OPEN

Giant Coronary Artery Aneurysm With Fistula to the Pulmonary Artery Complicated by Frequent Ventricular Premature Contractions

A Case Report

Hao Cao, MD, PhD, Liang Ye, MD, Paul Chan, MD, PhD, Huimin Fan, MD, PhD, and Zhongmin Liu, MD, PhD

Abstract: Giant coronary artery aneurysm with a fistula is a rare condition. The presence of a giant aneurysm imposes considerable health risks.

We report a case of a 67-year-old woman who presented with frequent ventricular premature contractions caused by a giant coronary aneurysm arising from a branch of the left anterior descending coronary artery that had a fistulous connection to the pulmonary artery.

The patient was referred for cardiac surgery. The giant aneurysm was resected, and the proximal and distal openings were closed directly. The main pulmonary artery was opened longitudinally and the fistula was also closed directly.

The patient's symptoms of frequent ventricular premature contractions disappeared postoperatively as confirmed by electrocardiography.

Although the standard therapeutic strategies of the disease are not well established because of the rarity of this condition, our clinical results indicate that the surgical treatment is an effective choice.

(*Medicine* 94(7):e530)

Abbreviations: CAA = coronary artery aneurysms, CABG = coronary artery bypass grafting, CAF = coronary artery fistula, LAD = left anterior descending, PA = pulmonary artery, RCA = right coronary artery.

Editor: Dai Yamanouchi.
Received: October 12, 2014; revised: January 9, 2015; accepted: January 12, 2015.
From the Department of Cardiovascular Surgery (HC, LY, HF, ZL), Shanghai East Hospital, Tongji University School of Medicine, Shanghai, China; and Cardiology Division, Department of Medicine (PC), Wan Fang Hospital, Taipei Medical University, Taipei, Taiwan.
Correspondence: Huimin Fan, Department of Cardiovascular Surgery, Shanghai East Hospital, Tongji University School of Medicine, Shanghai 200120, China (e-mail: frankfan64@hotmail.com).
HC and LY contributed equally to this work.
Specialized Research Fund for the Doctoral Program of Higher Education of China (20130072120062), Shanghai focus on specialties of medicine (ZK2012A27), Shanghai leading talents (2012035), Shanghai Municipal Science and Technology Commission projects (12441902500), funded by Key Disciplines Group Construction Project of Pudong Health Bureau of Shanghai (Grant No. PKzxkq2014–01), funded by Outstanding Leaders Training Program of Pudong Health Bureau of Shanghai (Grant No. PKR2011–01), funded by Research Grant for Health Science and Technology of Pudong Health and Family Planning Commission of Shanghai (Grant No. PW2013A-2), funded by Health industry Project of Pudong Health Bureau of Shanghai (Grant No. PW2013E-1), funded by Qihang Talents Training Program of Shanghai East Hospital, Tongji University (Grant No. DFQH-M4).
The authors have no conflicts of interest to disclose.

ISSN: 0025-7974
DOI: 10.1097/MD.0000000000000530

INTRODUCTION

Coronary artery aneurysms (CAA) are localized dilatations of the coronary artery exceeding the diameter of the adjacent normal segment by 50% and are detected incidentally in only 0.15% to 4.9% of patients who undergo coronary angiography.[1] ''Giant'' CAA, which are defined as having a diameter of 2 cm or greater, are rarer with a reported prevalence of 0.02% and are more likely to develop complications.[2,3] Many patients with CAA are asymptomatic and are diagnosed during coronary angiography while undergoing investigation for some other pathology. Giant aneurysms, however, may become symptomatic; size and location determine their manifestations. CAAs can present with myocardial ischemia due to thrombosis secondary to sluggish blood flow in the aneurysm. Coronary artery fistula (CAF) is also a rare coronary artery abnormality in which blood is shunted into a cardiac chamber, great vessel, or other structure with an incidence ranging from 0.1% to 0.8% in the adult population.[4] In adulthood, the clinical importance of CAFs is due to an increased risk of complications, including heart failure, myocardial ischemia, infective endocarditis, arrhythmias, and rupture.[5] Although evidence-based treatment guidelines are limited, surgical resection may be selected due to potential life-threatening complications, such as rupture, thrombosis, heart failure, myocardial ischemia, infective endocarditis, arrhythmias, and distal coronary embolization.

