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Case Report

Successful radiofrequency catheter ablation of life-threatening atrial tachycardia in an infant with asplenia syndrome

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ABSTRACT

A 1-year-old infant with asplenia syndrome and congenital heart disease consisting of common atrium, common inlet left ventricle with a common atrio-ventricular (AV) valve, pulmonary atresia, and total anomalous pulmonary venous connection was admitted to our hospital for radiofrequency catheter ablation (RFCA) of supraventricular tachycardia (SVT) before total cavo-pulmonary connection. After antiarrhythmic medications were discontinued for RFCA, she suffered from SVT that resulted in the rapid deterioration of hemodynamic status. Antiarrhythmic medications and cardioversion were not effective in terminating SVT. The baseline electrocardiogram confirmed the existence of twin AV nodes; however, this SVT was revealed to be focal atrial tachycardia (AT) with enhanced automaticity. The origin of AT was not related to surgical scar. Emergent RFCA for AT was successful in our case of asplenia syndrome. AT is a life-threatening complication in a single ventricle and delayed treatment can be fatal. It is important to perform RFCA promptly when drug treatment is not effective. We suggest that the AV node is not always the target site for ablation in patients with asplenia syndrome and twin AV nodes.

<**Learning objective:** In the case of supraventricular tachycardia with asplenia and twin atrio-ventricular (AV) nodes, atrial tachycardia (AT) as well as AV reentrant tachycardia could occur. AT is a life-threatening complication in infants with single ventricle. If drug therapy is not effective, emergent catheter ablation should be performed. AV node is not always the target site for ablation in patients with asplenia and twin AV nodes.>

Introduction

Supraventricular tachycardia (SVT) is a life-threatening and common complication in single ventricular morphology with heterotaxy syndrome. Abnormal cardiac conduction systems, including twin atrio-ventricular (AV) nodes, have been described in asplenia syndrome or right isomerism (RI) [1,2]. In asplenia syndrome, AV reentrant tachycardia (AVRT) using twin AV nodes has been well described; however, atrial tachycardia (AT) rarely occurs. There have been few reports describing radiofrequency catheter ablation (RFCA) in infants with asplenia syndrome for focal AT with enhanced automaticity, the origin of which is not related to surgical scar. We report a case of successful emergent RFCA for AT in an infant with asplenia syndrome and twin AV nodes before total cavo-pulmonary connection (TCPC).

Case report

A 1-year-old infant was admitted to our hospital for electrophysiologic study and RFCA before TCPC. Her body weight was 7.5 kg. She was prenatally diagnosed with asplenia syndrome including common atrium, common inlet left ventricle, pulmonary atresia, common AV valve (CAVV), and total anomalous pulmonary venous connection (TAPVC). She had a common inlet AV connection with single left ventricle. She was born by a caesarean section at 38 weeks of gestation with a birth weight of 2528 g. Blalock–Taussig shunt operation was performed at the age of 19 days, then bidirectional Glenn operation and TAPVC repair were

