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CASE REPORT

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Ostial atresia of the coronary sinus in patients with supraventricular arrhythmias

Ken-ichi Hiroshima MD | Masahiko Goya MD | Michio Nagashima MD | Masato Fukunaga MD | Masatsugu Ohe MD | Yoshimori An MD | Yu Makihara MD | Kentaro Hayashi MD | Kenji Ando MD

Department of Cardiology, Kokura Memorial Hospital, Kitakyushu, Japan

Correspondence
Masahiko Goya, Heart Rhythm Center, Tokyo Medical and Dental University, Yushima 1-5-45, Bunkyo-ku, Tokyo, 113-8519, Japan.
Email: mgoyamd@yahoo.co.jp

Abstract

Of 3577 patients with supraventricular arrhythmias, 3 demonstrated an atresia of the coronary sinus (CS) ostium. Two patients had the accessory pathways. One had atrial fibrillation. No unroofed CS or apparent persistent left superior vena cava was observed. Venous drainage through a small cardiac vein located on the lateral portion of the tricuspid annulus was observed in all patients. Those cases demonstrated that the incidence of ostial atresia of the CS was 0.084%. Accessory pathways were often accompanied by this anomaly. An abnormal venous orifice located on the lateral tricuspid annulus often functioned as the drainage of the CS flow.

KEYWORDS

coronary sinus, occlusion of the coronary sinus ostium, ostial atresia, persistent left superior vena cava, supraventricular tachycardia

1 | INTRODUCTION

Ostial atresia of the coronary sinus (OACS) is a rare cardiac abnormality but is clinically important because an electrode catheter insertion inside the CS is not feasible. Several case reposts have been published; however, still little is known about this anomaly.

Between September 2004 and March 2014, 3577 patients who underwent catheter ablation of supraventricular arrhythmias were retrospectively analyzed. The target arrhythmias were atrioventricular nodal reentrant tachycardia (AVNRT) in 560, atrioventricular reentrant tachycardia (AVRT) in 410, atrial flutter/atrial tachycardia (AFL/AT) in 658, and atrial fibrillation (Afib) in 1949 patients.

2 | CASE REPORTS

2.1 | Case 1

A 53-year-old woman was referred due to paroxysmal supraventricular tachycardia. A 12-lead electrocardiogram (ECG), the physical examination, chest X-ray, and echocardiograms were normal. We punctured the right internal jugular vein and tried to insert a multielectrode catheter into the ostium of the CS. However, the CS could not be cannulated despite prolonged attempts. Left coronary angiography revealed an OACS and that the CS drained into the right atrium (RA) via a small cardiac vein and collateral vein running along the tricuspid annulus along the lateral portion (Figure 1A). A concealed accessory pathway (AP) on the left lateral mitral annulus (Figure 1A, arrow with the oblique line) was successfully ablated.

2.2 | Case 2

A 25-year-old man with weekly palpitation attacks and a structurally normal heart was referred for catheter ablation. The ECG exhibited no delta waves. Repeated attempts to cannulate the CS ostium were unsuccessful. Then coronary angiography (Video S1) demonstrated an OACS and antegrade drainage located at the lateral portion of the tricuspid annulus (Figure 1B). A left lateral concealed AP (Figure 1B, arrow with the oblique line) was successfully ablated.

Dr. Goya moved from Kokura Memorial Hospital to the Heart Rhythm Center, Tokyo Medical and Dental University.

