


CASE REPORT

Open Access



Giant aneurysm of left circumflex artery branch with fistula to the coronary sinus: a case report

Yasuyuki Toyoda^{1*} , Takahiro Takemura¹, Kazuaki Shiratori¹, Yoshikazu Yazaki², Takahiro Tachibana², Hirokazu Niitsu¹ and Takashi Kunihara³

Abstract

Background: Aneurysm of a coronary artery branch with a fistula is extremely rare. Here, we present a case of giant aneurysm of the left circumflex artery branch with a fistula to the coronary sinus treated successfully with aneurysmectomy.

Case presentation: A 58-year-old woman was referred to our hospital due to an abnormal pericardial mass found by multidetector computed tomography. Imaging examination revealed a dilated left circumflex artery branch with a 30-mm aneurysm. Coronary angiography confirmed a left circumflex artery branch aneurysm with a fistula to the coronary sinus. As percutaneous occlusion of the aneurysm by catheterization was considered unsuccessful, the aneurysm was resected, and the fistula was occluded surgically with excellent outcome. Pathological examination suggested that congenital factors may have contributed to the development of the aneurysm. Computed tomography showed no recurrence of the aneurysm at 1-year postoperative follow-up.

Conclusions: We presented a case of giant aneurysm of the left circumflex artery branch with a fistula to the coronary sinus. This is the first report of the combination of a giant coronary artery branch aneurysm with a fistula to the coronary sinus. Surgical aneurysmectomy should be considered in such cases to avoid fatal aneurysmal complications.

Keywords: Coronary artery fistula, Coronary artery aneurysm, Aneurysmectomy

Background

Coronary artery fistula is an uncommon congenital malformation, with 10% of cases accompanied by a coronary artery aneurysm [1]. Aneurysm of a coronary artery branch with a fistula is extremely rare. Here, we present a case of giant aneurysm of the left circumflex artery (LCX) branch with a fistula to the coronary sinus (CS) and describe our successful surgical strategy.

Case presentation

A 58-year-old asymptomatic woman was referred to our hospital due to an abnormal pericardial mass found incidentally on chest computed tomography (CT) during a medical check-up. On physical examination, she had normal heart sounds, no cardiac murmur, and no lung rales. She had a history of diabetes mellitus, hypertension, and hyperlipidemia, for which she was receiving appropriate medical therapy. She had no history of congenital heart disease, Kawasaki disease, or cardiac trauma, and no relevant family history.

*Correspondence: ciu91590@yahoo.co.jp

¹ Department of Cardiovascular Surgery, Saku Central Hospital Advanced Care Center, 3400-28 Nakagomi Saku, Nagano 385-0051, Japan
Full list of author information is available at the end of the article



Investigation

Electrocardiography showed normal sinus rhythm and no significant ST-T changes. Chest X-ray showed neither cardiomegaly nor cardiac congestion. Transthoracic echocardiography demonstrated normal wall motion with left ventricular ejection fraction (LVEF) of 64%. Multidetector CT revealed a huge coronary branch aneurysm originating from close to the terminal portion of the main LCX trunk with a fistula directly draining to the CS (Fig. 1a, b). The end of the main LCX was very thin, indicating abundant blood flow to the coronary branch aneurysm. Coronary angiography showed a giant branch aneurysm arising from the LCX, whereas the left anterior descending artery and right coronary artery appeared normal (Fig. 2). The distal side of the aneurysm drained into the CS without a capillary bed, indicating the presence of a coronary artery fistula between the aneurysm

