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CASE REPORT

ADVANCED

CLINICAL CASE

# Successful Surgical Treatment for Ruptured Aneurysm of Coronary-Pulmonary Artery Fistula Complicated With Cardiac Tamponade

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## ABSTRACT

A 74-year-old woman with no past medical history showed cardiac tamponade caused by rupture of a coronary-pulmonary artery fistula–related aneurysm. Preoperative pericardial puncture and multidetector computed tomography imaging enabled patient condition optimization and accurate morphologic evaluation of fistula and aneurysm, leading to complete surgical resection of the aneurysm. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2022;4:1283-1287) © 2022 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

## INTRODUCTION

Coronary-pulmonary artery fistula (CPAF) is a rare coronary abnormality that can lead to aneurysm development, which can subsequently rupture and cause cardiac tamponade.[1] Reported here is successful management of a case of ruptured CPAF aneurysm complicated with cardiac tamponade.

## LEARNING OBJECTIVES

- To review diagnostic modalities for accurate evaluation of the complex morphology of CPAF aneurysm.
- To recognize the importance of preoperative evaluation for complete resection of CPAF aneurysm.

## PAST MEDICAL HISTORY

A 74-year-old woman with no past medical history presented with lost consciousness on rest and low blood pressure. Contrast computed tomography (CT) performed by a primary physician revealed pericardial effusion and an enhanced mass sized 1 cm at the left lateral side of the main pulmonary artery (PA) (**Figure 1**). Under suspicion of a ruptured coronary artery aneurysm, the patient was referred to our hospital.

## INVESTIGATIONS

On admission, the patient's systolic blood pressure was 79 mm Hg and heart rate was 60 beats/min. Electrocardiogram showed normal sinus rhythm and no significant ST-T wave changes. Transthoracic echocardiography revealed significant pericardial effusion and severe compression of the right heart,

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**ABBREVIATIONS AND ACRONYMS**

**CAG** = coronary artery angiography

**CPAF** = coronary-pulmonary artery fistula

**CT** = computed tomography

**LAD** = left anterior descending artery

**MDCT** = multidetector computed tomography

**PA** = pulmonary artery

**RCA** = right coronary artery

indicating cardiac tamponade. Coronary artery angiography (CAG) showed tortuous abnormal vessels originating from the proximal left anterior descending artery (LAD) and right coronary artery (RCA), suggesting that the cardiac tamponade was caused by a ruptured CPAF aneurysm (**Figures 2A and 2B,** Video 1). These fistulae were classified as type A according to the Sakakibara classification,[2] originating from the proximal third of the coronary artery. Although communication of the RCA into the main PA was observed with CAG, regions distal of the abnormal vessel from the LAD and ruptured aneurysm were not enhanced. Following stabilized hemodynamics after pericardial puncture, multidetector CT (MDCT) was performed for further morphologic evaluation of the CPAF and aneurysm, which clearly showed the origin, course, and drainage sites of 2 fistulae, one originating from the LAD and the other from the RCA (**Figures 3A and 3B**). Furthermore, those findings indicated that the CPAF from the LAD formed aneurysmal dilatations with a string-of-beads appearance, in which an aneurysm with low-density hematoma was found (**Figure 3C**). Small abnormal vessels communicating with the RCA and main PA were not dilated and did not have communication with the aneurysms. In this case, catheter-based interventions, including coil embolization, were anatomically difficult because MDCT showed thrombosis filled in the aneurysm, which was the cause of poor enhancement of regions distal to the fistula during CAG. Based on these results, a surgical strategy for complete resection of the aneurysms and ligation of the CPAF was established.

