Single Coronary Ostium with Obstructive Hypertrophic Cardiomyopathy-Case Report

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Case Report

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Abstract

Background

Hypertrophic cardiomyopathy (HCM) is the monogenic inherited cardiovascular disorder. In addition, single coronary artery (SCA) is a rare congenital anomaly of the coronary arteries. However, single coronary artery concomitant with severe obstructive hypertrophic cardiomyopathy has never been reported in the literature.

Case presentation

Herein, we present a 64-year-old female diagnosed as a single left coronary artery with severe obstructive hypertrophic cardiomyopathy. Obstructive hypertrophic cardiomyopathy was treated with "MORROW procedure". The patient was discharged on the seventh postoperative day and has been asymptomatic during the follow-up.

Conclusion

To our knowledge, single left coronary artery with severe obstructive hypertrophic cardiomyopathy is the first to be reported in the literature. In addition, myocarial pretection by cardioplegia antegrade perfusion in the single coronary artery was safe in the patient with single coronary artery and obstructive hypertrophic cardiomyopathy.

Introduction

Hypertrophic cardiomyopathy (HCM) is a monogenic inherited cardiovascular disease affecting 1 in 500 people (0.2%)[1]. Furthermore, single coronary artery (SCA) is a rare congenital coronary anomaly. According to the literature reports, the incidence of a single coronary artery is 0.024–0.066%[2]. In 1979, Lipton et al. proposed a very useful method for classifying single coronary anomalies, which was further improved by Yamanaka and Hobbs in 1990[3]. SCA is more frequent when it coexists with other congenital anomalies while it could also be an isolated congenital heart disease sometimes[3]. In addition, several cases of HCM coexisting with abnormal coronary origin have been reported in the published literature[4–6]. However, severe obstructive hypertrophic cardiomyopathy with SCA was never reported in the literature. Our current case report would provide the treatment of the patient with coexistence of HCM and SCA.

Case Presentation

A 64-year-old woman was admitted to hospital because of chest tightness and shortness of breath. Physical examination revealed a grade III/VI systolic murmur with tremor in the third intercostal space of the left sternal border. The patient had no other significant clinical manifestations during hospitalization. Hypertension family history was found through inquiry (both her father and two older sisters had

hypertension) and this patient suffered from grade II hypertension (very high risk). The patient underwent left upper lobectomy for lung cancer 5 years ago, and the postoperative recovery was good. Holter electrocardiogram (ECG) showed occasional atrial premature beats (APB), frequent multi-source premature ventricular contractions (VPC), and ST-T changes (Fig. 1). Echocardiography showed left ventricular ejection fraction (LVEF) was 63%, and the interventricular septum (IVS) was locally thickened (diastole, 20 mm). The hypertrophic myocardium clearly protrudes into the left ventricular outflow tract (LVOT) (diastole, Fig. 2A), which caused severe obstruction of LVOT. The mitral valve regurgitation (severe) and SAM sign were observed in the systolic stage (Fig. 2B). In addition, the diastolic function of the left ventricle was reduced (Additional file 1: Movie 1). The echocardiography also displayed systolic blood flow in a specific direction from the left ventricle to the aorta, with a peak velocity of 6.08 m/s, a maximum transvalvular pressure of 147.73 mmHg and a mean pressure of 63.37 mmHg (systole, Fig. 2C, Additional file 2: Movie 2). Coronary artery CTA showed significant stenosis of the LVOT (Fig. 2D), and the right coronary artery (RCA) was found to be undeveloped. Three-dimensional coronary artery CTA imaging revealed the abnormal origin and course of the coronary artery (Fig. 3A/B). The subsequent coronary angiography (DSA) examinations further confirmed the diagnosis of a single coronary artery (SCA). The left coronary artery (LCA) originated from the left sinus of the aortic sinus and spread throughout the entire heart. The great circumflex (CX) branch of the LCA supplied the area that right coronary artery should supply in the normal condition (Fig. 4). After the comprehensive evaluation of the preoperative condition and exclusion of the surgical contraindications, the "MORROW" procedure was performed under cardiopulmonary bypass. We adopted anterograde perfusion of the aortic root during the operation, and then cardiac arrest was successful. The intraoperative findings were consistent with the preoperative data: the myocardium with obvious hypertrophy of the ventricular septum was seen protruding into the LVOT (Fig. 5A), and RCA was undetected as well (Fig. 5B). During the operation, hypertrophic myocardium was removed carefully and appropriately, which unclogged the LVOT. During the operation, about 3×2×0.8cm hypertrophic myocardium was cut down longitudinally from 8-10mm below the midpoint of the inferior margin of the right coronary valve to the junction of the left and right coronary valves. Further exploration showed that the mitral valve and subvalvular apparatus was normal. The patient was discharged on the seventh postoperative day. One day before discharge, the Echocardiography showed that the maximum transvalvular pressure between the LVOT and the aorta artery was 15 mmHg, the mitral valve regurgitation was mild (Fig. 6A), the LVOT had been dredged (Fig. 6B) and the SAM sign was disappeared (movie 3). After more than 3 months of follow-up,the patient has been asymptomatic.