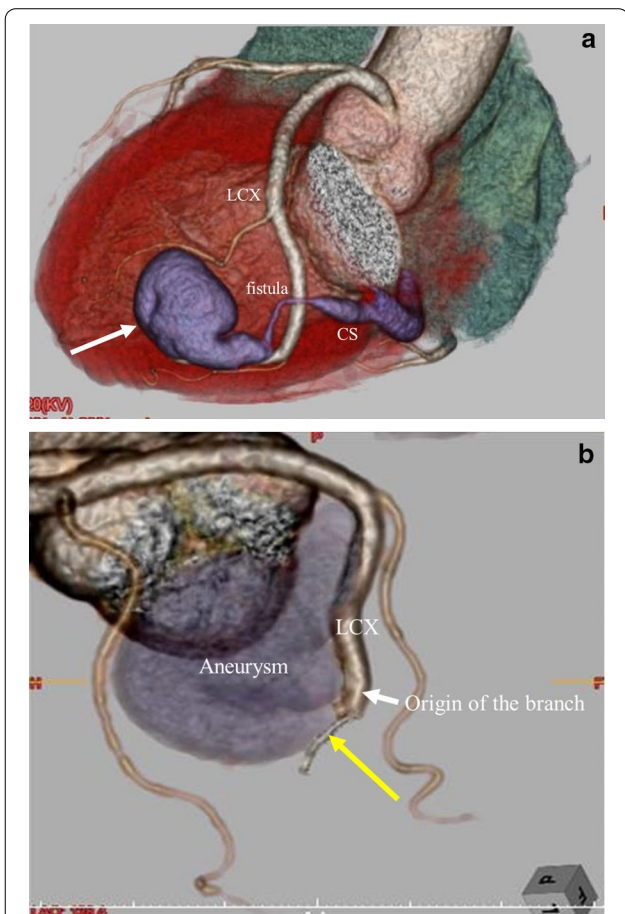


Fig. 1 **a** Preoperative coronary artery computed tomography revealed a giant coronary branch aneurysm originating from near the terminal portion of the main LCX trunk with a fistula draining into the CS (white arrow). **b** The end of the main LCX was very thin (yellow arrow). CS, coronary sinus; LCX, left circumflex artery

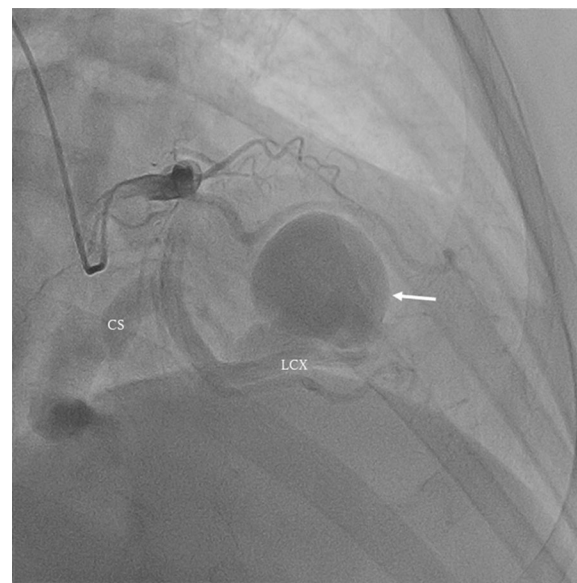


Fig. 2 Preoperative coronary artery angiography showed that the abnormal aneurysm branched from the end of the LCX (white arrow). The aneurysm drained into the CS. CS, coronary sinus; LCX, left circumflex artery

and the CS. The trunk of the LCX itself was of normal caliber, but it became aneurysmal about 5 mm distal to its branch, measuring 30 mm in diameter. There were no side branches arising from the aneurysm. Percutaneous occlusion of the fistula by catheterization was initially considered to reduce the blood flow into the giant aneurysm, but this was difficult because cannulation was impossible. The heart team referred the patient for surgical closure.

Management

Surgical treatment was performed through a median sternotomy. Cardiopulmonary bypass was established using aortic cannulation and bicaval cannulation, and the aneurysm was found at the branch of the LCX (Fig. 3). After clamping of the ascending aorta, cardiac arrest was achieved by antegrade cardioplegia. The apex was lifted, the aneurysm was incised, and its proximal and distal orifices were closed with 5–0 polypropylene sutures. The aneurysm was excluded, and the patient was smoothly weaned from cardiopulmonary bypass. The operation time, cardiopulmonary bypass time, and aortic clamp time were 174, 71, and 25 min, respectively.

The postoperative course was uneventful with no clinically significant changes on electrocardiography. Postoperative blood examination revealed no relevant elevation of serum creatinine kinase MB level (max. 11 IU /L). Postoperative multidetector CT demonstrated absence of

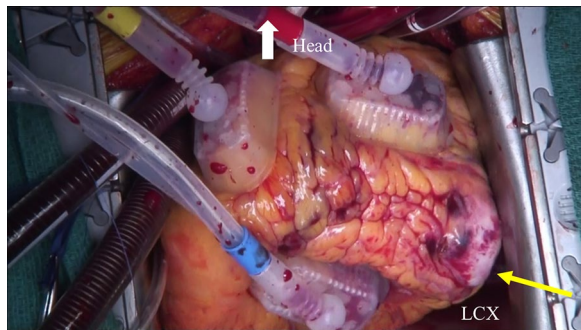


Fig. 3 Operative view of the LCX branch aneurysm with fistula (yellow arrowhead). LCX, left circumflex artery