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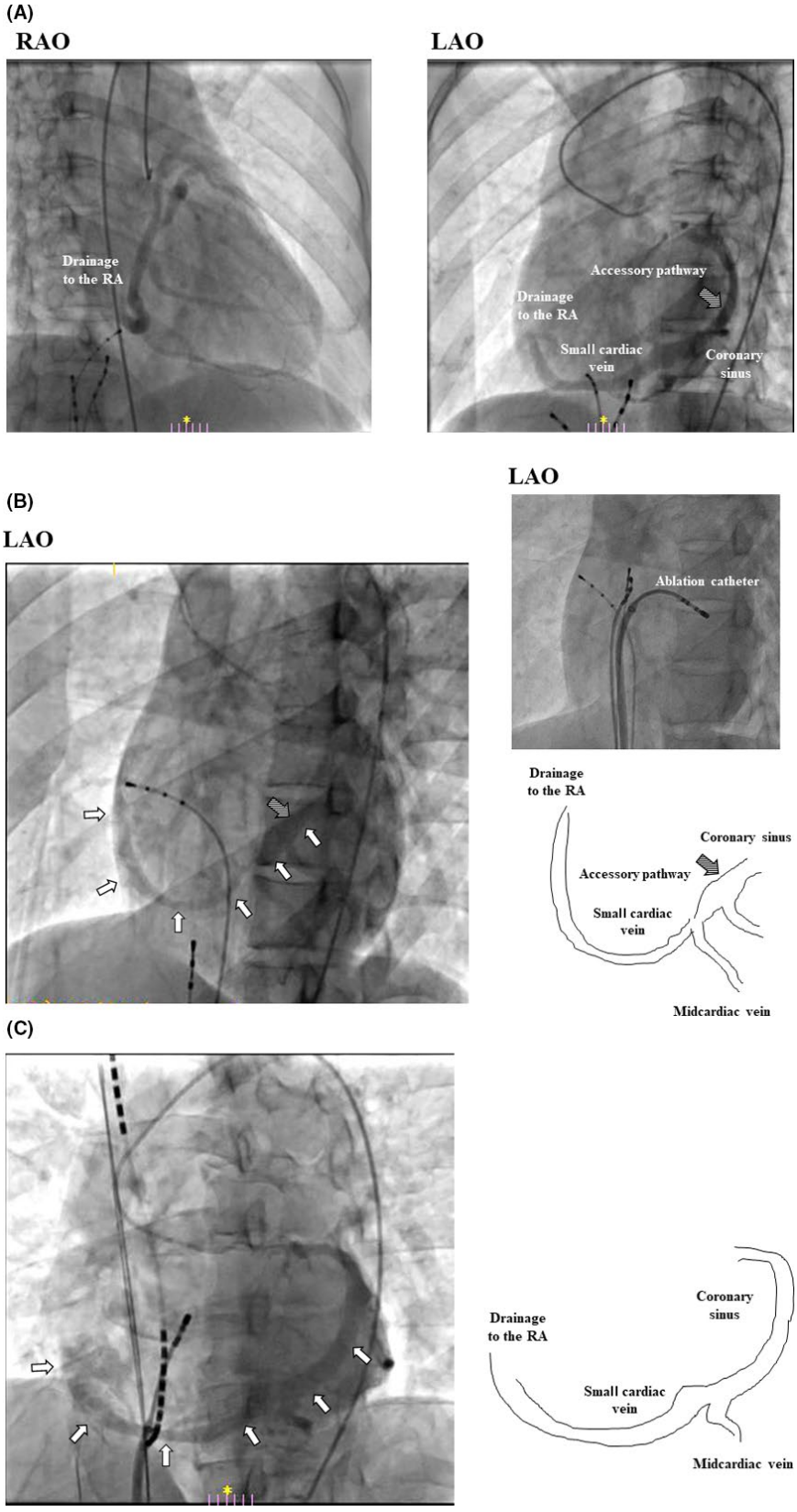

FIGURE 1 A, Left coronary angiography in Case 1—ostial atresia of the coronary sinus (CS) and a dilated small cardiac vein drawing into the right atrium on the lateral tricuspid annulus are visible. The location of the accessory pathway is demonstrated by the arrow with the oblique line. B, Left: A still frame of left coronary angiography and schematic diagram in Case 2—ostial atresia of the CS and a dilated small cardiac vein drawing into the right atrium on the anterolateral tricuspid annulus are visible. The location of the accessory pathway is demonstrated by the arrow with the oblique line. Right upper: Location of ablation catheter during successful energy application. Right lower: Schematic representation of coronary vein system in Case 2. C, Left coronary angiography in Case 3 showed ostial atresia of the CS with a dilated small cardiac vein drawing into the right atrium on the lateral tricuspid annulus. RAO, right anterior oblique; LAO, left anterior oblique; RA, right atrium

2.3 | Case 3

A 49-year-old man was referred due to drug refractory paroxysmal Afib. During the ablation procedure, a 6Fr, 20-electrode catheter was inserted through the right internal jugular vein and was attempted to be cannulated into the CS ostium. However, the CS ostium was atretic and coronary angiography demonstrated the venous drainage was located on the lateral portion of the tricuspid annulus (Figure 1C). After

a transseptal puncture, all four pulmonary veins were successfully isolated. No other tachycardia was inducible during the procedure.

3 | DISCUSSION

The incidence of OACS was 3 out of 3577 patients (0.084%) in our study population. CS atresia was observed in 1 out of 1949 patients with Afib (0.051%), none with AT/AFL or AVNRT, and 2 out of 410 with AVRT (0.488%). There were no statistically significant difference in the prevalence between these three groups, but OACS might be documented more often in patients with AVRT. Shum et al[1] reported OACS detected by multidetector computed tomography (CT) and magnetic resonance imaging. They found 15 cases with ostial CS atresia among 13,842 patients (0.11%). These data suggest that the incidence of OACS is around 0.1%. Shum's study involved various indications for cardiac CT or MRI, but our study population was selected for the evaluation and ablation of supraventricular arrhythmias. From this point of view, our study was new and the consistency is interesting.