Discussion And Conclusions

Hypertrophic cardiomyopathy (HCM) developed as a monogenic inherited cardiac muscular disease with an incidence of about 0.2%[1]. The clinical presentation of HCM is diverse and the pathophysiology is complex, yet therapeutic strategies are effective and available. It has transitioned from a rare and nearly uncurable disease to a common genetic disorder up to now[7]. It is considered as an important cause of arrhythmic cardiac arrest, heart failure, and atrial fibrillation (with embolic stroke)[8]. Clinical diagnosis is

primarily based on unexplained left ventricular hypertrophy as determined by echocardiography or cardiovascular magnetic resonance imaging (MRI)[9].

Single coronary artery (SCA) is a rare congenital coronary artery developing abnormality with an incidence of 0.024–0.066% that only one coronary artery emerges from a coronary ostium, and supplies the entire heart[2]. In 1979, Lipton et al. proposed a very useful classification of single coronary artery malformations based on a large number of clinical cases. The classification was further improved by Yamanaka and Hobbs in 1990, it was divided into different subtypes according to the origin and course of coronary artery[3]. The first level of classification is divided into right type (R) and left type (L) according to the origin of the single coronary artery; the second level of classification is divided into type I, type II and type III according to the course of coronary branches. Type I: The artery runs along the anatomical route of the left coronary artery (LCA) or the right coronary artery (RCA). The CX provides the posterior descending artery, runs in the posterior atrioventricular groove, and also CX branches supply the right atrium and right ventricle; in the absence of LCA, a very large right coronary artery (RCA) is located in the posterior atrioventricular groove, and extends to the anterior base of the heart, where it forms an anterior descending artery (LAD). Type II: The other coronary artery originates proximal to the normally located one, passing through the base of the heart before reaching the normal part of the native coronary artery. Type III: single coronary artery originates from the right coronary sinus, while the left anterior descending (LAD) and left circumflex (LCX) branches originate from the common trunk, respectively. According to this classification, this patient belongs to the L-I type originating from the left coronary sinus.

In fact, HCM and coronary artery abnormalities are considered to be the leading causes of exercise-related cardiac arrest, especially in young adults[10]. SCA can be an isolated congenital heart disease. However, when it coexists with certain other congenital anomalies, it is found much more frequently. In the published literature, congenital anomalies coexisting with SCA include coronary aneurysm [11], coronary arteriovenous fistula [12], ventricular septal defect[13], transposition of great vessels[14], patent foramen ovale [15], tetralogy of Fallot [16], trunk arteriosus [17], patent ductus arteriosus[18], and bicuspid aortic valve [19]. In addition, several cases of HCM coexisting with abnormal coronary origin but not SCA have been reported in the literature[4–6]. Therefore, the current case was first reported in the literature which provides evidence for the coexistence of HCM and single-vessel coronary anomalies.

Various rare courses of SCA can lead to accidental ligation or injury of important main vessels during cardiac surgery. Therefore, every cardiac surgeon and coronary angiographer should be familiar with the presence and anatomy of this congenital anomaly. Because of the rarity of this condition and the complications during surgery, it is recommended that coronary angiography should be performed routinely before cardiac surgery. In addition, SCA should also be paid attention to during cardiopulmonary bypass surgery because the myocardial protection was related to the strategy of cardioplegia perfusion. We discussed the coronary angiography and echocardiographic imaging data of the patient in detail before operation, and considered that there was no obvious abnormality of the aortic valve. Finally, we agreed that anterograde perfusion in the aortic root could be safe and effective, and direct perfusion of a

single coronary artery would be performed when necessary. This article is only for individual cases, patients with the same disease need individual solutions. Our current case report provides the evidence for the coexistence of HCM and SCA, and looks forward to providing corresponding support for the recurrence of the same case in the future.