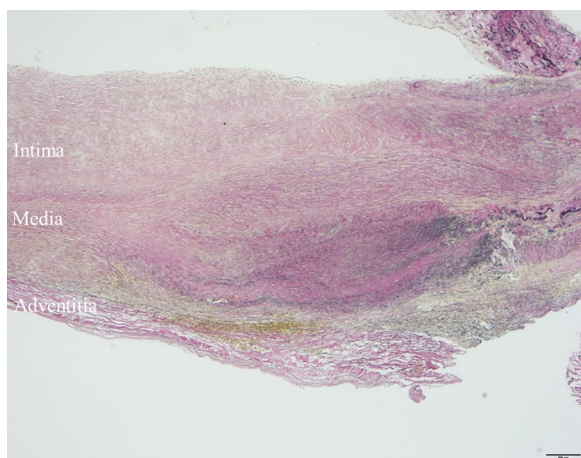


Fig. 4 The resected left circumflex artery branch aneurysm specimen had a thickened intima with no hyaline arteriosclerosis, thin fibrous media with small amounts of muscular components, and no internal elastic membrane. Stain: Elastin van Gieson

the fistula and no recurrence of the aneurysm. CT also showed no expansion of the LCX branch aneurysm at 1-year postoperative follow-up. Pathological examination of the surgical specimen revealed a thickened intima without hyaline arteriosclerosis, thin media with minimal smooth muscle, and lack of an internal elastic membrane, which was markedly different from the typical media. These findings suggested that inherent weakness of the coronary branch artery wall may have contributed to the aneurysm (Fig. 4).

Discussion and conclusions

Coronary artery aneurysm with an associated fistula (CAAAF) is a rare congenital malformation. Neufeld et al. [2] first reported a surgical case of CAAAF in a 10-year-old girl in 1961. In 1963, Habermann et al. [3] reported a CAAAF in an adult patient who died of

aneurysm rupture. The number of cases of CAAAF has gradually increased with the widespread adoption of coronary angiography but reports of this disease are still limited. Coronary artery branch aneurysm with a fistula is extremely rare. Hirose et al. [1] performed a literature review of 45 reports of CAAAF, of which 23 were written in Japanese, and 12 of the 22 reports written in English were from Japan. The authors found that CAAFs occurred more often in female than in male patients. One third of the patients with CAAFs were asymptomatic and were usually diagnosed incidentally by cardiac murmur or abnormalities on chest radiographs. The clinical presentation depends on the severity of the left-to-right cardiac shunt. Clinical symptoms, such as fatigue, dyspnea, angina, myocardial ischemia, or myocardial infarction, may appear as the shunt enlarges. The LCX is the origin of the fistula in 18.3% of cases with a coronary artery fistula, with the left anterior descending artery and the right coronary artery accounting for 42% and 50–60% of cases, respectively. The drainage site of the fistula is the CS in 7% of cases [4].

Libertini et al. [5] and Kemmochi et al. [6] reported cases of giant LCX aneurysms with CS fistulas. Almansori and Tamim [7] also reported a case of a giant coronary artery fistula from the LCX to the CS. However, these cases involved a main LCX aneurysm, whereas our case involved a branch aneurysm arising from the LCX. Edwards et al. [8] reported a case of a giant CS with a focal aneurysm secondary to multiple fistulous connections arising from a dilated tortuous LCX. However, the LCX in our patient was of normal caliber and the branch of the LCX was aneurysmal, which were different from these former cases. To our knowledge, this is the first report of the combination of a giant coronary artery branch aneurysm with a fistula to the CS.

Previous reports have also suggested that surgery should be performed for all patients with a CAAAF, even when asymptomatic, due to the risk of future development of cardiac complications, such as rupture, ischemia, or thrombosis [1]. Indeed, Yoshino et al. reported rupture of a coronary artery fistula to the coronary sinus with giant aneurysm of the coronary artery [9]. We followed the strategy recommended in these previous reports. El Nihum et al. recently reported transcatheter embolization of a coronary artery fistula [10]. However, we did not select this procedure as our case involved a coronary branch aneurysm and we considered that cannulation of the fistula would have been difficult and embolization of the LCX main trunk would be associated with a risk of ischemia. Moreover, transcatheter embolism would not prevent aneurysm rupture, and coil embolization could affect the blood supply to the normal coronary artery. In our case, there were no coronary side branches

arising from the aneurysm that would necessitate coronary bypass grafting, and we therefore performed closure of the orifices in the aneurysm.

The precise mechanisms underlying aneurysm development with a coronary artery fistula are unknown. In our case, pathological examination of both the fistula and aneurysm revealed congenital structural weakness of the coronary artery wall, which may have contributed to the development of the CAAAF. Genetic factors may also be associated with the process of aneurysm formation [1].

