



THE ANNALS OF THORACIC SURGERY



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Ann Thorac Surg 1999;68:1832-1833

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An Adult Case of Bland White Garland Syndrome With Huge Right Coronary Aneurysm

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We report the case of a 62-year-old male patient with left main coronary artery originating from the pulmonary trunk, severe mitral insufficiency, and huge right coronary artery aneurysm. He is the oldest such patient among those reported in the literature, surviving to the sixth decade without any anginal symptoms. He is also the first such case with such a huge and calcified right coronary artery aneurysm and a prominent collateral from the noncoronary circulation.

(Ann Thorac Surg 1999;68:1832-3)

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Bland-White-Garland syndrome is a rare congenital anomaly seen mostly in infancy and childhood. Due to severe disturbance of the normal physiology of the myocardium, survival to adulthood is rare. Associated mitral valve insufficiency of ischemic origin can be seen theoretically. This may disappear after the correction of the coronary pathology.

A 62-year-old man was admitted to our hospital with the complaints of effort dyspnea and palpitation that had progressively increased during the previous 3 years. He was in New York Heart Association class III when admitted to the hospital. He weighted 82 kg and measured 177 cm in height. His blood pressure was 122/78 mm Hg and his pulse rate was 80 bpm regularly. History revealed coronary artery disease in his family. On auscultation, a continuous murmur (Levine 1-2/6) over the precordium and pansystolic murmur (Levine 4/6) more prominent at the apex was heard. Chest roentgenogram films revealed cardiomegaly (cardiothoracic index: 60%) and left atrial enlargement. Electrocardiography displayed poor R progression in leads V₁₋₄ and ST-segment depression in leads V₅₋₆, suggesting left ventricular hypertrophy. No signs of previous myocardial infarction or even ischemia were present.

Two-dimensional echocardiography showed severe mitral insufficiency (grade IV), left atrial enlargement, and left ventricular dilatation (left ventricular end systolic diameter = 51 mm; left ventricular end diastolic diameter = 64 mm). A mass localized in the right atrium was also suspected. Coronary angiography and cardiac catheterization was performed. The left main coronary artery (LMCA) could not be visualized by selective left coronary angiography; however, there was a prominent retrograde collateral blood flow from the right coronary artery (RCA) to left anterior descending (LAD) and

circumflex (Cx) artery. Selective right coronary artery injection showed a huge right coronary artery aneurysm on an early image and a prominent retrograde collateral blood flow from RCA to the LAD and Cx on a delayed image. Although there was no prominent difference in O₂ saturation between inferior vena cava and right ventricle outflow, it was found to be 85% in the main pulmonary artery, showing step-up oxygenation, which suggested steal of the coronary oxygenated blood flow in to the main pulmonary artery. O₂ saturation in the left and right distal pulmonary arteries was found to be 77% and 78%, respectively, and the calculated Qp/Qs was found to be 1.3. Mitral valve and right atrial mass exploration and revascularization of LAD and Cx artery was planned.

After classical median sternotomy and cannulation, extracorporeal circulation was initiated. Perfusate temperature was decreased to 26°C. After cross-clamping of the aorta, retrograde cold crystalloid cardioplegic solution was infused through the aortic root; however, diastolic arrest was not feasible. The main pulmonary artery was dissected and we tried to give the cardioplegic solution from the LMCA ostium. Excessive back-flow was prominent, making this attempt unsuccessful. Because of unsaturated blood flow from the noncoronary collaterals on the cross-clamped heart, retrograde cardioplegia through the coronary sinus was considered as an ineffective alternative approach. Thus, deep systemic and topical hypothermia was used for myocardial protection. It was reassured by transverse aortotomy and pulmonary artery incisions that the LMCA originated from the pulmonary artery next to the posterior cusp. The so-called right atrial mass in echocardiography was a huge RCA aneurysm that extended to the posterior descending coronary artery. It was heavily calcified (Fig 1). Wide collaterals over the right ventricle, connecting RCA and LAD, and aneurysmatically enlarged proximal portions of LAD and Cx, could easily be noticed at the operation. There was an extensive collateral channel over the entire myocardium (Fig 1). Because of a wide collateral at the Waterston's groove, it was only feasible to reach the left atrium by transatrial septal approach. The two leaflets of the mitral valve were seen to be wide apart because of annular dilatation. Excessive calcification, predominantly at the papillary muscles and chordae tendinea and much less at the leaflets, produced marked prolapsus of the leaflets. The valve was replaced with Carbomedics No. 33 valve prosthesis (Sulzer Carbomedics, Austin, TX) and the orifice of the LMCA was closed from inside the pulmonary artery by a Teflon pledgeted polypropylene (Impra Inc, Tempe, AZ). The postoperative course was uneventful; the patient was extubated at the fifth hour and discharged on the seventh day with the prescription of digoxin, Coumadin (Du Pont Pharmaceuticals, Wilmington, DE), and aspirin. To evaluate the origin of the noncoronary collateral flow, we performed the second coronary angiography; however, it was not possible to visualize bronchial arteries. Selective RCA angiography showed that there was no more retrograde filling of the left coronary system from the RCA, suggesting a prominent noncoronary collateral circulation to the left coronary system preventing retrograde filling.