List Of Abbreviations

Cardiopulmonary bypass CPB

Single coronary artery SCA

Hypertrophic cardiomyopathy HCM

Electrocardiogram ECG

Left ventricular ejection fraction LVEF

Left ventricular outflow tract LVOT

Circumflex CX

Left coronary artery LCA

Right coronary artery RCA

Anterior descending artery LAD

The interventricular septum IVS

Declarations

Ethics approval and consent to participate:

This study was approved by the Ethics Committee of Qilu Hospital of Shandong University, Jinan, China. Written informed consent was obtained from the patient reported in this study. All methods were performed in accordance with the Declaration of Helsinki.

Availability of data and materials:

As this paper is a case report, all data generated or analysed are included in this article.

Consent for publication:

The authors confirm that written consent for submission and publication of this case report, including the images and the associated movie, has been obtained from the patient. A copy of the written consent is

available for review by the Editor-in-Chief of this journal.

Competing interests:

Authors have no competing interests associated with this study.

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Authors' contributions:

LX and LJH collected the data. LX and LCZ wrote the manuscript. WRY, MLW, KXJ, and WKM analysed the patient data. CGQ was Chief surgeon. LJH and LX was assistant. LK was perfusionist. The authors read and approved the final manuscript.

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Conflicts of Interest:

No Disclosure.

