supraventricular tachycardia (SVT) after a total cavo-pulmonary connection with an extra-cardiac conduit is challenging and the diagnosis of the SVT is difficult. However the electrophysiological features observed during the electrophysiological study using a three-dimensional mapping system, anatomical features observed with intracardiac echocardiography, and pharmacological features seen during a rapid intravenous injection of adenosine triphosphate can lead to an accurate diagnosis, and moreover lead to a successful RFCA.

© 2015 Japanese College of Cardiology. Published by Elsevier Ltd. All rights reserved.

Introduction

A total cavo-pulmonary connection (TCPC) is a palliative surgical procedure resulting in a total right heart bypass for patients with complex congenital heart disease (CHD) [1]. A TCPC with an extra-cardiac conduit (EC) has a lower incidence of post-surgical tachyarrhythmias than other types of TCPCs. However, some intrinsic arrhythmogenic conditions, such as heterotaxy syndrome, especially a right isomerism, are still associated with a high incidence of arrhythmias [2]. However, the anatomical complexity after a TCPC with an EC causes a major difficulty for radiofrequency catheter ablation (RFCA) [3].

Furthermore, cases in which the tachycardia after the TCPC is sinoatrial nodal reentrant tachycardia (SANRT) have rarely been reported. We describe a successful RFCA case of SANRT in a patient with heterotaxy syndrome after a TCPC.

Case report

A 32-year-old female was referred to our institute for RFCA therapy of a tachycardia. She had a single ventricle morphology, common atrioventricular (AV) valve, and heterotaxy syndrome (right isomerism and dextrocardia). She underwent staged palliations with a Blalock–Taussig shunt at 3 years old, and then a TCPC using an EC at 17 years old (Fig. 1a and b). She began to feel palpitations 4 years previously. Although it had sensitivity to verapamil, it became refractory and more frequent. Therefore she was admitted to our hospital for RFCA in November 2013.

The patient’s vital signs were normal and her SpO2 was 98% (room air). In a 12-lead electrocardiogram (ECG) series, two types of P wave polarities were observed. One was positive in lead I and flat in lead aVL, and another was negative in leads I and aVL. That indicated that she had two sinus nodes.

During the electrophysiological study (EPS), no ventriculoatrial (VA) conduction was observed, and the tachycardia [cycle length (CL); 390 ms] was repetitively induced with single atrial extrastimuli. There was an inverse correlation between the coupling interval of the atrial extra stimulation and the return CL from the
stimulation to the tachycardia. The P wave morphology of the tachycardia exhibited a low voltage with a wide width, however the P wave polarity of the tachycardia was almost similar to that of sinus rhythm (Fig. 1c).

A rapid intravenous injection of 10 mg of adenosine triphosphate (ATP) prolonged the atrial CL with a change in the subsequent ventricular CL of the tachycardia prior to termination. An EPS revealed evidence of twin AV nodes, however the ventricle was not involved in the tachycardia.

Via a Brockenbrough puncture from the EC into the atrium with intracardiac echocardiography (ICE), the activation map of this tachycardia showed a centrifugal propagation pattern originating from the high atrial wall adjacent to the EC (Fig. 2). It was located close to the sinus node. However, the P wave morphology was a
little different, because we speculated that the propagation velocity surrounding the earliest site was slow at the tachycardia rate. At that earliest site, a folded structure resembling the crista terminalis was recognized with ICE (Fig. 3 Left). Therefore by increasing the energy up to the maximum output of 35 W it was finally possible to successfully eliminate the tachyarrhythmia.

The mechanism of this tachycardia was speculated to be reentry, and the diagnosis was suggested to be SANRT due to its electrophysiological, anatomical, and pharmacological features. After confirming the function of the sinus node on the other side, the session was completed. In the 12-lead ECG after the RFCA, the polarity of the P wave temporarily changed to that of the left side node. The subsequent follow-up has been uneventful.

**Discussion**

Heterotaxy syndrome is primarily induced by disorders of the left–right axis determination during the early embryonic development [3]. This syndrome is characterized by a lot of CHDs, e.g. single common atrium and/or ventricle, common AV valve, pulmonary atresia or stenosis, double outlet right ventricles, and total anomalous pulmonary venous drainage.

Patients with heterotaxy syndrome are subdivided into right and left isomerism cases. In right isomerism cases, a bilateral sinus node can exist and/or twin AV nodes can exist on the anterior and posterior sides, and they can cause supraventricular tachycardias, e.g. AV reciprocating tachycardia (AVRT) involving twin AV nodes [4]. In left isomerism cases, hypoplasia or aplasia of the sinus node or dysfunction of the AV node can lead to sick sinus syndrome or AV block [5].

In TCPC, the lateral tunnel method or intra-atrial rerouting method causes a pressure overload on part of the atrium and leaves a suture in the atrium. This is related to a high arrhythmogenicity in the late phase after the surgery. On the other hand, a TCPC using an EC has a lower incidence of tachyarrhythmias because of the avoidance of the atrial suture lines and maintaining a common atrium with a low-pressure environment, and moreover it provides a reduction in the risk of sinus node dysfunction [6-8]. However, some intrinsic arrhythmogenic conditions, such as heterotaxy syndrome, are associated with a high incidence of arrhythmias [2].

RFCA after a TCPC is usually difficult because of the complexity of the anatomical features after the surgery [3]. In this case, we approached the atrium from the EC with the needle puncture method carefully while using ICE.

This tachycardia’s features were as follows. At first, it was sensitive to ATP and verapamil. In the 12-lead ECG obtained before the RFCA, two types of P wave polarities were recognized, and thus, different atrial rhythms or twin sinus nodes appeared to be present. A His bundle ECG was recorded at two separate sites, and thus, the existence of twin AV nodes was recognized. In the EPS, no VA conduction was observed, and the tachycardia was repetitively induced with single atrial extrastimuli. There was an inverse correlation between the coupling interval of the atrial extra stimulation and the return CL from the stimulation to the tachycardia. Moreover, a rapid intravenous injection of 10 mg of ATP prolonged the atrial CL of the tachycardia resulting in a change in the subsequent ventricular CL and then terminated the tachycardia. An activation map using a 3D mapping system revealed a centrifugal propagation pattern from the high atrial wall adjacent to the EC.

From the above, the tachycardia was determined to be an ATP sensitive atrial tachycardia (AT). Moreover, it was not an AVRT involving the twin AV nodes, which has a relatively high incidence in patients with heterotaxy syndrome [9]. The P wave morphology of the tachycardia exhibited a low voltage with a wide width, however the P wave polarity of the AT was similar to that of the sinus rhythm. The AT was repetitively induced with single atrial extrastimuli, and was sensitive to ATP. Moreover, by using ICE, a folded structure resembling the crista terminalis was recognized at the site of the earliest activation. Therefore we finally needed a high output energy to eliminate the tachyarrhythmia. However the target was close to the sinus node. Therefore we increased the energy after terminating the tachycardia while observing the reaction of sinus rhythm. After the RFCA, we created a sinus rhythm map with a 3D mapping image (Fig. 3 Right), and the mapping of the shifted rhythm showed that the foci moved to the fold of the right pulmonary vein in the opposite atrium, which
embryologically was suggested to be the crista terminalis with a right isomerism. The mechanism of this tachycardia was suggested to be an SANRT due to its electrophysiological, anatomical, and pharmacological features [10].

Conflict of interest

None declared.

Acknowledgments

We thank Mr. John Martin for the linguistic assistance in the preparation of this manuscript.

References