CASE REPORT

A 67-year-old woman was referred to our hospital due to frequent ventricular premature contractions for 2 months. Her medical history was unremarkable from the cardiologic point of view. Initial vital signs showed a body temperature of 36.8°C, blood pressure of 142/83 mm Hg, respiration rate of 18/min, and heart rate of 85 beats/min. A continuous Levine grade 2/6 cardiac murmur was heard at the second intercostal space at the right sternal border. Chest x-ray was normal and electrocardiography revealed frequent ventricular premature contractions. Laboratory studies showed a white blood cell count of 6.9×10^9/L, hemoglobin level of 137 g/L, platelet count of 225×10^9/L, C-reactive protein concentration of <1 mg/L, and the serum N-terminal pro-B-type natriuretic peptide level was 302.20 ng/L. Arterial blood gas analysis evaluated under room air showed partial pressures of oxygen and carbon dioxide of 89.1 mm Hg and 35.8 mm Hg, respectively. The transthoracic and transesophageal echocardiography showed a large cystic mass containing smoke-like echoes adjacent to the ectatic left anterior descending artery. Blood flow into the pulmonary artery (PA) could be observed as color Doppler signals (Figure 1A and B). Axial section of the computed tomography scan showed a CAA with the dimensions of 32×33 mm

(Figure 1C). Three-dimensional reconstruction of the multislice CT scan images showed a giant coronary aneurysm located near the proximate-portion of the LAD, and the CAF was dilated and tortuous and connected with the PA (Figure 1D). Coronary angiography showed a giant aneurysm originating from a branch of the LAD, which had a fistulous connection to the PA (Figure 1E). Electrocardiogram showed sinus rhythm with frequent ventricular premature contractions (Figure 1G).

Under cardiopulmonary bypass via median sternotomy, the giant aneurysm located at a coronary artery branching from the left anterior descending (LAD) was opened and resected with no intra-aneurysmal thrombus found, and the proximal and distal openings were closed directly. Coronary artery bypass graft surgery was not performed. The main PA was opened longitudinally and the fistula was also closed directly. The patient's symptoms of frequent ventricular premature contractions disappeared postoperatively as confirmed by electrocardiography (Figure 1H), and the patient was discharged 2 weeks after surgery. Postoperative coronary angiography revealed the CAA and CAF had disappeared (Figure 1F).

The case study was approved by the Institutional Review Board or Ethics Committee of Shanghai East Hospital, Tongji University School of Medicine, Shanghai, China. Informed consent was obtained from the patient before data collection.

DISCUSSION

CAAs are usually discovered incidentally with right coronary artery (RCA) lesions accounting for 50% of the total, and aneurysms of the left main coronary artery are rare, contributing less than 1% of all CAAs.[6] CAA can be asymptomatic. However, they can portend life-threatening complications, such as thrombosis with possible myocardial infarction,[7] rupture that