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Figures

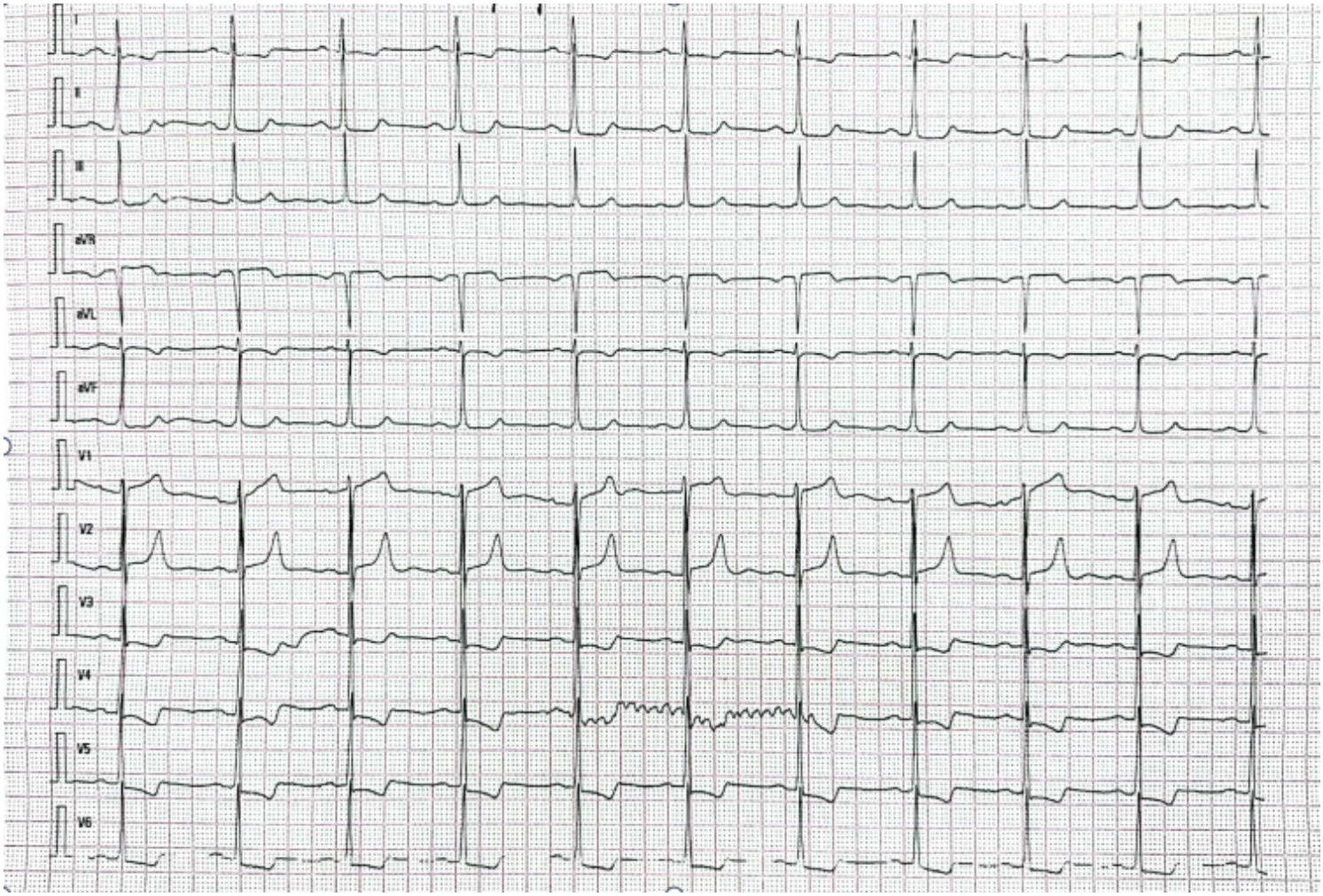

Figure 1

The electrocardiogram (ECG) preoperatively

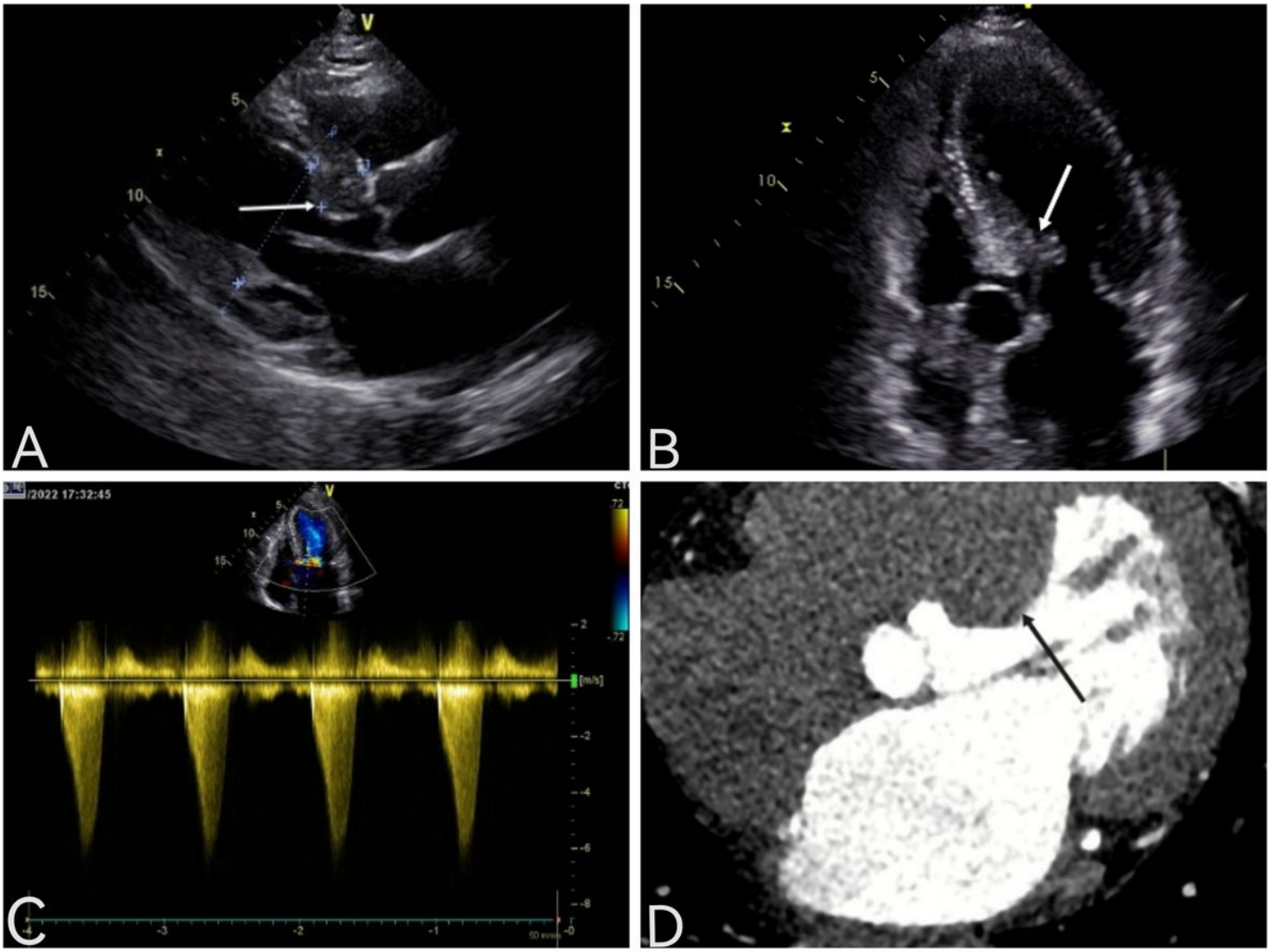

Figure 2

Echocardiography preoperatively

The interventricular septum (IVS) was locally thickened (A, arrow).

