

A Rare Late Complication Following Composite Graft Replacement of Aortic Root in A Patient with Marfan Syndrome : A Case Report and Literature Review

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Abstract

Composite graft replacement for aortic aneurysm and dissection more currently is used to prevent the life-threatening cardiovascular complications associated with Marfan syndrome, albeit this procedure carries a potential risk of various late complications. Stenosis in the left coronary ostial anastomosis is a rare late complication following composite graft replacement with separate Dacron coronary grafts. This complication may be a possible cause of late cardiac events such as sudden death and global myocardial infarction. We present a case with Marfan syndrome who developed tight stenosis in the left main coronary ostial anastomosis seven months following composite graft replacement with separate coronary graft. Percutaneous transluminal coronary angioplasty with stent implantation was performed to clear the stenosis. The experience showed that the stent supported left main coronary angioplasty may be considered an option to be used as a bridge of surgical revascularization. There is a need to assess the long-term outcomes of Marfan 掇 patients treated with stenting for unprotected left main coronary stenosis. (J Intern Med Taiwan 2003;14: 77-82)

Key Words : Composite graft replacement, Left main coronary stenosis, Percutaneous transluminal coronary angioplasty, Coronary stenting

Introduction

Marfan syndrome is an autosomal dominant di-sorder of connective tissue characterized by abnormalities involving the skeletal, ocular and cardiovascular systems 1. The syndrome is one of the commonest inherited disorders of the connective tissue, with a prevalence of about 1 in 10,000. The phenotypic

manifestation are due to the production of abnormal fibrillin, which is the main component of extracellular microfibrils and is encoded by the FBN1 gene on chromosome 15q21.2. Abnormal microfibrils cause degeneration of the elastic laminae and subsequently lead to aortic dilatation. The cardiovascular features typically include aortic dissection or rupture. In addition, the dilatation of the aortic ring can cause aortic valve incompetence and regurgitation.

The primary life-threatening complication of Marfan syndrome is rupture of aortic aneurysm or dissection, which can be prevented by composite graft replacement, a procedure first described by Bentall and De Bono in 1968. The procedure has also become the standard treatment of aortic regurgitation associated with ascending aortic aneurysm. The Cabrol operation is an alternative, using a small Dacron tube (8 mm in diameter) to reimplant the coronary arteries. This procedure can be used to avoid excessive tension on coronary anastomoses and prevent late pseudoaneurysm. The Cabrol operation was subsequently modified by Piechler and Pluth. Separate coronary grafts were used for coronary reattachment.

Post-operatively, the common late complications (occurring more than one month after operation) such as dissection or rupture of the residual aorta or thromboembolic events are cumbersome problems. Among the complications, stenosis in the left main coronary ostial anastomosis is a rare late complication following composite graft replacement association with Cabrol modification. This complication may cause late cardiac events such as sudden death or global myocardial infarction and thus needs careful management. We presented a woman who suffered from chest pain seven months following composite graft replacement association with Cabrol modification. Complication of post-operative stenosis in the left main ostial anastomosis was noted by coronary angiography. We further discussed possible mechanisms and management of this complication.

Case Report

A 39-year-old woman with Marfan syndrome had ascending aortic aneurysm and chronic type A aortic root dissection. She underwent the Bentall operation with hemi-Cabrol modification at Taipei Mackay Memorial Hospital in March 2002. Preoperative coronary artery angiography was unremarkable. After operation, she was followed up regularly and took warfarin daily. She suffered from crushing chest pain in the substernal area with radiation to the back since 2:00 AM on October 7, 2002. The duration of chest pain was about twenty minutes. Dizziness and sweating were also noted. She was immediately sent to the Emergency Department of Taitung Mackay Memorial Hospital, where the electrocardiographic examination revealed ST segment depression and T wave invert in V2-V6, I, and aVL (Fig 1). Elevated levels