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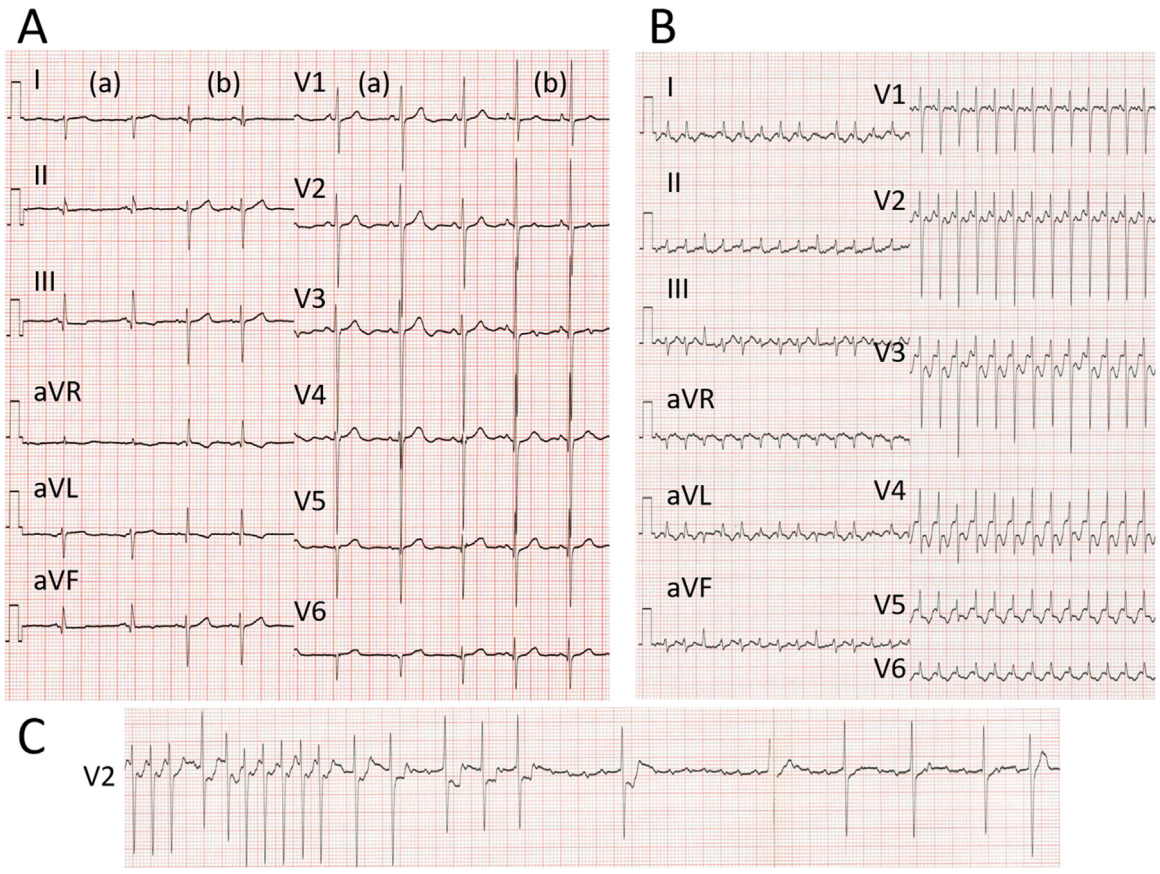

Fig. 1. Twelve-lead surface electrocardiogram (ECG). (A) Baseline ECG showing two types of P waves and different QRS complexes. (a) QRS axis of +135°, P axis of +120°, and V1 with R > S. (b) QRS axis of −80°, P axis of +45°, and V1 with R < S. (B) During a narrow-QRS tachycardia at a rate of 287 bpm a variable QRS axis is noted. (C) After the intravenous infusion of adenosine triphosphate isoelectric baseline with discrete P waves is observed.

performed at the age of 11 months. She was noted to have SVT since the 7th day after birth. It was controlled by the use of flecainide, atenolol, and digoxin.

The baseline electrocardiogram (ECG) exhibited two types of P waves and different QRS complexes (Fig. 1A), suggesting that she had twin sinoatrial nodes and twin AV nodes. All antiarrhythmic medications were discontinued from the day before hospitalization. At night of the day of admission, narrow-QRS tachycardia at a rate of 230–300 bpm was then initiated (Fig. 1B). The tachycardia was the same as the one that occurred when she was 7 days old. An isoelectric baseline with discrete P waves was documented after infusion of adenosine triphosphate (ATP) (0.1 mg/kg) (Fig. 1C), and the tachycardia was demonstrated to be AT on the 12-lead ECG. Cardioversion (1 J/kg) was not effective in terminating this tachycardia. We started infusion of landiolol at 1 μg/kg/min to depress the AV conduction. However, the dose of landiolol was increased to 10 μg/kg/min by 1 μg/kg/min every 15 min, rate control was not achieved. Infusion of nifekalant (0.4 mg/kg/h) was added, nevertheless AT was not terminated. She subsequently fell into shock and lost consciousness. We started mechanical ventilation and infusion of amiodarone at loading dose (5.0 mg/kg), followed by an infusion of 10 mg/kg/day. Drug therapies were not effective and AT was still continuing. Low cardiac output syndrome developed, therefore we decided to perform emergent RFCA.