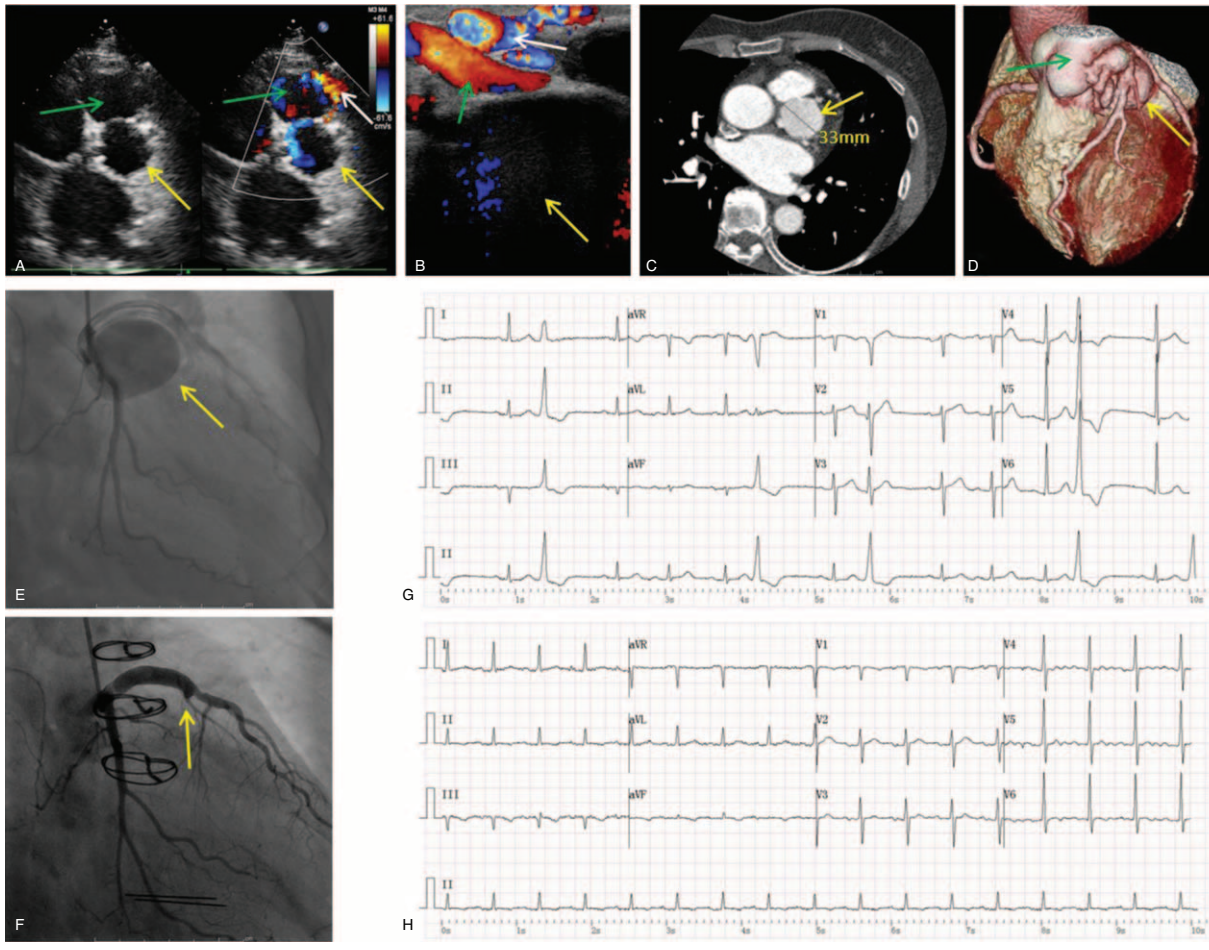

FIGURE 1. Preoperative cardiac images. A and B. Two-dimensional transthoracic and transesophageal echocardiography. A large cystic mass containing smoke-like echoes was observed adjacent to the ectatic left anterior descending artery. Blood flow into the pulmonary artery could be observed as color Doppler signals (yellow arrow: coronary artery aneurysm, white arrow: coronary artery fistula, green arrow: pulmonary artery) C. Axial section of CT image. Coronary artery aneurysm (dimensions 32 × 33 mm) with heterogenic contrast enhancement is seen (yellow arrow). D. Three-dimensional heart reconstruction of CT images. A giant coronary artery aneurysm was present at the branch artery of the left anterior descending artery (LAD), and the proximal portion of LAD was dilated and tortuous (yellow arrow: coronary artery aneurysm, green arrow: pulmonary artery). E. Coronary angiography. Left coronary artery angiography demonstrated a giant coronary aneurysm originating from branch of LAD with a fistulous connection to the pulmonary artery (yellow arrow). Postoperative cardiac images. F. Coronary angiography. The coronary artery aneurysm and coronary artery fistula disappeared (yellow arrow). Electrocardiography: G. Preoperative electrocardiography revealed frequent ventricular premature contractions. H. Postoperative electrocardiography revealed no frequent ventricular premature contractions.

