Surgical treatment of giant coronary artery aneurysm

Dianyuan Li, MD,^{a*} Qingyu Wu, MD,^{a*} Lizhong Sun, MD,^a Yunhu Song, MD,^a Wei Wang, MD,^a Shiwei Pan, MD,^a Guohua Luo, MD,^a Yongmin Liu, MD,^a Zhitao Qi, MD,^a Tianfu Tao, MD,^a Jian-Zhong Sun, MD,^b and Shengshou Hu, MD^{a*}

Dr Hu

Objective: Giant coronary artery aneurysm is an extremely uncommon disease. Most previous reports have involved only single cases. This report describes 6 patients with giant coronary artery aneurysm, examines its causes, and aims to establish the optimal surgical strategies for this exceptional and rare pathology.

Methods: From July 1996 to October 2004, a total of 30,268 patients underwent heart surgery at Fuwai Hospital in Beijing. Among these, 6 patients had giant coronary artery aneurysm diagnosed and underwent operation. Various surgical strategies were used for the operations of these 6 patients, such as coronary artery aneurysm resection, coronary artery reconstruction, and concomitant coronary bypass. Additional procedures, such as fistula closure, aortic valve replacement, aortoplasty, and embolectomy, were done at same time for the patients with complications of coronary fistula, aortic valve insufficiency, or thrombus. Patients were followed up from 8 to 87 months, with a mean of 48 months. Doppler echocardiography, ultrafast computed tomography, and 3-dimensional aerial image studies were performed during follow-up.

Results: Five of these six cases were found combined with coronary artery fistula, and the cause for these giant coronary artery aneurysms was congenital. The remaining case was caused by atherosclerosis. After surgery, all patients recovered uneventfully, without in-hospital mortality. None died during the follow-up, nor did any have recurrence of the symptoms or giant coronary artery aneurysm.

Conclusion: Giant coronary artery aneurysm is a rare entity that is commonly caused by congenital malformation and combined with other cardiac anomalies. An optimal surgical operation should be based on the specific cardiac anomaly of the individual patient.

From the Department of Cardiovascular Surgery, Cardiovascular Institute and Fuwai Hospital, Chinese Academy of Medical Science and Peking Union Medical College, Beijing, The People's Republic of China,^a and the Department of Anesthesiology, Thomas Jefferson University, Philadelphia, Pa.^b

Received for publication Feb 16, 2005; revisions received March 26, 2005; accepted for publication April 5, 2005.

Address for reprints: Dianyuan Li, MD, Department of Surgery, Fuwai Hospital, 167 Bei Li Shi RD, Xicheng District, Beijing 100037, The People's Republic of China (E-mail: lidianyuan@founder.com).

*These authors are equal contributors. S.H. is co-corresponding author.

J Thorac Cardiovasc Surg 2005;130:817-21
0022-5223/\$30.00

Copyright © 2005 by The American Association for Thoracic Surgery

doi:10.1016/j.jtcvs.2005.04.004

Coronary artery aneurysm is an uncommon disease, found in only 0.15% to 4.9% of patients who undergo coronary angiography.¹ Coronary artery aneurysm is defined as coronary dilation that exceeds the diameter of normal adjacent segments or the diameter of the patient's largest coronary vessel by 1.5 times. On rare occasions, a coronary artery aneurysm grows large enough to be called *giant coronary artery aneurysm*, for which a precise definition is still lacking.^{2,3} In the literature, the reported diameter of giant coronary artery aneurysm in adults varies from 50 to 150 mm. In a review of the literature, most previous reports on giant coronary artery aneurysm have only involved single cases.³ Here for the first time we describe the clinical features and surgical treatments of 6 patients with giant coronary artery aneurysm from a single hospital, Fuwai Hospital in Beijing, China.