As far as we are aware, 10 papers regarding OACS have been published (Table 1). In total, 17 patients with congenital OACS have been reported, including those from our study. Interestingly, 8 of 17 patients had AVRT, while 3 had AVNRT, and only 2 had Afib. Chiang et al[2] reported that major CS abnormalities were found more frequently in patients with APs than in patients with AVNRT. The authors suggested that the major CS abnormalities may be anatomically related to the distribution of the APs, because the APs were located exclusively on the left free wall or in the posteroseptal region in their series. Among 17 patients, 8 had APs. The location of the pathways was the left free wall in five and interatrial septum in three patients. This distribution was similar to that of Chiang's report. Takatsuki et al[3] speculated that during cardiogenesis, the CS develops from the proximal left sinus horn of the sinus venosus at 7–8 weeks of the embryonic age. APs are considered a remnant of an incomplete separation of the atrial and ventricular myocardium by the annulus fibrosus, which develops during the same stage as the CS in the embryo. Therefore, OACS and the APs might be embryologically related to each other, and this might be the reason why AVRT has more OACS.

Santoscoy et al[4] reported that OACS is often associated with other cardiac abnormalities such as a persistent left superior vena cava (PLSVC) or unroofed CS. In our group patients, no patients had an unroofed CS or functional PLSVCs. Because if a functional PLSVC exits, blood returns inside the CS in a retrograde direction, passing upward to the left SVC, and left innominate vein during coronary angiography.

TABLE 1 Clinical studies on ostial atresia of the CS and supraventricular arrhythmias

Author	Year	No of cases	Concomitant cardiac anomaly	Target arrhythmia
Takatsuki et al[3]	2000	1	PLSVC	AP around the CS ostium
Khairy et al.	2005	3 (4)	PLSVC(2), unroofed CS (1)	Left-sided AP, uncommon AVNRT, right mideseptal AP
Okuyama et al.	2005	1	Tiny PLSVC	Common AVNRT
Luik et al.	2006	3	PLSVC(3)	Focal AT originating from the anterior MVA(1), AFL/Afib(1), AFL(1)
Kim et al.	2008	1	None	Left-sided AP
Yoshida et al.	2012	1	None	Left posteroseptal AP
Kawata et al.	2013	1	PLSVC, unroofed CS	Common AVNRT
Hirao et al.	2017	1	PLSVC	AT originating from the CS ostium
Inamura et al.	2017	1	None	AF induced by AESs originating from the CS and VOM
Kataoka et al.	2017	1	PLSVC	Left-sided AP
Hiroshima et al.	Present	3	None	Left-sided AP (2), Afib (1)

Abbreviations: AES, atrial extrasystole; Afib, atrial fibrillation; AFL, atrial flutter; AP, accessory pathway: AT, atrial tachycardia; MVA, mitral valve annulus; AVNRT, atrioventricular nodal reentrant tachycardia; CS, coronary sinus; PLSVC, persistent left superior vena cava; VOM, vein of Marshall.

According to previous reports, only 2 out of 17 patients (11.8%) have demonstrated an unroofed CS while 10 patients (58.8%) had PLSVCs. That data suggest that in patients with this anomaly a PLSVC is often documented, and in contrast, an unroofed CS is less than expected.

In patients with OACS, there must be some extraordinary drainage of the venous flow of the CS. As previously mentioned, 10 patients had a PLSVC and 2 had an unroofed CS. Those two types of abnormal venous return morphologies can function as the drainage. Further, there were no obvious additional anomalies in six patients, including our three patients. Angiography demonstrated antegrade drainage located on the lateral portion of the tricuspid annulus through a dilated small cardiac vein in four patients. This suggested that an abnormal venous orifice located on the lateral tricuspid annulus often functioned as the drainage of the CS flow in this anomaly. We did not attempt it, but it might be technically feasible to cannulate the coronary sinus through a small cardiac vein using a small-sized electrode catheter.

4 | CONCLUSIONS

Our study suggested that the incidence of OACS was around 0.1% in patients with supraventricular arrhythmias. Further, AVRT via an accessory pathways was often accompanied by this anomaly (0.488%) due to embryological reasons than residual supraventricular arrhythmias. An abnormal venous orifice located on the lateral tricuspid annulus often functioned as the drainage of the CS flow in this anomaly.

CONFLICT OF INTEREST

The authors declare no conflict of interests for this article.

ORCID

Masahiko Goya https://orcid.org/0000-0002-7210-0671

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SUPPORTING INFORMATION

Additional supporting information may be found online in the Supporting Information section at the end of the article.

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