results in cardiac tamponade,[8] rapid increase in the size of aneurysm that results in increasing the risk of rupture,[9] or compression of the atria or ventricles with subsequent heart failure.[10] In addition, some literatures confirmed that cardiac compression could lead to arrhythmia.[11–13] Therefore, surgery should be considered, certainly in the case of a giant, symptomatic aneurysm.[14] Various surgical techniques can be applied, such as isolating or resecting the aneurysm and reconstructing the coronary course, for instance by using an interpositional graft or by maintaining distal coronary flow via concomitant coronary artery bypass grafting (CABG).

CAFs are rare, and the RCA is more commonly affected than the left coronary vessels. The fistulae drain into the right cardiac chambers more commonly than the left, with the most common drainage sites being the right ventricle, right atrium, or PA, and less frequently the coronary sinus, left atrium, left ventricle, or superior vena cava.[15,16] The clinical presentation of CAFs is mainly dependent on the severity of the left-to-right shunt.[17] The majority of adult patients are usually asymptomatic; however, a smaller percentage of pediatric patients tend to be asymptomatic.[18] This may be the result of the larger fistulas being more likely to cause symptoms in the pediatric age group. The clinical presentations include fatigue, dyspnea, orthopnea, angina, endocarditis, arrhythmias, stroke, myocardial ischemia, or myocardial infarction.[17,18] Rarely, pericardial effusion and sudden death were found as a presenting feature in these patients.[19,20] Potential complications if a large left-to-right shunt exists are pulmonary hypertension and congestive heart failure; others include rupture or thrombosis of the fistula or associated arterial aneurysm or coronary steal phenomena.

The main indications for closure are clinical symptoms especially of heart failure and myocardial ischemia, and in asymptomatic patients with high-flow shunting, to prevent occurrence of symptoms or complications, especially in the pediatric population.[17] Surgery and catheter techniques can be used to close the fistulous connection, and both approaches have similar early effectiveness, morbidity, and mortality.[21] Patients treated conservatively should be followed up closely for appearance of symptoms. Most of the adult patients who are asymptomatic remain free of symptoms for long periods.[22,23]

We report a patient who was referred to our hospital due to frequent ventricular premature contractions, and who was eventually found to have a giant CAA originating from the LAD with a fistulous connection to the PA. Surgical treatment was selected because of the presence of giant CAA and frequent ventricular premature contractions caused by CAF associated with giant CAA causing a steal syndrome. Surgical removal of the aneurysm and closure of the proximal opening by epicedial patch were successfully performed, which led to disappearance of the patient's symptoms of frequent ventricular premature contractions. Due to the rarity of such a condition, selection of appropriate therapeutic strategies and investigation of underlying conditions should be carefully performed.

ACKNOWLEDGMENTS

The authors specially thank the staff of the department of echocardiograph and radiology for the diagnosis of the disease.