of cardiac enzymes (CK=140U/L, CKMB=12.1U/L, and positive Troponin I) and increased prothrombin time international normalized ratio (PTINR) (2.92) along with clinical presentation suggested an unstable angina. After admission, echocardiogram revealed regional wall motion abnormality with mild hypokinesis in the anterior septal area of left ventricle and normal ejection fraction. Other laboratory data were as follows: cholesterol =174 mg/dL, triglyceride =83 mg/dL and glucose (AC) = 93 mg/dL. After placing on oral aspirin and intravenous nitroglycerin, her chest pain was improved. Although there were no risk factors for coronary artery disease, coronary artery occlusion was highly suspected. The patient was transferred to Taipei Mackay Memorial Hospital where coronary artery angiography revealed 80% stenosis in the anastomosis of aortic graft and the left main coronary artery (Fig 2). The right coronary artery was patent (Fig 3). Percutaneous transluminal coronary angioplasty (PTCA) was performed and an Express stent (4.5×12 mm) was implanted (Fig 4). Subsequently, the patient recovered with no chest discomfort. She took aspirin and warfarin on a daily basis and there is no chest discomfort for recent five months.

Discussion

In order to improve the overall outcome of Marfan syndrome, early prophylactic treatments for cardiovascular complications are important. Although the beta blockers might lower the risk and slow the progression of aortic dissection by reducing the systolic ejection impulse⁹, current thoughts propose that the dissection can be surgically intervened as a prophylactic means. Groenink and colleagues¹⁰ provided the following guidelines for prophylactic aortic root replacement: (1) aortic root diameter ≥ 55 mm, (2) positive family history of aortic dissection and aortic root diameter ≥ 50 mm, and (3) aortic root growth ≥ 2 mm/year. Selective Marfan 掇 patients meeting the above criteria are at low operative risk. However, it still carries a potential risk of various late postoperative events. Common late complications (occurring more than one month after operation) include rupture/dissection of the residual aorta, thromboembolic events (stroke or prosthetic aortic valve thrombosis) and prosthetic valve endocarditis⁷. Vincent L. et al⁷ reported aortic root replacement in 271 Marfan patients of their 24-year experience. Two hundred thirty-five Marfan patients underwent elective aortic root replacement with no 30-day mortality. Two early deaths occurred among 36 patients who underwent urgent or emergent operation. Eighty-three percent of patients in this series are currently alive. The actuarial freedom from thromboembolism, endocarditis, and reoperation on the residual aorta 20 years postoperatively was 93%, 90% and 74%, respectively.

Cabrol operation can prevent late pseudoaneurysm in coronary anastomoses following

Bentall operation. Cabrol and colleagues reported that the long-term results of composite graft replacement with small coronary grafts were satisfactory 5. However, other authors reported the occurrence of left main anastomotic stenosis following the Cabrol operation, in which urgent coronary artery bypass was required 8. The present case is unique in who received stent supported left main coronary angioplasty to clear her left main anastomotic stenosis. To our knowledge, this approach in such a clinical setting has not been reported in the literature.

The two possible mechanisms of stenosis in the left main coronary ostial anastomosis following the modified Cabrol operation are proposed as follows. Firstly, iatrogenic small intimal tear of coronary ostium may have been caused by retrograde cannula of cardioplagia during operation. This small intimal tear could have triggered chronic inflammatory infiltration and activated platelet proliferation subsequently inducing platelet-rich thrombus formation gradually 11. Secondly, the size difference between the 8-mm Dacron coronary grafts and native coronary arteries might have caused turbulent flow at the coronary orifice. The ostial intimal hyperplasia resulting from turbulent flow might be the reason for late anastomotic stenosis of coronary graft 12. In fact, it is not only the turbulent flow that may cause the injury at the anastomotic site. This coronary graft is not compliant (elastic) in their cross-sectional area. An additional factor to be considered is that the coronary arteries are embedded in the surrounding tissues, and the main coronary flow is supplied during the diastolic phase. The cross-sectional area of the arteries is almost ideally down-sized in the length. The arteries can thus oppose the systolic hydrodynamic pressure peak without injury. In contrast, the small (8mm) fabric graft has no support surrounding it except the original support of the coronary artery just distal from the main ostial button 13. The systolic pressure peak, "hammering" against the button, may increase the movement of the dissected artery 13. A question arises with regard to how such an injury may be prevented to avoid obstruction of left main coronary anastomoses. Vincent 13 suggested use of a graft as short as possible and, immobilization of the anastomoses as much as possible. The turbulent flow may be caused by the anastomotic discongruency, from 8 mm down to 3 mm in diameter, for example. Vincent further suggested tapering the anastomosis by oblique end-to-end anastomosis, partially sacrificing the traditional button to preserve some laminar flow.