SAM sign was obvious (B, arrow).

Systolic blood flow image in a specific direction from the left ventricle to the aorta (C, arrow).

Coronary artery CTA showing the hypertrophic myocardium clearly protrudes into the left ventricular outflow tract (LVOT) (D, arrow).

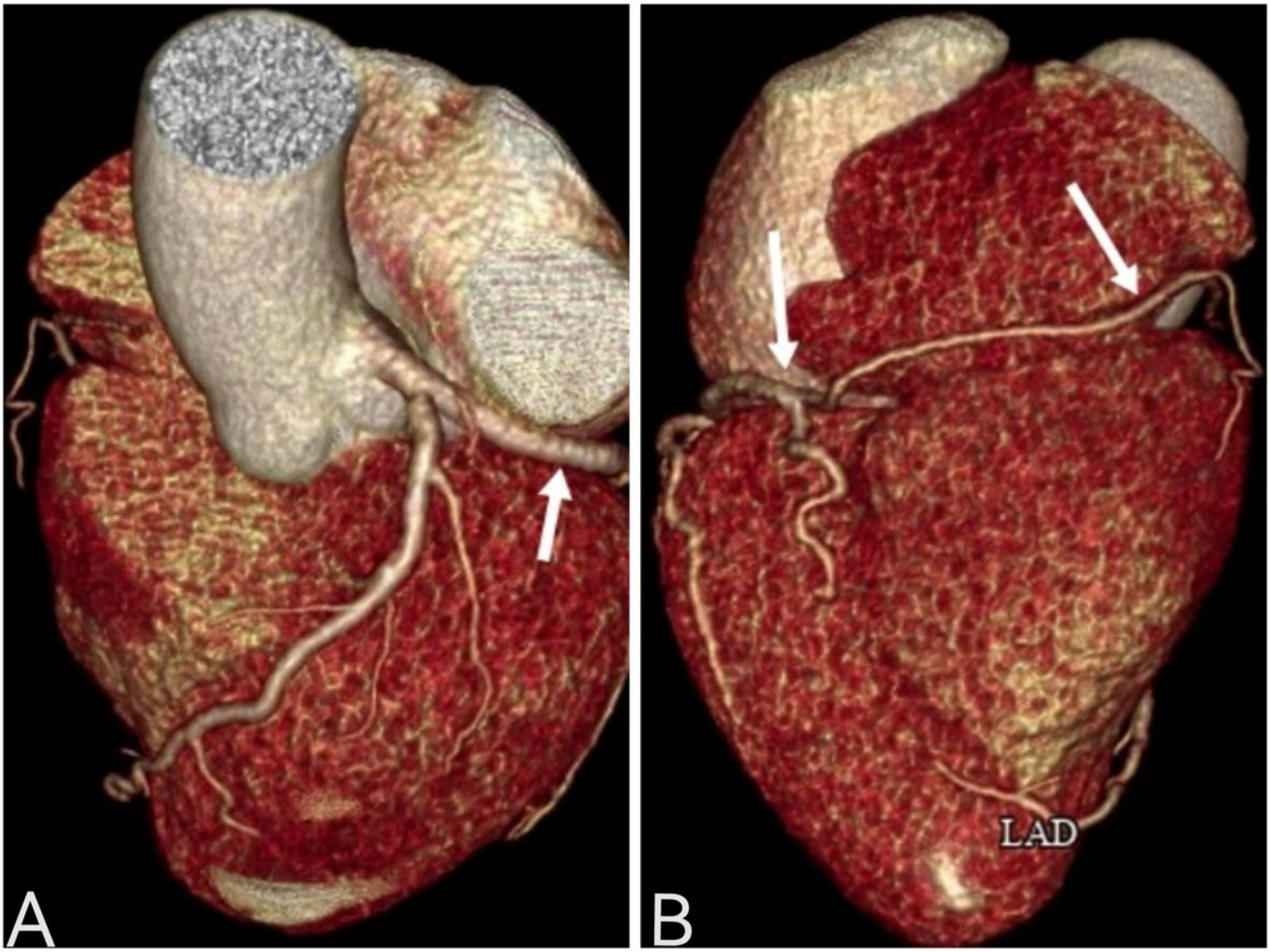

Figure 3

Three-dimensional coronary artery CTA imaging showing the abnormal origin and course of the coronary artery

The giant circumflex (CX) branch of the LCA supplied the right coronary artery blood supply area (A and B, arrow).