Clinical Summary

Patients

As noted previously, the definition of giant coronary artery aneurysm remains unclear. In this report, on the basis of clinical presentations and pathologic findings, we have used the

TABLE 1. Cases of giant coronary artery aneurysm

Case	Sex	Cause	Origin of CAA	Fistula	Size (mm)	Additional procedures				
						1	2	3	4	5
1	M	Congenital	RCA	LV	138 × 121 × 110	CABG × 2		AVR		
2	F	Congenital	LM + LAD	LV	130 × 112 × 92	CABG × 4				
3	F	Congenital (operation 24 y previously)	RCA	LV	50 × 35 × 27				AP	Embolectomy
4	M	Atherosclerotic	LAD	—	22 × 18 × 15		CARC	AVR		
5	M	Congenital	RCA	LV	56 × 54 × 52		CARC	AVR	AP	
6	M	Congenital	Diagonal	LV	45 × 43 × 40		CARC			

CAA, Coronary artery aneurysm; RCA, right coronary artery; LV, left ventricle; CABG, coronary artery bypass grafting; AVR, aortic valve replacement; LM, left main coronary artery; LAD, left anterior descending coronary artery; CARC, coronary artery reconstruction; AP, aortoplasty.

standard of a diameter of more than 20 mm to define a giant coronary artery aneurysm. From July 1996 through October 2004, a total of 30,268 patients underwent heart surgery in our hospital, and 6 of them were found to have giant coronary artery aneurysms according to this standard. The incidence was thus 0.02% among our surgical patients. An additional 7 patients were found to have coronary artery aneurysms, but in all cases the diameter of the coronary artery aneurysm was less than 18 mm and the cause of the pathologic changes was Kawasaki disease. Those 7 patients were therefore excluded from this report.

Clinical Presentation

Clinical features of 6 patients (2 female and 4 male) with giant coronary artery aneurysms are presented in Table 1. The mean age of patients was 40.5 ± 3.5 years (30-56 years). Patient 3 had undergone surgical closure of a coronary fistula 24 years previ-

ously, and he was brought back to our hospital for the recurrent symptoms of palpitation, shortness of breath, and syncope. The other 5 patients all had symptoms of palpitation, shortness of breath, fatigue, angina, chest pain, and hypertension. Among these 5 patients, patient 1 had acute congestive heart failure. On physical examination of these 5 patients, a grade 3/6 murmur could be heard along the left intercostal space.

Laboratory Studies

Chest radiography demonstrated a very big mass adjacent to the right or left heart border in all patients. The mean cardiothoracic ratio was 0.63 ± 0.03 (0.58-0.72). Echocardiography, ultrafast computed tomography (UFCT), and 3-dimensional aerial imaging

Figure 1. UFCT in patient 1 reveals giant right coronary aneurysm with diameter of 138 mm and coronary fistula draining to left ventricle.

Figure 2. Coronary angiography in patient 5 reveals giant right coronary artery (RCA) aneurysm (CAA) with diameter of 56 mm. Coronary fistula drains into left ventricle (LV) through interventricular septum.

Figure 3. Through incision of giant right coronary aneurysm in patient 1, aneurysm is found to communicate to left ventricle. Fistula, which can be seen at top of suction apparatus, is surrounded by arterial wall of giant coronary artery aneurysm. Diameter of fistula is 22 mm.

revealed a large aneurysm on the right coronary artery (patients 1, 3, and 5), the left main proximal and left anterior descending artery (patient 2), the left anterior descending artery (patient 4), and the diagonal artery (patient 6). The biggest aneurysm, in patient 1, measured $138 \times 121 \times 110$ mm and was 30 mm proximal to its ostium. It drained into left ventricle in the left infraposterior wall. The aneurysm was so big that it compressed the right atrium and right ventricle and narrowed inlet portion of the right ventricle (Figure 1). The smallest aneurysm, in patient 4, measured $22 \times 18 \times 15$ mm and was located in the left anterior descending coronary artery. This patient also had aortic valve insufficiency, but without coronary fistula. The other 5 patients had coronary fistulas, including the patient on whom coronary fistula closure had been done 24 years previously. Interestingly the fistulas in these 5 patients all opened into left ventricle, as demonstrated by coronary arterioangiography in Figure 2. Two fistulas (patients 1 and 5) drained into the left ventricle through the interventricular septum near the cardiac apex. Another 2 fistulas (patients 2 and 6) drained into the left ventricle directly. The patient previously operated on (patient 3) had an enlarged right coronary sinus and proximal of right coronary artery, and a thrombus was located at the bottom of the aneurysm. His distal right coronary artery was closed.