After discontinuing all antiarrhythmic medication, AT was stopped soon after induction of general anesthesia. Induction of general anesthesia was performed by the intravenous administration of propofol, fentanyl, and muscle relaxants with endotracheal intubation. Sevoflurane was used for the maintenance of general anesthesia. Two 2-Fr octapolar catheters, a 4-Fr quadripolar catheter, and a 7-Fr Navistar quadripolar catheter (Biosense Webster, Diamond Bar, CA, USA) were positioned in the single atrium and single ventricle, in the esophagus with using the CARTO 3 system (Biosense Webster) for 3D mapping. AT was not induced by programmed atrial stimulation. Subsequently, sevoflurane was discontinued and propofol infusion started; however, AT was not induced. Additionally, dobutamine infusion did not induce AT. Finally, premature atrial contractions (PACs) were induced after isoproterenol infusion. The P wave morphology of AT was the same with isoproterenol-induced PACs on the 12-lead ECG. At the earliest activation site of the PAC, a low-amplitude fragmented electrical activity was recorded at the annulus of the CAVV (Fig. 2A, B). Electroanatomical mapping of the single atrium revealed that there was no reentry circuit. Therefore, we diagnosed that the mechanism of AT was focal with enhanced automaticity and the origin of AT was not related to surgical scar. Soon after RF energy application (30 W, 60 °C) at that site for 60 s (Fig. 2C), PACs disappeared immediately (Fig. 2D). After ablation, AT and PACs were not induced by atrial burst pacing, atrial extra stimulation, and isoproterenol infusion. Consequently, we assessed that the ablation was successful. His bundle ECG was not recorded; therefore, we confirmed the earliest sites of ventricular activation by performing atrial pacing. There are two different ventricular activation patterns by pacing at two different atrial sites, indicating the possibility of the existence of twin AV nodes (Fig. 2E, F). However, 1:1 ventriculo-atrial conduction was recorded at ventricular pacing rate of 100–110 ppm, Wenckebach ventriculo-