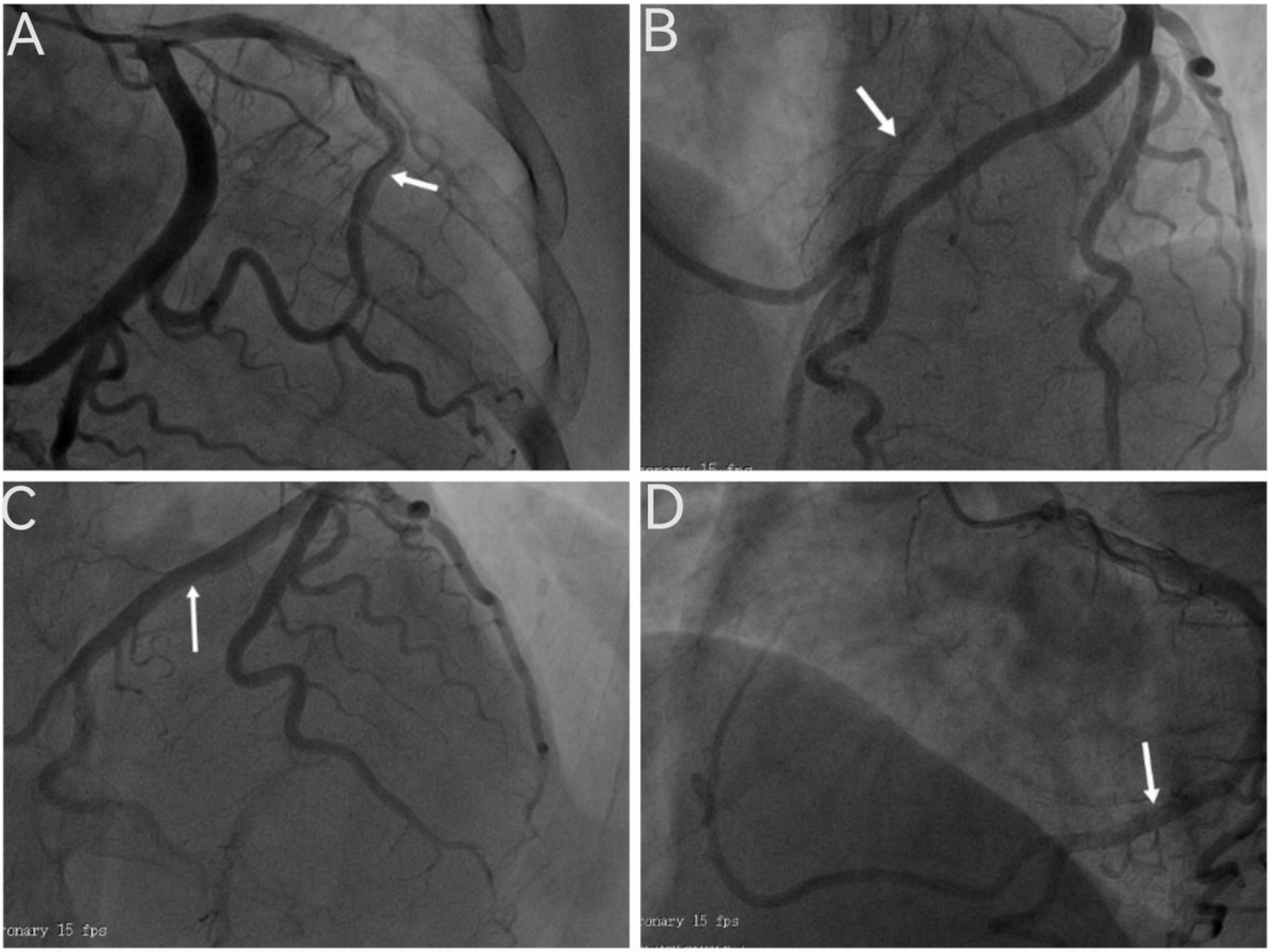

Figure 4

Coronary angiography (DSA) showing a single coronary artery SCA

Anterior descending artery (LAD) (A and B, arrow).