REFERENCES

1. Syed M, Lesch M. Coronary artery aneurysm: a review. *Prog Cardiovasc Dis.* 1997;40:77–84.
2. Alcock R, Naoum C, Ng AC. Giant right coronary aneurysm: a case of mistaken identity. *Eur Heart J.* 2011;32:2712.
3. Holinski S, Dohmen PM, Lembcke A, et al. Surgical management of multiple coronary artery aneurysm, including giant form. *Tex Heart Inst J.* 2009;36:238–240.
4. Fernandes ED, Kadivar H, Hallman GL, et al. Congenital malformations of the coronary arteries: the Texas Heart Institute experience. *Ann Thorac Surg.* 1992;54:732–740.
5. Vitarelli A, De Curtis G, Conde Y, et al. Assessment of congenital coronary artery fistulas by transesophageal color Doppler echocardiography. *Am J Med.* 2002;113:127–133.
6. Everett JE, Burkhart HM. Coronary artery aneurysm: case report. *J Cardiothorac Surg.* 2008;3:1.
7. Chiang JY, Wu CK, Luo JL, et al. A giant left main coronary artery aneurysm with fistula to the pulmonary artery, manifested with ST-segment elevation myocardial infarction. *JACC Cardiovasc Interv.* 2014;7:e55–e56.
8. Wan S, LeClerc JL, Vachiery JL, et al. Cardiac tamponade due to spontaneous rupture of right coronary artery aneurysm. *Ann Thorac Surg.* 1996;62:575–576.
9. Bajaj A, Sehgal V, Bajwa SJ, et al. Expanding giant right coronary artery aneurysm: an acute need for new management strategies. *Heart Views.* 2014;15:13–15.
10. Kumar G, Karon BL, Edwards WD, et al. Giant coronary artery aneurysm causing superior vena cava syndrome and congestive heart failure. *Am J Cardiol.* 2006;98:986–988.
11. Flores-Ríos X, Campo-Pérez R, Rodríguez Fernández JÁ. Giant aneurysm of the right coronary artery: a rare cause of atrial fibrillation. *Rev Esp Cardiol (Engl Ed).* 2013;66:993.
12. Starck CT, Zientara A, Alkadhi H, et al. Uncommon cause of ventricular tachycardia in a patient with a giant pseudoaneurysm of the ascending aorta. *Thorac Cardiovasc Surg.* 2013;61:603–605.
13. Krishnasamy P, Gillespie JS, McGlinchey PG, et al. *Giant true saphenous vein graft aneurysm causing cardiac compression: a rare cause of atrial flutter Int J Cardiol.* 2007;121:317–319.
14. John LC, Hornick P, Davies DW, et al. The role of surgery in the management of solitary coronary artery aneurysm. *Eur J Cardiothorac Surg.* 1991;5:440–441.
15. Nakamura M, Matsuoka H, Kawakami H, et al. Giant congenital coronary artery fistula to left brachial vein clearly detected by multidetector computed tomography. *Circ J.* 2006;70:796–799.
16. Mangukia CV. Coronary artery fistula. *Ann Thorac Surg.* 2012;93:2084–2092.
17. Balanescu S, Sangiorgi G, Castelvecchio S, et al. Coronary artery fistulas: clinical consequences and methods of closure: a literature review. *Ital Heart J.* 2001;2:669–676.
18. Wang NK, Hsieh LY, Shen CT, et al. Coronary arteriovenous fistula in pediatric patients: a 17-year institutional experience. *J Formos Med Assoc.* 2002;101:177–182.
19. Lau G. Sudden death arising from a congenital coronary artery fistula. *Forensic Sci Int.* 1995;73:125–130.
20. Ozeki S, Utsunomiya T, Kishi T, et al. Coronary arteriovenous fistula presenting as chronic pericardial effusion. *Circ J.* 2002;66:779–782.
21. Armsby LR, Keane JF, Sherwood MC, et al. Management of coronary artery fistulae. Patient selection and results of transcatheter closure. *J Am Coll Cardiol.* 2002;39:1026–1032.
22. Said SA, Landman GH. Coronary-pulmonary fistula: long-term follow-up in operated and non-operated patients. *Int J Cardiol.* 1990;27:203–210.
23. Sunder KR, Balakrishnan KG, Tharakan JA, et al. Coronary artery fistula in children and adults: a review of 25 cases with long-term observations. *Int J Cardiol.* 1997;58:47–53.