Journal of Cardiology Cases 19 (2019) 29–32

Contents lists available at ScienceDirect

Journal of Cardiology Cases

journal homepage: www.elsevier.com/locate/jccase

Case Report

Surgical repair of unroofed coronary sinus with severe mitral regurgitation in an elderly patient

Keiji Yamada (MD)[a], Shinichiro Miyazaki (MD)[a], Mai Oboshi (MD)[a], Chisato Suezawa (MD)[a], Yutaka Sakakibara (MD PhD)[b], Kazunobu Nishimura (MD PhD)[b,*]

[a] Department of Cardiology, Takamatsu Red-Cross Hospital, Kagawa, Japan
[b] Department of Cardiovascular Surgery, Takamatsu Red-Cross Hospital, Kagawa, Japan

ARTICLE INFO

Article history:
Received 23 June 2018
Received in revised form 19 September 2018
Accepted 25 September 2018

Keywords:
Unroofed coronary sinus
Mitral regurgitation
Surgical repair

ABSTRACT

An unroofed coronary sinus (URCS) is a rare anomaly that produces communication between the left atrium (LA) and the coronary sinus (CS), resulting in a left-to-right shunt. Due to the lack of symptoms and particular anatomical characteristics, this disease is difficult to diagnose, and prone to be overlooked. An 85-year-old man was admitted to our hospital because of anorexia and shortness of breath. On physical examination, a systolic murmur was heard at the apex, and pitting edema was present in both legs. Transthoracic echocardiography showed severe regurgitation of the mitral valve and tricuspid valve. Transesophageal echocardiography confirmed a shunt between the LA and the CS. Because of uncontrolled heart failure, we performed surgical repair 50 days after admission. Under cardiopulmonary bypass and heart arrest, the URCS was detected in the LA and directly sutured. Repair of the mitral and tricuspid valves and the Maze procedure were also performed. The patient had a good postoperative course, and has been doing well for 2 years. Transesophageal echocardiography is helpful for diagnosis of URCS. Mitral regurgitation might raise the left atrial pressure and result in increase in shunt flow, causing left and right heart failure in elderly patients.

<**Learning objective:** Diagnosis of an unroofed coronary sinus (URCS) is often overlooked in adult patients because of the lack of symptoms and/or particular anatomical characteristics. We herein describe an octogenarian patient who was diagnosed with URCS in association with severe mitral and tricuspid regurgitation. Transesophageal echocardiography helped to identify the location of the URCS. The cause of the heart failure was mitral regurgitation, which raised the left atrial pressure and increased shunt flow.>

© 2018 Japanese College of Cardiology. Published by Elsevier Ltd. All rights reserved.

Introduction

An unroofed coronary sinus (URCS) is a rare congenital anomaly characterized by a defect between the left atrium (LA) and the coronary sinus (CS), resulting in a left-to-right shunt [1]. Because of lack of symptoms and particular anatomical characteristics of URCS, this disease is prone to be misdiagnosed [2]. Before the era of echocardiography, precise diagnosis of URCS was only possible during surgery [3] or autopsy. Even with routine use of echocardiography, the diagnosis of URCS is still difficult. We herein report an octogenarian patient who developed left and right heart failure and was diagnosed with an URCS in association with severe mitral and tricuspid regurgitation.

Case report

An 85-year-old man was admitted to our hospital because of anorexia and shortness of breath. He was previously suspected to have an atrioventricular septal defect (AVSD) at another hospital, and was recommended to undergo an operation at that time, but he refused because of minimal symptoms. Physical examination revealed a grade ll/Vl systolic murmur at apex, and edema in both hips and legs. The heart was markedly enlarged with a cardiothoracic ratio of 72% on chest radiograph. Electrocardiogram revealed an incomplete right bundle block with sinus tachycardia and occasional atrial flutter (Fig. 1). Blood test showed a high bilirubin level at 3.0 mg/dl, and a high N-terminal pro–B-type

* Corresponding author at: Department of Cardiovascular Surgery, Takamatsu Red-Cross Hospital, 4-1-3, Ban-cho, Takamatsu-shi, Kagawa 760-0017, Japan.
E-mail address: nishimura-kazunobu@takamatsu.jrc.or.jp (K. Nishimura).

https://doi.org/10.1016/j.jccase.2018.09.002
1878-5409/© 2018 Japanese College of Cardiology. Published by Elsevier Ltd. All rights reserved.