Surgical Procedures

All patients underwent operation, as shown in Table 1, including 1 (patient 1) who underwent an emergency operation for acute congestive heart failure. Access was through a median sternotomy. Routine cardiopulmonary bypass was performed in 4 cases, and femoral arterial cannulation for cardiopulmonary bypass was performed in 2 cases (patients 1 and 5) because of very large aneurysms. Cardiac arrest was obtained by antegrade and retrograde cold blood cardioplegia. The mean time of cardiopulmonary bypass was 144 ± 26 minutes (67-207 minutes). The mean time of aortic clamping was 104 ± 21 minutes (56-172 minutes). In

Figure 4. Postoperative UFCT in patient 1 shows giant right coronary aneurysm to be replaced by saphenous vein graft (yellow conduit). Blue portion represents right atrium and ventricle.

patients 1 and 2, the diameter of the fistulous ostia was more than 10 mm, so the aneurysm was incised and the ostium closed with a patch (4-0 polypropylene suture). In patients 5 and 6, the origin of the fistula was ligated or closed with interrupted sutures under direct vision without injury to the native coronary circulation. Patient 3, with previous coronary fistula closure, underwent coronary artery aneurysm resection, embolectomy, and plastic repair of the aortic wall with partial excision of the free aortic wall. Additional procedures were needed in all 6 cases, as shown in Table 1. Figure 3 illustrates the operative procedure.

Clinical Outcomes

The postoperative courses were uneventful. All patients recovered satisfactorily and were discharged. Mean follow-up was 48 ± 11 months (8-87 months). UFCT and echocardiography were used for follow-up examination and revealed no abnormal communication related to the aneurysm and dilated portion of the coronary artery (Figure 4). During follow-up, all patients were free of symptoms, and none died or had recurrence of giant coronary artery aneurysm.

Discussion

The first pathologic description of a coronary artery aneurysm was by Morgagni⁴ in 1761, and the first clinical case of coronary artery aneurysm was reported by Bourgon⁵ in 1812. Coronary artery aneurysm is an uncommon disease, although it has been diagnosed with increasing frequency since the advent of coronary angiography. Giant coronary artery aneurysm is rarely seen. This is the first report of a

series from a single hospital of 6 patients with giant coronary artery aneurysm.

Incidence

From July 1996 through October 2004, a total of 30,268 patients in our hospital had undergone heart surgery, including 5707 with coronary artery disease, 16,423 with congenital heart disease, and 85 with coronary artery fistula. We found a total of 13 patients with coronary artery aneurysm, among them 6 patients with giant coronary artery aneurysm (diameter exceeding 20 mm). The incidences of coronary artery aneurysm and giant coronary artery aneurysm in our cardiac surgical population were thus only about 0.04% and 0.02%, respectively. However, giant coronary artery aneurysm is found in 5.9% (5/85) of the patients with congenital coronary artery fistula, although the latter is found in only 0.1% of the patients who undergo coronary angiography,⁶ indicating a possible histopathologic connection between giant coronary artery aneurysm and congenital coronary artery fistula.

Etiology

The main causes of coronary artery aneurysm include atherosclerosis, Kawasaki disease, complication of percutaneous transluminal coronary angioplasty, and endocarditis.¹ The etiology of coronary artery aneurysm, however, varies geographically. For example, in Europe or North America, atherosclerosis causes 50% of coronary artery aneurysms, congenital heart disease causes 17%, and Kawasaki disease causes 10%.^{1,2} In Japan, Kawasaki disease is the main cause of coronary artery aneurysm.^{7,8} In China, Kawasaki disease also seems to be responsible for the most cases (7 cases from total of 13 patients with coronary artery aneurysm) in our observation. With regard to the etiology of giant coronary artery aneurysm, however, most cases are congenital, as in this report (5 cases of 6, 83%) and a previous report (7 cases of 17, 41%).³ We therefore conclude that the etiology of coronary artery aneurysm is different from that of giant coronary artery aneurysm; that is, coronary artery aneurysms are mainly due to atherosclerosis in Europe and North America or to Kawasaki disease in China and Japan, whereas giant coronary artery aneurysms are mainly due to congenital malformation.

Location

In our series, giant coronary artery aneurysm was more often seen around the atrium, which is consistent with a previous report.³ This indicates that the area of atrium is a weak point for the formation of giant coronary artery aneurysm.