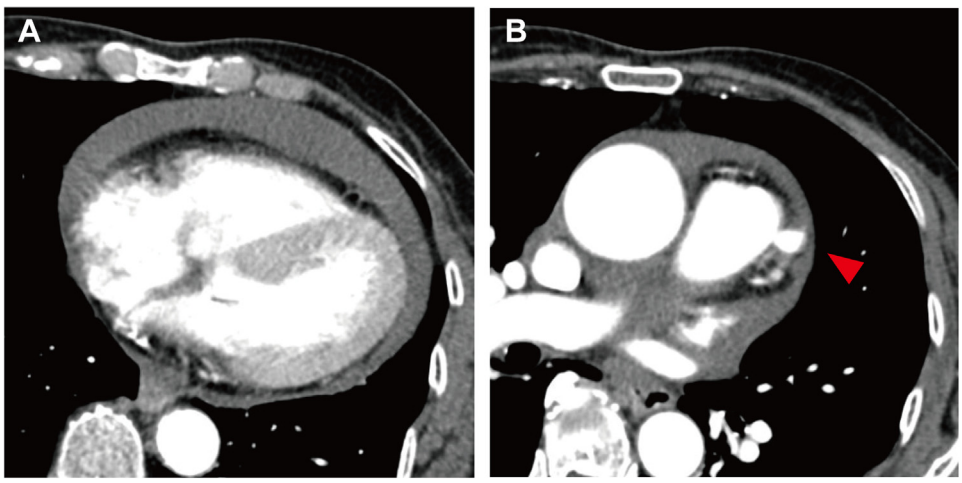
## MANAGEMENT

Nine days after admission, surgery was performed via a median sternotomy (Video 2). After establishing a cardiopulmonary bypass, cardiac arrest was achieved with a retrograde infusion of cardioplegia. The PA trunk was transected, and then abnormal vessels located anterior to the proximal LAD were found below the PA. As the abnormal vessels were dissected distally, an aneurysm 2 cm in diameter was found in which a rupture site was detected (**Figure 4A**). Following its incision (**Figure 4B**), efferent and afferent holes in the aneurysm, as well as the origin of a fistula arising from the proximal LAD, were directly closed with pledgeted 4-0 polypropylene mattress sutures. A drainage hole from the CPAF was found inside of the proximal PA trunk, and that orifice was similarly closed with pledgeted mattress sutures.

## FOLLOW-UP

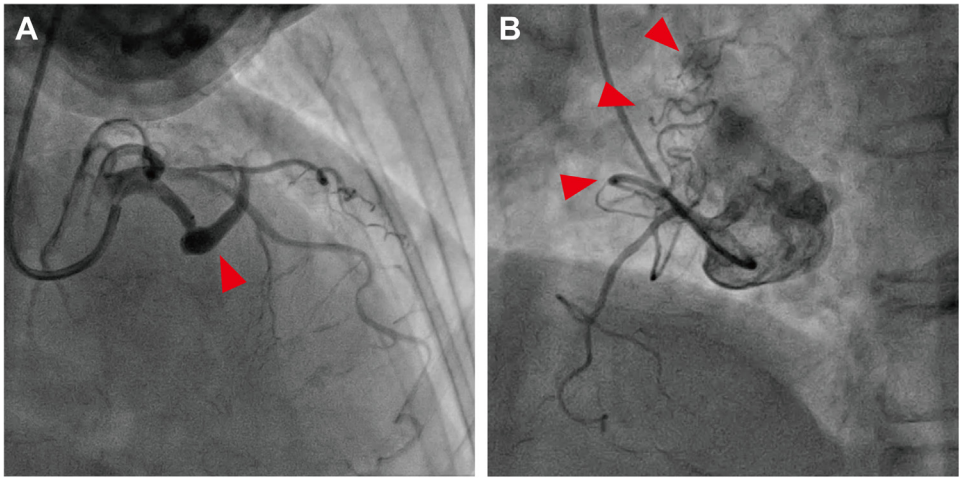
MDCT contrast imaging findings showed that neither the resected aneurysm nor fistulae were enhanced. The patient was discharged uneventfully and found to be doing well at 6 months after surgery.

**FIGURE 1 Initial Computed Tomography**




Initial contrast computed tomography showing **(A)** pericardial effusion and **(B)** an enhanced mass **(red arrowhead)** at the left lateral side of the main pulmonary artery.

**FIGURE 2** **Coronary Angiography**




Coronary angiography revealing **(A)** an abnormal tortuous vessel **(red arrowhead)** originating from left anterior descending artery and **(B)** small tortuous vessels **(red arrowheads)** originating from the right coronary artery that drained into the main pulmonary artery.