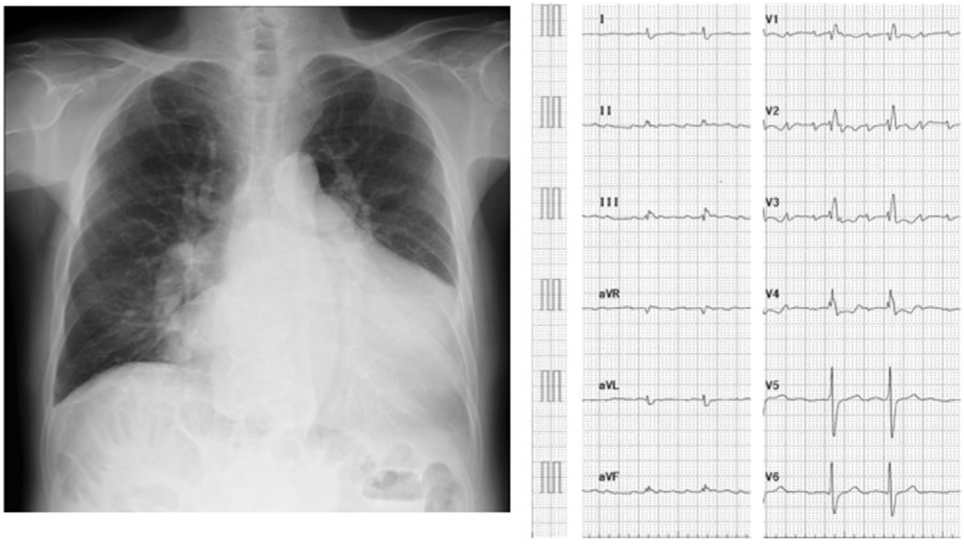

Fig. 1. The patient's chest radiograph (left) and electrocardiogram (right) on admission. The chest radiograph showed marked cardiomegaly with a cardiothoracic ratio of 72%. Electrocardiogram revealed an incomplete right bundle branch block with sinus tachycardia.

natriuretic polypeptide level at 1750 pg/ml. Transthoracic echocardiography (TTE) showed dilatation of the LA, the right atrium, and the right ventricle in association with severe regurgitation of the mitral valve and tricuspid valve. The size of the left ventricle and the ejection fraction were within the normal ranges. The CS was enlarged, the cause of which was unclear. Because the level attached to the septum of the mitral valve was the same as that of the tricuspid valve, the patient was diagnosed with an incomplete type AVSD. A septum primum was present, but no defect of the atrial or ventricle level was observed (Fig. 2). No shunt flow was detected by TTE. Transesophageal echocardiography (TEE) confirmed a shunt between the LA and the CS with a definitive diagnosis of URCS (Fig. 2). Cardiac catheterization showed elevation of the pulmonary artery pressure (64/21, mean: 30 mmHg), and the pulmonary capillary wedge pressure (mean: 19 mmHg). The left-to right shunt was measured with Qp/Qs of 2.36. The calculated pulmonary arterial resistance was 1.81 WU.

Because the patient was in poor condition due to the uncontrolled left and right heart failure, he decided to undergo surgical repair 50 days after admission. On cardiopulmonary bypass (CPB) under heart arrest, a left atrial incision was made by a right-sided approach. The URCS was detected just to the right (medial) side of the medial commissure of the mitral valve. The size of URCS was approximately 5 mm with outer wall resembling eaves of 15 × 20 mm (Fig. 3). We directly sutured the URCS defect. Examination of the mitral valve revealed prolapse of medial side of the anterior leaflet with chordal rupture, while a cleft was not observed. The mitral and tricuspid valves were properly repaired with artificial chordae and rings. The Maze procedure was also conducted to prevent paroxysmal atrial flutter. The aortic cross-clamp time and CPB time were 174 and 231 mins, respectively. The patient had a good postoperative course with a decrease in his cardiothoracic ratio to 50%, and he was discharged on day 32 postoperatively. He has been doing well for more than 2 years.

Discussion

A URCS is a spectrum of cardiac anomalies in which part or all of the common wall between the CS and the LA is absent. Most affected patients are diagnosed at a younger age, with a persistent left superior vena cava (PLSVC), and other type of congenital cardiac anomalies including a partial or complete AVSD [4]. Attenhofer Jost et al. reported that only 60% of patients were diagnosed with URCS before surgery [2]. In adults, this disease is prone to be misdiagnosed due to the lack of symptoms and particular anatomical characteristics [3]. Although limited agreement has been reached regarding the categorization of URCS, some reports have classified it by the location of the aperture and presence or absence of PLSVC [4]. The patient described herein was categorized as having an unroofed midportion of the coronary sinus according to Kirklin and Barratt-Boyces [1], and type ll URCS according to Xie et al. [4].