Accepted for publication March 5, 1999.

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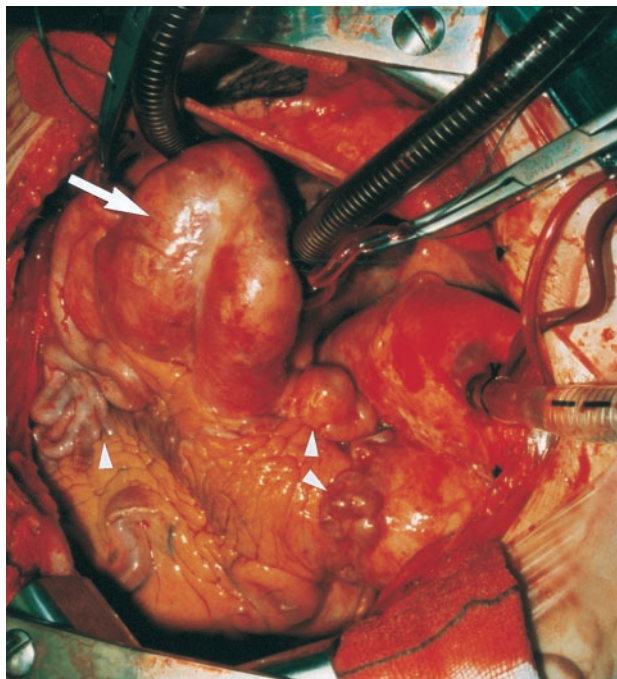


Fig 1. Giant right coronary artery aneurysm. Note the coronary collateral vessels.

Comment

Brooks was the first to demonstrate that coronary arteries might originate from the pulmonary artery [1]. Although this anomaly is reported in only one of 300,000, it is the most frequent anomaly among the origination of the coronary arteries. Because O₂ saturation and pressure are the same at the aorta and pulmonary artery of the fetus, this anomaly does not result in fetal loss. The main problem arises after the first month of life as physiologic pulmonary hypertension regresses. Only 25% of the patients may reach puberty, most of whom experience mitral insufficiency, myocardial infarction, symptoms of angina, and congestive heart failure [2]. In this case, it was not possible to demonstrate ischemia by electrocardiography or clinically. Electrocardiography showed only left ventricular hypertrophy, and the patient had symptoms resulting from left ventricular dilatation and mitral insufficiency, which had become more prominent in the last 3 years (New York Heart Association class III). The more collateral channels exist between RCA and left coronary system, the longer the patient survives; however, extensive collateral circulation may not prevent ischemic damage, because low pulmonary artery pressure results in coronary steal phenomenon, which may cause sudden deaths in about 80% to 90% of the infants who did not have appropriate therapy. Thus, Bland and colleagues have defined a symptom complex in infants with angina, dyspnea, excessive perspiration, dizziness, and paleness induced by crying and feeding, which they named Bland-White-Garland syndrome [3].

Treatment is by operation, and survival of up to 13 years after the closure of the origin of LMCA has been reported in the literature [4]. However, it has also been reported that mortality is higher in patients with simple ligation, than those with aortocoronary bypass. For the

infants, the preferred technique is reconstructing a coronary system of two vessels [5].

Due to the patient's advanced age, well-developed coronary and noncoronary collaterals, and aneurysms of coronary arteries, a simple ligation was performed. If a surgical approach for creating a double-coronary system were performed, it might have resulted in coronary imbalance and thrombosis in the aneurysmatic coronary arteries. In our patient, we did not perform any surgical intervention for the aneurysms, because they were huge and calcified. Also, we thought that such a dilated ventricle would not have tolerated any infarct zone that might appear as a result of the procedure.

In conclusion, the combination of advanced age, ischemic mitral insufficiency without any ischemic symptoms, huge RCA aneurysm, and proximal aneurysmatic dilatation of LAD and Cx made this case an interesting one, which also has enlightened the physiopathology of this syndrome.

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A Review of Aortopulmonary Fistulas in Aortic Dissection

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Aortopulmonary fistula is an exceedingly rare complication of aortic dissection. Only 4 cases in acute dissection and 8 cases in the chronic one have been published previously. We report the thirteenth case and a review of the literature. A man underwent an operation for type A aortic dissection. At surgery, a fistula was discovered between the false lumen and the main pulmonary artery, although the preoperative investigations did not suggest such a complication.

(*Ann Thorac Surg* 1999;68:1833-6)

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Accepted for publication March 18, 1999.

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