Surgical Treatment of Giant Coronary Artery Aneurysm Secondary to Kawasaki Disease

Chunhu Gu,^{1*} Shanhong Fan,^{3*} Heping Zhou,^{1*} Yunya Wang,^{2*} Dinghua Yi,¹ Rong Zhao,¹ Guocheng Sun¹

¹Department of Cardiovascular Surgery, ²Department of Nuclear Medicine, Xijing Hospital, Fourth Military Medical University, Xi'an, China; ³Department of Equipment and Supplies, Tangdu Hospital, Fourth Military Medical University, Xi'an, China

ABSTRACT

Aim: To investigate the clinical features and surgical management of giant coronary artery aneurysm during end-stage Kawasaki disease.

Methods: From May 2006 to October 2007, 5 patients, 2 to 57 years old, presented with giant coronary artery aneurysm and underwent surgical correction. The coronary aneurysm diameters were 1.5 to 2.5 cm. The coronary aneurysm lesion sites included the right main coronary artery in 1 case, the left main coronary artery in 2 cases, and both the left and right coronary arteries in 2 cases. Preoperative electrocardiogram revealed altered S-T segments in 5 cases and reduced ejection fraction values in 3 cases, resulting in 1 emergency admission for congestive heart failure. Surgical treatments included thromboendarterectomy, thrombectomy, and aneurysmal reconstruction under the orthophoria of extracorporeal circulation.

Results: There were no operative deaths. All patients recovered and received dopamine 2 to 4 μ g/min per kg and nitroglycerine 0.3 to 0.5 μ g/min per kg. Time spent by patients in intensive care was uneventful. Following surgery, 4 patients showed ischemic improvement of the S-T segment on electrocardiograms, and 4 patients presented with increased ejection fraction, according to cardiac ultrasound inspection. The improvement of ejection fraction value was not significant in only 1 case.

Conclusion: Surgery is necessary for stage-3 Kawasaki disease patients that have giant coronary artery aneurysm complications. Surgical treatment includes thromboendarterectomy, thrombus clearing, aneurysmal reconstruction, and coronary artery bypass grafting, followed by postoperative anticoagulation and immunotherapy. Myocardial ischemia and cardiac function can be greatly improved through surgery.

INTRODUCTION

Kawasaki disease is a self-limiting, immunological disease commonly found in infants. The pathological characteristics

*Chunhu Gu, Shanhong Fan, Heping Zhou, and Yunya Wang are co-first authors.

Received April 8, 2009; accepted May 5, 2009.

Correspondence: Guocheng Sun, Department of Cardiovascular Surgery, Xijing Hospital, Fourth Military Medical University, Xi'an, Shanxi, 710032, China; 86-29-84775311; fax: 86-29-83210092 (e-mail: gchtougao@126.com). are primarily made up of systemic vasculitis, especially of the coronary artery [Gong 2002]. Coronary artery aneurysm may occur during end-stage disease, and thromboembolism may give rise to myocardial ischemia, myocardial infarction, and acute cardiac failure, thus surgical treatment is mandatory [Huang 2004; Li 2006]. In the present study, 5 patients with giant coronary artery aneurysms occurring during end-stage Kawasaki disease received successive operations during the period May 2006 to October 2007. The clinical manifestations, pathological characteristics, and surgical indications and methods were investigated.

DATA AND METHODS

Clinical Data

Five patients (3 males and 2 females, age 2-57 years) were enrolled in this study. The patients primarily exhibited symptoms that included chest distress and precordial discomfort (1 patient, age 57 years), and cardiac insufficiency (1 patient, age 2 years, emergency admission). Electrocardiograms displayed an obvious change in S-T segments in all 5 cases, and echocardiography demonstrated an ejection fraction value of 38% to 61%. Right main coronary artery multiple giant coronary aneurysm in combination with thromboembolism was diagnosed in 1 patient, left coronary artery circumflex branch aneurysm with thromboembolism in 2 patients, and left and right coronary artery aneurysm in 2 patients. Coronary arteriography confirmed giant coronary aneurysm, coronary artery stenosis, thromboembolism, and ventricular wall motion disturbance. Of the 5 patients, 4 received medical treatment for >6 months.

Myocardial Preservation

Preoperatively, the patients received 2 µg/min per kg dobutamine along with diuretics to improve heart function. In addition, patients were administered aspirin orally (3 mg daily). During surgery, the heart was arrested with antegrade cardioplegia by cold crystal (2 cases) or cold oxygenated diluted blood (3 cases). The distal coronary artery was directly perfused by cannulation. The mean arterial blood pressure during extracorporeal circulation was maintained between 50 to 70 mmHg. Postoperative cardiac support included intravenous injection of dopamine 2 to 4µg/min per kg and nitroglycerine 0.3 to 0.5 µg/min per kg.

Moreover, the patients received an intravenous drip of fructose or phosphocreatine.