Diagnosis of URCS by TTE was difficult, particularly type ll URCS because of its anatomical location. Contrast echocardiography is an option for accurate diagnosis of this type of defect [4]. Computed tomography (CT) with contrast enhancement has recently become available for diagnosis of URCS [5]. However, the loss of information regarding shunt flow is a drawback of CT. TEE is also effective in adult patients [6], because the transducer is positioned just behind the LA posterior wall. Accurate diagnosis is essential to obtain good surgical results and to avoid missing abnormalities. In a previous report, a URCS was incidentally recognized during the cardiac operation [3]. TEE can show cardiac structures more precisely, and should be increasingly and aggressively used in future.

In combination with the URCS, the patient had severe mitral regurgitation (MR) with chordal rupture. In a previous report by Sun et al., MR was observed in 2 of 25 adult patients with URCS [6]. Esmaeilzadeh et al. also reported a case of partial URCS with MR in a 74-year-old woman [7]. Whether MR was a coincidence or influenced by the URCS was unclear. We recognized that the flow of the left-to right shunt was not a little in proportion to the size of the defect (5 mm), which we confirmed intraoperatively. We speculated that as grade of MR increased, the LA pressure rose, which resulted in an increase in shunt flow, thus, the patient developed both left and right heart failure.

In conclusion, TEE was extremely helpful to identify the location of the URCS. The cause of the patient's heart failure lately manifested was MR, which resulted in both increased LA pressure and a left-right shunt. Even if the defect of URCS is small, the left-to-right shunt would increase after developing MR, and accurate diagnosis is important before surgery.