We reported the successful surgical treatment of a giant LCX branch aneurysm with a fistula to the CS. Surgical aneurysmectomy should be considered in such cases to avoid fatal aneurysmal complications.

Abbreviations

CAAAF: Coronary artery aneurysm with an associated fistula; CS: Coronary sinus; CT: Computed tomography; LCX: Left circumflex artery; LVEF: Left ventricular ejection fraction.

Acknowledgements

None.

Author contributions

YT: Writing original draft, editing. TT, KS, HN, YY, and TT: review and editing. TK: Supervision. All authors read and approved the final manuscript.

Funding

None.

Availability of data and materials

All data generated or analyzed during this study are included in this published article. Data sharing is not applicable to this article as no datasets were generated or analyzed during the present study.

Declarations

Ethics approval and consent to participate

This was a case report and the requirement for approval from the Institutional Review Board of Saku Central Hospital Saku Sogo Byoin was waived.

Consent for publication

The patient provided informed consent for the publication of this report and accompanying images.

Competing interests

The authors declare that they have no competing interests.

Author details

¹Department of Cardiovascular Surgery, Saku Central Hospital Advanced Care Center, 3400-28 Nakagomi Saku, Nagano 385-0051, Japan. ²Department of Cardiology, Saku Central Hospital Advanced Care Center, Nagano, Japan. ³Department of Cardiac Surgery, Jikei University School of Medicine, Tokyo, Japan.

Received: 1 March 2022 Accepted: 15 August 2022

Published online: 21 August 2022

References

1. Hirose H, Amano A, Yoshida S, Nagao T, Sunami H, Takahashi A, et al. Coronary artery aneurysm associated with fistula in adults: collective review and a case report. *Ann Thorac Cardiovasc Surg.* 1999;5:258–64.

- Neufeld HN, Lester RG, Adams P Jr, Anderson RC, Lilleheri CW, Edwards JE. Congenital communication of coronary artery with a cardiac chamber or the pulmonary trunk ("coronary artery fistula"). *Circulation.* 1961;24:171–9. <https://doi.org/10.1161/01.CIR.24.2.171>.
- Habermann JH, Howard ML, Johnson ES. Rupture of the coronary sinus with hemopericardium: a rare complication of coronary arteriovenous fistula. *Circulation.* 1963;28:1143–4.
- Mangukia CV. Coronary artery fistula. *Ann Thorac Surg.* 2012;93:2084–92.
- Libertini R, Wallbridge D, Jones HR, Gunning M, Satur CMR. Giant circumflex artery with a coronary sinus fistula. *Ann Thorac Surg.* 2018;106:e223–5.
- Kenmochi R, Ohga Y, Kubo Y, Matsumoto M. Left circumflex artery aneurysm with fistula to coronary sinus. *Gen Thorac Cardiovasc Surg.* 2020;68:1196–8. <https://doi.org/10.1007/s11748-019-01247-8>.
- Almansori M, Tamim M. Giant coronary artery fistula. *Asian Cardiovasc Thorac Ann.* 2014;22:595–7. <https://doi.org/10.1177/0218492313478627>.
- Edwards NFA, Wijesekera VA, Anderson BA, Habibian M, Burstow DJ, Walters DL, et al. A rare case of a giant coronary sinus with focal aneurysm secondary to multiple fistulous connections arising from a dilated, tortuous left circumflex coronary artery. *CASE (Phila).* 2018;2:99–102. <https://doi.org/10.1016/j.case.2017.12.004>.
- Yoshino S, Minagoe S, Yu B, Kosedo I, Yamashita M, Ishizawa M, et al. Cardiac tamponade due to rupture of coronary artery fistula to the coronary sinus with giant aneurysm of coronary artery: usefulness of transthoracic echocardiography. *Heart Vessels.* 2013;28:536–40. <https://doi.org/10.1007/s00380-012-0279-9>.
- El Nihum LI, Thakkar AN, Chinnadurai P, Lin CH. Successful coil embolization of a large right coronary artery-coronary sinus fistula causing a significant left-to-right shunt: a case report. *Eur Heart J Case Rep.* 2021;5:1–5. <https://doi.org/10.1093/ehjcr/ytab121>.

Publisher's Note

Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

Ready to submit your research? Choose BMC and benefit from:

- fast, convenient online submission
- thorough peer review by experienced researchers in your field
- rapid publication on acceptance
- support for research data, including large and complex data types
- gold Open Access which fosters wider collaboration and increased citations
- maximum visibility for your research: over 100M website views per year

At BMC, research is always in progress.

Learn more biomedcentral.com/submissions