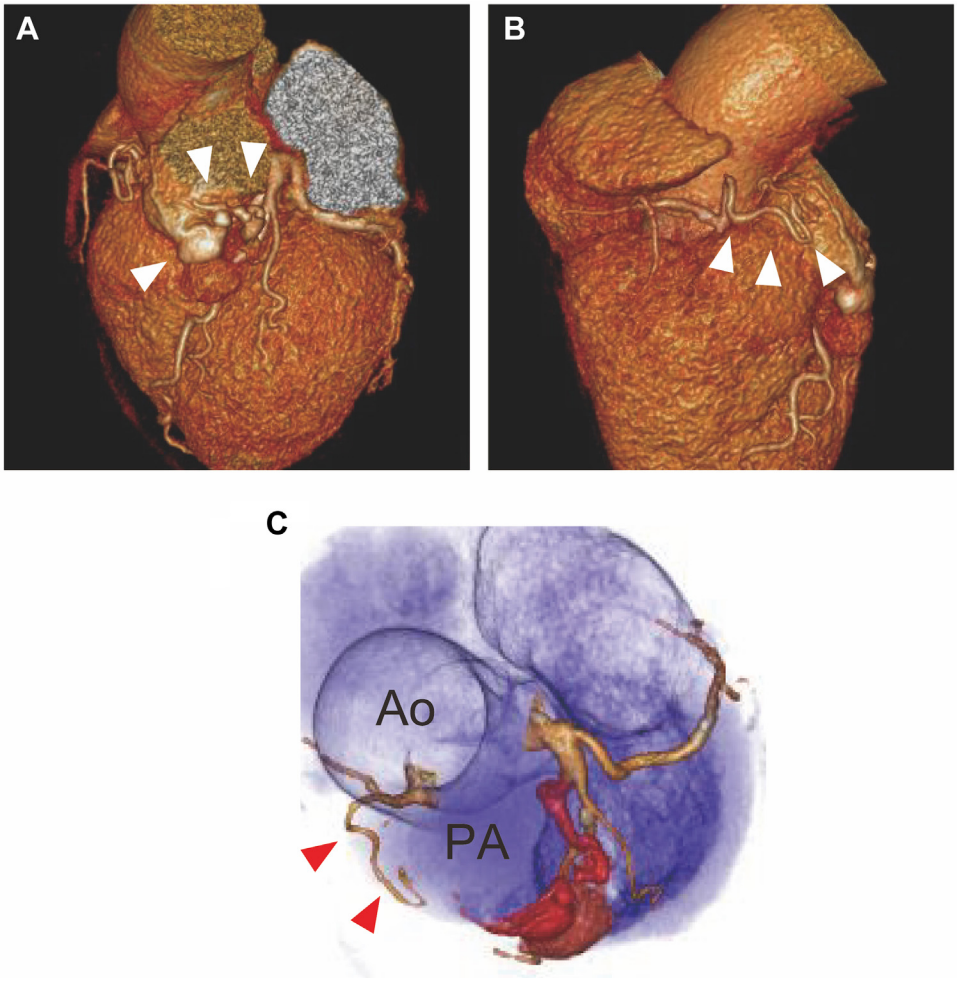
## DISCUSSION

Although 19% to 29% of CPAF cases have been reported to be complicated with an aneurysmal dilatation, rupture of a CPAF aneurysm is rare.[1,3] Generally, coronary artery aneurysm larger than 3 cm in size is an indication for surgery because of risk of rupture.[4] However, in patients with an aneurysm associated with a CPAF, some reports have noted a ruptured aneurysm sized 1 to 2 cm, the same as found in the present case.[4] The risk of rupture of a CPAF aneurysm may be greater compared to that of a native coronary artery aneurysm because of the structural fragility of the fistula. In a pathologic study, smooth muscle was shown to be replaced by fibrous tissue in the vessel media of a fistula.[1] Although rupture of an aneurysm associated with a CPAF is very rare, in patients with cardiac tamponade and abnormal mass around the main PA trunk, a ruptured CPAF aneurysm should be considered, even if the mass is <3 cm in size.