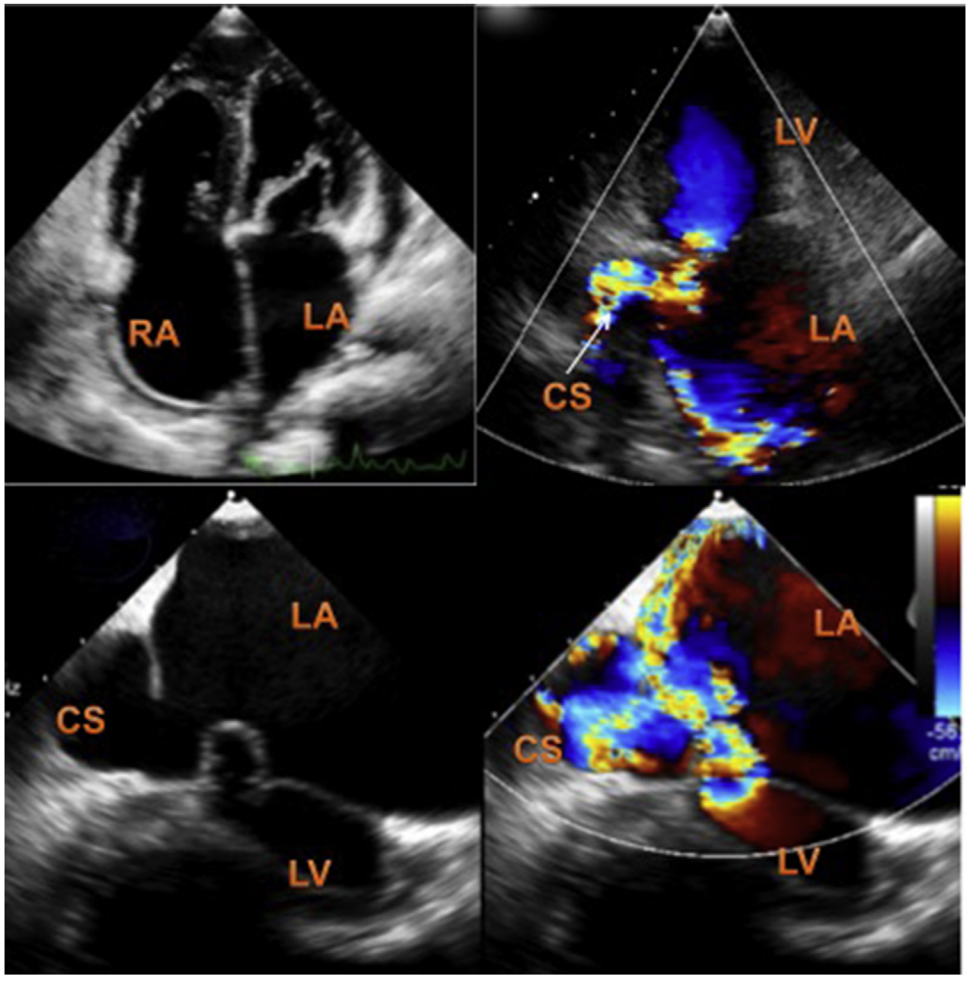

Fig. 2. Transthoracic echography (upper two pictures) and transesophageal echocardiography (lower two pictures). The upper left picture is the four-chamber view, which demonstrates that the level attached to the septum of the mitral valve was the same as that of the tricuspid valve. The upper right picture shows severe regurgitation of the mitral valve and jet flow into the enlarged CS; no shunt flow was detected. The lower left picture shows enlarged CS, wall of which was partially absent. The lower right picture shows the same phase image as the left with color doppler, suggesting the left-to right shunt.
CS, coronary sinus; LA, left atrium; LV, left ventricle; RA, right atrium.

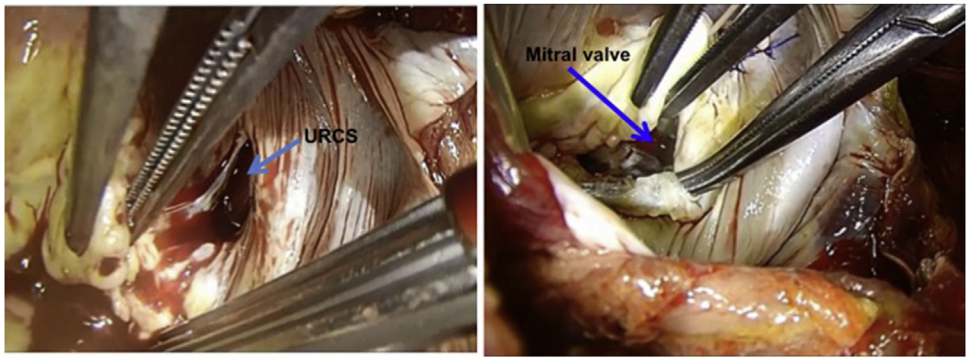

Fig. 3. Intraoperative photographs. (Left) A 5-mm defect of the unroofed coronary sinus is visible at the upper-right side of the medial mitral commissure. (Right) After direct suturing of the defect, the mitral valve with chordal rupture was inspected.
URCS, unroofed coronary sinus.

Acknowledgment

We thank Angela Morben, DVM, ELS, from Edanz Group (www.edanzediting.com/ac), for editing a draft of this manuscript.

Conflict of interest

All authors declare that they have no conflicts of interest.

References

[1] Kirklin JW, Barratt-Boyces BG. Cardiac surgery. 2nd ed. New York: Churchill Livingstone, Inc.; 1993
. p. 683–4.

[2] Attenhofer Jost CH, Connolly HM, Danielson GK, Dearani JA, Warnes CA, Tajik AJ. Clinical features and surgical outcome in 25 patients with fenestration of the coronary sinus. Cardiol Young 2007;17:592–600.

[3] Payne DM, Hamilton A, Milne D, Tanzora R, O'Reilly M. Intraoperative diagnosis of unroofed coronary sinus after aortic valve replacement. Ann Thorac Surg 2012;93:296–7.

[4] Xie MX, Yang YL, Cheng TO, Wang XF, Li K, Ren PP, et al. Coronary sinus septal defect (unroofed coronary sinus): echocardiographic diagnosis and surgical treatment. Int J Cardiol 2013;168:1258–63.

[5] Ouchi K, Sakuma T, Kawai M, Fukuda K. Incidence and appearance of coronary sinus anomalies in adults on cardiac CT. Jpn J Radiol 2016;34:684–90.

[6] Sun T, Fei HW, Huang HL, Chen OD, Zheng ZC, Zhang CJ, et al. Transesophageal echocardiography for coronary sinus imaging in partially unroofed coronary sinus. Echocardiography 2014;31:74–82.

[7] Esmaeilzadeh M, Salehi-Omran M, Hosseini S, Sadr-Ameli M. Partially unroofed coronary sinus with intact atrial septum in an elderly patient. Res Cardiovasc Med 2012;1:33–6.