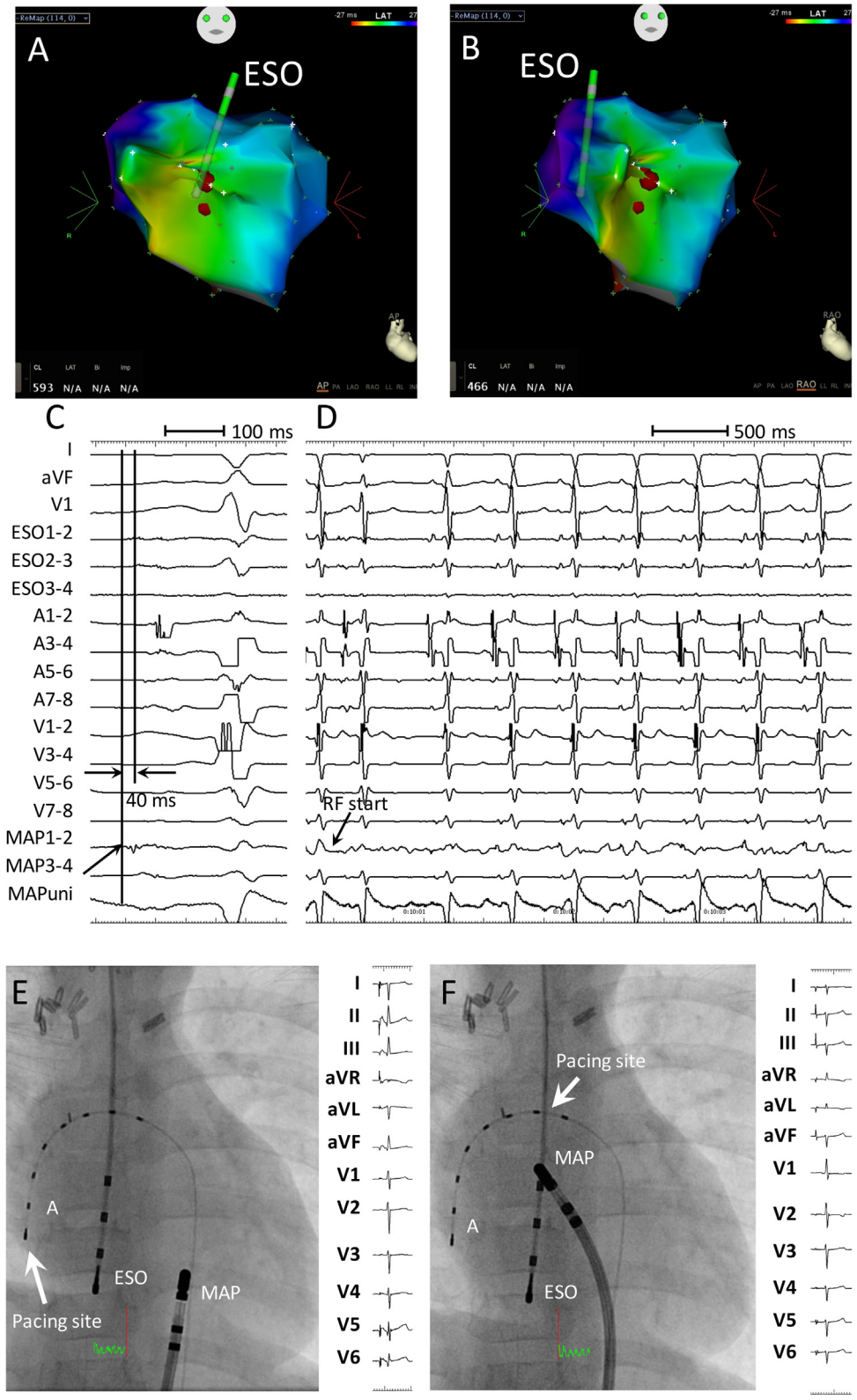

Fig. 2. (A) Electroanatomical mapping of the single atrium anteroposterior view. The red dots show the radiofrequency catheter ablation sites. (B) Right anterior oblique view. (C) Low-amplitude fragmented electrical activity that precedes the surface P wave by 40 ms is noted at the successful ablation site of the premature atrial contractions (PAC). (D) The PAC disappeared after the delivery of radiofrequency energy. (E) Pacing from distal atrial lead showed similar QRS morphology to that in Fig. 2A (a). (F) Pacing from proximal atrium represent a posterior atrioventricular node. The QRS morphology is similar to that in Fig. 2A (b).
ESO, esophagus.

atrial conduction was recorded at ventricular pacing rate of 120 ppm. The retrograde AV conduction property with relatively long refractory period was proved to be related with non-inducibility of AVRT.

We discontinued all antiarrhythmic drugs and AT had not recurred for 3 months. However, the recurrence of AT occurred 4 months after ablation. Recurrent AT was treated and controlled by flecainide. As follow up, we planned to perform 2nd RFCA session for recurrent AT after discontinuing flecainide. However, AT and PACs were not induced by programmed atrial stimulation, atrial extra stimulation, and isoproterenol infusion. Therefore, RFCA was not performed and we continued flecainide. The clinical course was uneventful without recurrence of AT for 3 years.

Discussion

Asplenia syndrome is associated with the conduction system characterized by twin AV nodes [1,2]. Wu et al. reported that twin AV nodes were found in 64% of patients [3]. SVT, which is defined by a narrow QRS tachycardia originating at or above the AV node including AVRT, AV nodal reentrant tachycardia or AT, is reported to be the most common arrhythmia in patients with asplenia syndrome. The occurrence of SVT is reported in 25–26% of the patients with asplenia syndrome [3,4]. SVT is most likely caused by the reentrant mechanism such as twin AV nodes may possibly constitute a reentry circuit and cause AVRT [4,5]. However, AT in asplenia syndrome is rare and little has been reported. Cheung et al. reported that in 85 patients with RI, only 1 patient (1%) had AT and that patient died suddenly 3 days after discontinuing amiodarone on his own [6]. The report from Miyazaki et al. showed that the incidence of SVT after extracardiac Fontan procedure was 27% in patients with RI, and furthermore, 50% of SVTs were not reentrant SVTs involving AT and unknown etiology with ATP insensitivity [7]. In postoperative patients with asplenia syndrome, macro-reentrant AT is known to be common; however, focal AT with enhanced automaticity is extremely uncommon. There have been scarce reports describing RFCA for focal AT with enhanced automaticity in asplenia syndrome, which is not related to surgical scar.

Taking our experience of RFCA for AT into account, not only reentrant SVT including AVRT but also AT should be taken into consideration in asplenia syndrome and twin AV nodes. Automaticity focal AT in children generally appears as an incessant arrhythmia and is often resistant to medical therapy. AT easily causes hemodynamic deterioration in single ventricle patients. If antiarrhythmic drug therapy is ineffective, RFCA is an important therapeutic option. Besides, the catheter access to the atrium and ventricle may become restricted after TCPC, therefore, RFCA should be scheduled before TCPC without hesitation. In the case of RFCA for asplenia syndrome and twin AV nodes, AV node is not always the target site for ablation.

Conflict of interest

The authors declare that there is no conflict of interest.

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