According to previous reports, CPAF cases have morphologic variations. Regarding the origin of a CPAF, a fistula may arise from the LAD, left circumflex artery, or RCA and can also originate from both the left and right coronary arteries.[1,3] As for the drainage site of CPAF, the left lateral side of the main PA trunk is most common (82%), whereas the others reported are the right lateral (12%) and anteromedial (6%) sides.[5] As a result, a CPAF in most cases is found to be located on the left anterolateral side of the PA, with the remaining showing no dominant portion.[3,5] In the present patient, the CPAF originating from the LAD formed an aneurysm, whereas CPAF originating from the RCA was small and did not show communication to the aneurysm. Based on those findings, the origin of the fistula from the LAD was ligated, and the drainage site in the PA was closed. Notably, preoperative MDCT imaging provided vital information for complete resection of the aneurysm.

Prompt diagnosis and treatment are required for patients with a ruptured CPAF aneurysm with cardiogenic shock, but accurate morphologic evaluation is also essential for precise surgical treatment. Fortunately, hemodynamics were dramatically improved after pericardial drainage in the present case, which allowed us to schedule the MDCT and an elective operation. On the other hand, Hijikata et al[6] reported a case of ruptured CPAF aneurysm that required emergency surgery. Nevertheless, even in that acute clinical setting, they were able to immediately evaluate the morphology of the CPAF with MDCT and subsequently performed its complete resection. Evaluation with MDCT followed by pericardial drainage and CAG seems to be an effective strategy for treatment of a ruptured CPAF.

**FIGURE 3 Multidetector Coronary Computed Tomography**




Multidetector coronary computed tomography showing tortuous abnormal vessels originating from **(A)** the left anterior descending artery (LAD) **(white arrowheads)** and **(B)** right coronary artery (RCA) **(white arrowheads)**. **(C)** An abnormal vessel communicating with the proximal LAD and main pulmonary artery (PA) formed aneurysmal dilatations with a string-of-beads appearance (red vessel), whereas small abnormal vessels communicating with the RCA and main PA were neither dilated nor communicating with the aneurysms **(red arrowheads)**.
Ao = ascending aorta.

## CONCLUSIONS

We report successful surgery for a ruptured aneurysm associated with CPAF in a patient with cardiac tamponade. Preoperative MDCT imaging provided vital information for complete resection of the aneurysm. In cases with a ruptured CPAF aneurysm, a smooth sequence that includes hemodynamic stabilization, diagnosis, and surgical treatment is important.

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The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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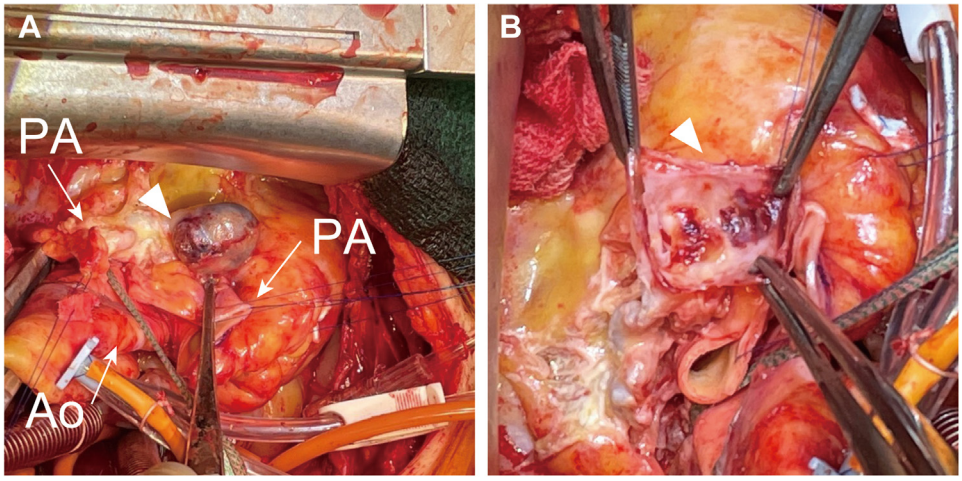
**FIGURE 4** **Intraoperative Lesions**




**(A)** After transection of the pulmonary artery trunk, an aneurysm with a rupture site **(white arrowhead)** was detected. **(B)** The incised aneurysm **(white arrowhead)** after hematoma removal. Ao = ascending aorta; PA = pulmonary artery.

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**KEY WORDS** **bilateral coronary-pulmonary artery fistula, coronary artery aneurysm, coronary computed tomography angiography**

**APPENDIX** **For supplemental videos, please see the online version of this paper.**