PTCA is usually performed in degenerative vascular disease of coronary arteries. PTCA for atheromatous left main coronary stenosis has been reported but the intervention is associated with suboptimal short-term and long-term results because of immediate elastic recoil and high incidence of restenosis and complications 14. The use of unprotected left main PTCA is recommended only in carefully selected (inoperable) patients. Stents seem to improve unprotected left main PTCAs by

effectively dealing with flow obstructing dissection and reducing restenosis 15. The long-term prognosis of patients after stenting of unprotected left main coronary stenosis was reported to be favorable in selected patients with normal left ventricular function 16. Recently, the stent supported left main coronary angioplasty has been used in the management of complication after arterial switch operation in patients with simple transposition of the great arteries 17. It is also an alternative treatment for left main coronary stenosis in Takayasu 癩 arteritis which increases the risk of failure of surgical revascularization during active disease 18. For the present case, we have satisfactorily improved the left main coronary stenosis after the modified Cabrol operation by stent supported left main coronary angioplasty which procedure may be considered an option to be used as a bridge to surgical revascularization.

According to Dake and colleagues 19, stenting has also been reported to be feasible in the treatment of selected patients with type B aortic dissection. For selected patients with acute type A or B aortic dissection, stent-graft placements have become a treatment option 19. The stent-grafts are made of self-expanding stainless-steel covered with woven polyester or polytetrafluoroethylene material. However, this new treatment for selected patients with aortic dissection requires further evaluation 19.

Repeated postoperative studies with CT scans and earlier surgical intervention before the onset of aortic dissection are mandatory for patients with Marfan syndrome 20. Chest discomfort in patients with Marfan syndrome following the Bentall operation with Cabrol modification must be carefully monitored in consideration of various postoperative events including coronary artery stenosis. Recent advances in transesophageal echocardiography provide direct visualization and assessment of the left main coronary artery 21. Satoshi et al 12 recommended that this noninvasive method be added to routine CT scan or magnetic resonance imaging for follow-up after composite graft replacement with Cabrol modification to facilitate the diagnosis and timely management of coronary stenosis.

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Marfan 症候群病人經主動脈重建術後罕見之晚期併發症：病例報告及文獻回顧

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摘 要

Marfan 症候群病人常因主動脈剝離或破裂而危及生命。雖然目前已能經由人工血管或合併瓣膜置換術的使用，使死亡率大幅降低，晚期術後併發症的危險仍潛在。不管手術如何進步，仍有相當比例之病人，因人工血管接縫至遠端大動脈處發生病變，而再次接受手術治療。從文獻統計得知，最常見的晚期併發症包括殘餘主動脈剝離或破裂、出血性或梗塞性腦中風、主動脈瓣膜栓塞，或人工瓣膜性心內膜炎。然而，左冠狀動脈主幹接縫狹窄，是相當罕見之晚期併發症，也是造成猝死和心肌梗塞之重要原因。我們報導一位女性 Marfan 病人，經主動脈重建術後七個月，左冠狀動脈主幹至人工血管接縫處發生顯著性狹窄。經由氣球擴張術合併血管支架植入，已能有效治療此一併發症，但我們仍需長期追蹤及進一步的大型研究來評估此療法的長期成效。