Complications and Treatments

The complications of coronary artery aneurysm include thrombosis, embolization, rupture, and vasospasm. In cases

with fistula formation of coronary artery aneurysm, complications are congestive heart failure and infectious endocarditis. The natural history and prognosis of giant coronary artery aneurysm remain obscure. With regard to therapy, simple observation may be justifiable for small aneurysms that produce no symptoms. Because of the rarity of giant coronary artery aneurysm, it is difficult to standardize treatment. Conservative treatment consists of attempts to prevent thromboembolic complications with anticoagulant and antiplatelet drugs; however, an operation should be considered for large aneurysms that produce significant symptoms, such as the cases described in this report. Surgical treatment is most widely used to avoid complications, such as extension, thrombosis, rupture, and coronary embolization. Various surgical strategies have been adopted, such as reconstruction, resection, and isolation with concomitant coronary bypass. In cases with fistula, closure of the fistula is needed. When native coronary blood flow is compromised, a bypass graft is indicated.

The distinction of our aneurysms is that most were symptomatic, complicated with coronary artery fistula, and in danger of rupturing if left untreated. Patients with giant coronary artery aneurysm who have significant symptoms should undergo surgery as early as possible, and for the procedure they should be placed under cardiopulmonary bypass with moderate hypothermia for resection of the aneurysm and closure of the fistula. In patients with coronary fistulas and giant aneurysms, the ligation of the original fistula site of coronary artery should be performed carefully without injury to the native coronary circulation in addition to closure of the distal openings of the fistula. Incision of the aneurysm wall made it easier to identify the original fistula site and examine the native coronary artery. If the diameter of fistulous ostium is more than 10 mm, we recommend repairing with a patch. We seldom closed the distal opening of the fistula from the recipient cavity because in our experience this cannot completely prevent recurrence of the fistula.

When a giant coronary artery aneurysm is very large, femoral artery cannulation for cardiopulmonary bypass may be necessary to relieve the compression of ventricle inflow tract for the resection of giant aneurysm. In our series, giant coronary artery aneurysms were often combined with other cardiac disease, such as aortic dilation, aortic valve insufficiency, and thrombus. Additional procedures should be done at the same time for those patients.

Coronary artery fistula can be diagnosed by preoperative echocardiography,⁹ but for the patients with a giant coronary aneurysm it is necessary to do coronary arterioangiography for the diagnosis and surgical procedure. In our study, UFCT and 3-dimensional aerial imaging delineated the size and configuration of coronary aneurysm and revealed their relation to the cardiac chambers. For the follow-up studies,

we have applied UFCT, 3-dimensional aerial imaging, and echocardiography. Results were reliable, and the cost was much less than that of coronary angiography.¹⁰

We thank Nicholas Lamelza from Philadelphia and Chao Yan from Tongren Hospital in Beijing for valuable suggestions and correction of the manuscript.

References

1. Syed M, Lesch M. Coronary artery aneurysm: A review. *Prog Cardiovasc Dis.* 1997;40:77-84.
2. Vranckx P, Pirot L, Benit E. Giant left main coronary artery aneurysm in association with severe atherosclerotic coronary disease. *Cathet Cardiovasc Diagn.* 1997;42:54-7.
3. Mawatari T, Koshino T, Morishita K, Komatsu K, Abe T. Successful surgical treatment of giant coronary artery aneurysm with fistula. *Ann Thorac Surg.* 2000;70:1394-7.
4. Morgagni JB. De sedibus et causis morborum. Vnctus Tom I, Epis 27, Art 28, 1761.
5. Bourgon A. *Biblioth Med.* 1812;37:183, cited by Packard M, Weehsler HF. Aneurysm of the coronary arteries. *Arch Intern Med.* 1929;45: 135-7.
6. Vavuranakis M, Bush CA, Boudoulas H. Coronary artery fistulas in adults: incidence, angiographic characteristics, natural history. *Cathet Cardiovasc Diagn.* 1995;35:116-20.
7. Kato H, Ichinose E, Kawasaki T. Myocardial infarction in Kawasaki disease: clinical analysis of 195 cases. *Pediatr Cardiol.* 1986;108: 923-7.
8. Suzuki A, Kamiya T, Kuwahara N. Coronary artery lesions in Kawasaki disease: cardiac catheterization findings in 1100 cases. *Pediatr Cardiol.* 1986;7:3-9.
9. Chen CC, Hwang B, Husing MC, Chiang BN, Meng, LC, Wang DJ, et al. Recognition of coronary fistula by Doppler two-dimensional echocardiography. *Am J Cardiol.* 1984;53:392-4.
10. Wu QY, Li DY, Hu SS, Pan SW, Qi ST. Surgical treatment of giant coronary artery aneurysm. *Asian Cardiovasc Thorac Ann.* 2001;9: 215-7.