The giant circumflex (CX) branch of the LCA (C and D, arrow).

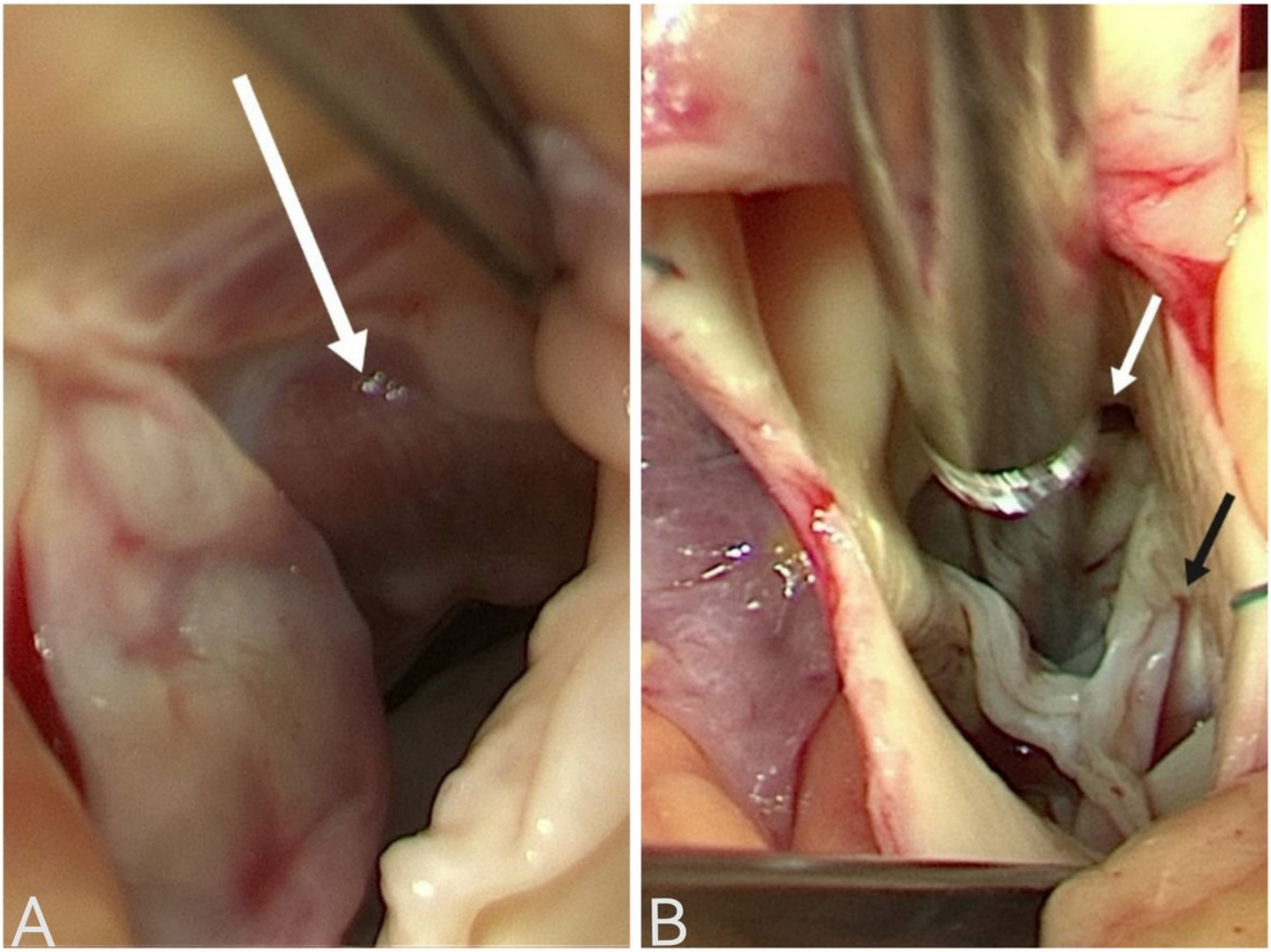

Figure 5

Imaging of the operation

The hypertrophic myocardium of interventricular septum (IVS) (A, arrow).