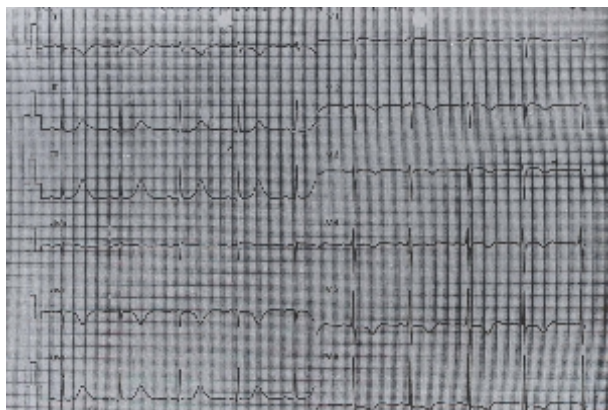


Fig.1. Electrocardiography showing ST segment depression and T wave invert in V2-V6, I1, and aVL during chest pain attack.

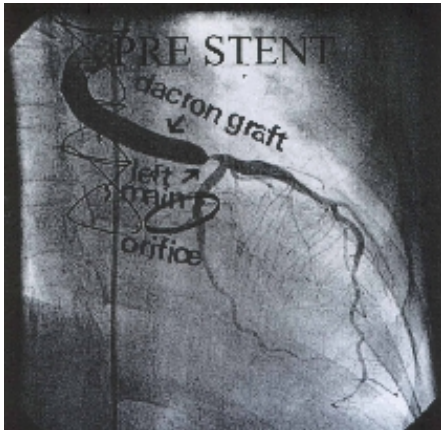


Fig.2. Significant stenosis in the anastomosis of the Dacron graft and the left main coronary artery following composite graft replacement with Cabrol modifications.

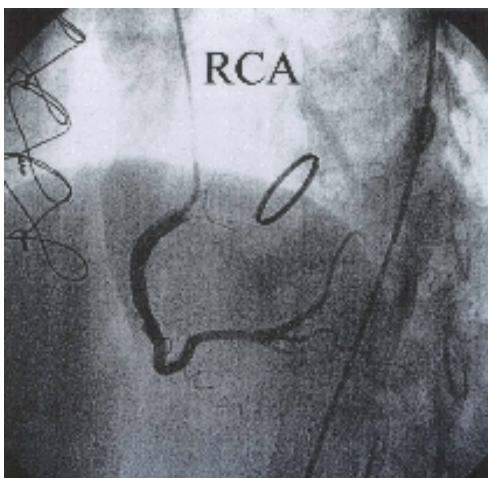


Fig.3. Patent right coronary artery (RCA).

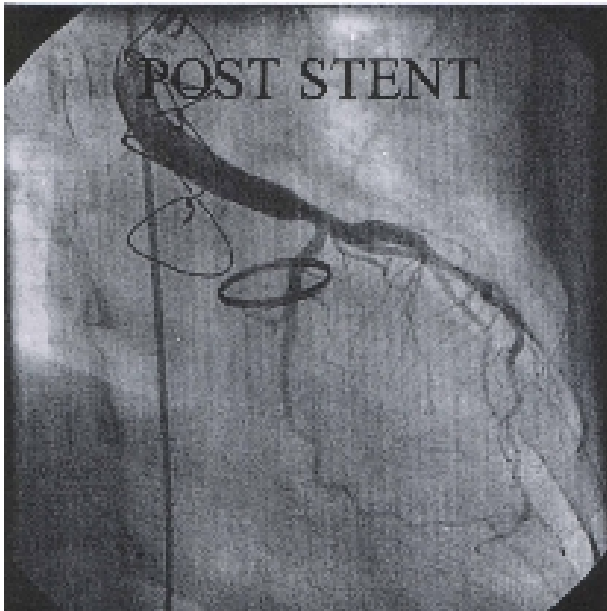


Fig.4. No residual stenosis in the left coronary ostium after an Express stent implantation.