Surgical Treatment and Operative Pathology

Coronary aneurysms were 1.5 to 2.5 cm wide and 1.5 to 5 cm long and were hard, with an absence of fluctuation (Figure, part a). After cardioplegia arrest, the aneurysmal body was incised longitudinally while the incision extended the normal coronary artery to determine the interface between the thickening intima and the fibrous layer. The intima was completely dissected. The intima was obviously thickened by 2 to 3 mm and was characterized by a gray-white color, plaques, and red or mixed thrombus (Figure, part b). Because the mixed thrombus was not fully organized and the intima surface was smooth in 2 cases, only the thrombus was cleared, with normal intima remaining (Figure, parts c and d). The coronary artery at the ends of the aneurysm remained soft and intact and allowed a diameter of 2 to 3 mm (4 cases) (Figure, part e). Four patients underwent coronary angioplasty with a running suture of 6-0-prolene thread (Figure, part f). Among these patients, 1 presented with right main coronary artery multiple giant coronary aneurysm complicated by thromboembolism, which was fully organized and integrated to the vascular intima. The main stem and collateral branch were completely obstructed (>5 cm). The remaining patient underwent right posterior descending branch bypass grafting with the great saphenous vein. The ascending aorta was deaired and opened and then rewarmed to 36.5°C when extracorporeal circulation was stopped.

RESULTS

There were no operative deaths or severe complications, with the exception of ventricular cardiac arrhythmia in 1 case. Chest pain and chest distress were relieved in all patients. Electrocardiogram demonstrated myocardial ischemia, and cardiac 2-dimensional ultrasound and color Doppler outcome revealed that the ejection fraction value and ventricular wall motion function were remarkably improved in 4 cases. In 1 patient, age 57 years, ejection fraction did not obviously improve, possibly due to patient age, poor condition of target vessel, long-term chronic myocardial ischemia, or myocardial revascularization insufficiency. The follow-up time ranged from 2 to 18 months, and the results were satisfactory. No recurrence or reappearance of coronary aneurysm was observed. According to the postoperative pathological reports, the coronary aneurysm wall displayed fibrosis and hyaline degeneration, and the thrombi were accompanied by organization and calcium mineralization.

DISCUSSION

Kawasaki disease is a self-limiting, immunological disease affecting systemic circulation and small vessels, in particular the coronary artery. The etiology remains undetermined [Gong 2002; Huang 2004]. Coronary aneurysms primarily occur during the middle and end stages of disease. The incidence rate accounts for 20% of children with the disease.



A representative surgical treatment and operative pathology. The coronary aneurysm diameter was 1.5 to 2.5 cm, and the length was 1.5 to 5.0 cm. It was large and hard, with an absence of fluctuation (a). The intima was obviously thickened by 2 to 3 mm, and was characterized by a gray-white color, plaques, and red or mixed thrombus (b). The thrombus was cleared with normal intima remaining (c, d). The coronary artery at the ends of the aneurysm remained soft and intact (e). The case patient underwent coronary angioplasty with a running suture of 6-0-prolene thread (f).

Therefore, coronary aneurysm is the main cause of acquired coronary artery disease in infants [lliadis 2002; Kitamura 2002; Reinerth 2006].

The pathological manifestations of coronary aneurysm include intimal fibroplasias of the vascular wall, rupture of the middle fibrous lamina, and bulging of the vascular wall. In our patients, the aneurysmal diameter was many times greater than the naive normal coronary artery and was complicated by thromboembolism. Approximately 5% to 19% of lesions develop into coronary stenosis and give rise to acute myocardial infarction or sudden death [Yoshikawa 2000]. Although in our patients the cardiological symptoms were not observed clinically, other complaints were evident, such as chest distress, precordial discomfort, pericarditis (myocarditis and endocarditis), cardiac arrhythmia, or cardiac insufficiency. In the present study, a 2-year-old patient had an emergency admission because of severe heart failure with an ejection fraction of 38%. Using coronary arteriography, we assessed the coronary aneurysm and developed a surgical strategy. On the basis of the surgical treatment in the 5 patients enrolled in this study, the conclusions were as follows:

Operative Indication

Patients with giant coronary artery aneurysm secondary to end-stage Kawasaki disease underwent treatment. Patients with obvious clinical symptoms or electrocardiogram changes, coronary aneurysm and coronary stenosis, or coronary aneurysm complicated with thromboembolism also underwent treatment. Asymptomatic giant coronary aneurysm posed a potential risk for rupture. Patients with coronary aneurysm with severe multiple coronary disease and cardiac anomalies required surgical treatment for both conditions at the same time.

Surgical Methods

Surgical methods included fibrosis intima and thromboembolism dissection, excision of the aneurysmal wall, and aneurysmal reconstruction. Operative coronary stenosis lesions received scaffold implantation or coronary artery bypass graft. Intimal dissection was suitable for patients with a hard aneurysmal wall, intimal fibrosis with calcification, or thromboembolism complicated by organization. Subsequent to removal of parts of the aneurysmal wall, aneurysmal reconstruction was performed.

Among patients with single-branch multiple coronary artery aneurysm with intima fibrosis, as well as completely organized and obstructed thrombus, coronary artery bypass grafting was recommended [Kitamura 2002].

Among patients with coronary aneurysm that was complicated by distal local coronary stenosis and normal intima, the above-mentioned methods were feasible. Simultaneously, scaffolds were implanted to relieve stenosis and prevent intima breakage. In the present study, 1 patient was accordingly treated and achieved satisfactory short-term effects. No restenosis was observed. In 5 patients with giant coronary aneurysm, intimal fibroplasia and thromboembolisms were evident in the aneurysmal wall, and the thrombus was organized to obstruct vessels. The aneurysmal walls remained hard without potential rupture, characteristics that may differ from changes in the coronary aneurysm wall due to a coronary artery fistula. These findings also provided evidence for the choice of surgical methods.

CONCLUSION

In our study, 4 patients were children. Therefore, intimal and thrombus dissection, as well as coronary angioplasty, were adopted to allow for long-term coronary artery bypass grafting effects and secondary effects. The feasibility for these methods, as well as the long-term outcomes, require further studies.

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