A single left coronary ostium (B, white arrow).

There is no right coronary artery opening in the right sinus of aortic sinus (B, black arrow).

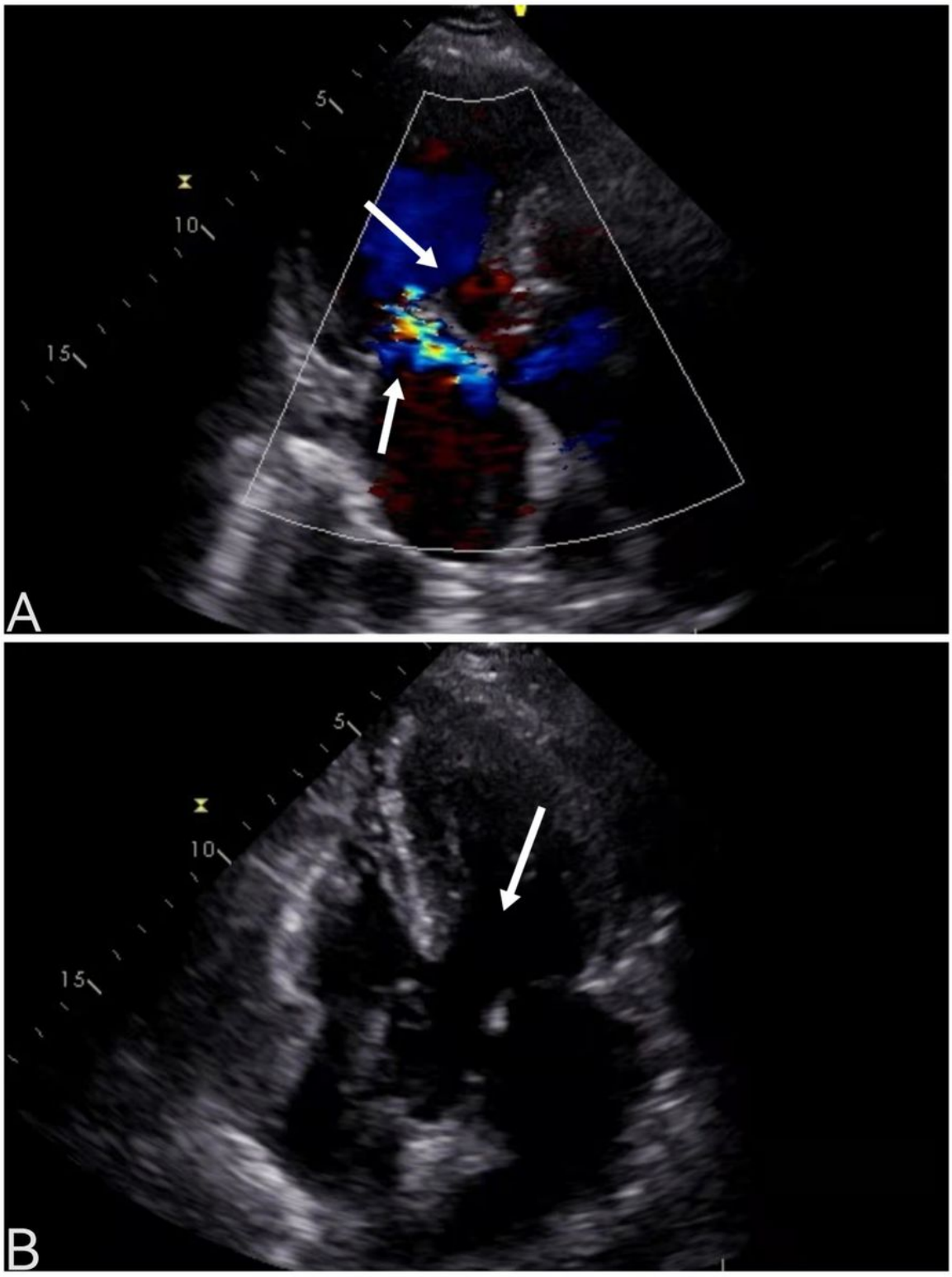

Figure 6

Echocardiography postoperatively

The mitral valve regurgitation was mild (A, the arrow below). LVOT blood flow is unobstructed (A, The arrow above). The SAM sign disappeared (B, arrow).

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- Movie1.avi
- Movie2.avi
- Movie3.avi