

Cardiac Reoperation in a Patient with Osteogenesis Imperfecta: A Case Report

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A case of a 40-year-old man with dehiscence of the prosthetic aortic valve and recurrence of mycotic aneurysm of the left ventricular outflow tract with osteogenesis imperfecta is presented. He had an operation of aortic valve replacement and direct closure of the mycotic aneurysm for infective endocarditis twenty-one months ago. We performed reoperation of prosthetic aortic valve, patch closure of the mycotic aneurysm and graft replacement of the ascending aorta. He was complicated with multiple fractures of the bilateral scapula and dislocation of the left shoulder on the first postoperative day. Fortunately, cardiac reoperation was performed successfully in this patient despite anticipated difficulties with tissue friability with osteogenesis imperfecta. (Ann Thorac Cardiovasc Surg 2001; 7: 241-5)

Key words: osteogenesis imperfecta, cardiac reoperation, aortic valve replacement, mycotic aneurysm, infective endocarditis

Introduction

Osteogenesis imperfecta (OI) is a heritable disorder of connective tissue. Cardiovascular operation in cases with OI have been associated with a high mortality rate.¹⁻⁵ We are reporting a patient with OI that underwent reoperation of prosthetic aortic valve, patch closure of the mycotic aneurysm of the left ventricular outflow tract and graft replacement of the ascending aorta.

Case

A man with osteogenesis imperfecta (OI) was referred to our hospital for cardiac evaluation when he was 38 years old in July 1998. He had laparoscopic cholecystectomy for acute cholecystitis five weeks before, and was complicated with pneumonia, sepsis and cerebral hemorrhage. He was a man of short stature with blue sclerae,

and a childhood history of atraumatic bone fracture. Hearing and dentition were normal. He had familial history in that his father, three brothers and a sister have OI traits. His past history was negative for rheumatic fever. Auscultation revealed a grade 4/6 diastolic decrescendo murmur at the left sternal border.

He had a low grade fever of 37.5°C and general fatigue. Blood tests showed a white blood cell count of 8900/mm³, C-reactive protein of 5.3 mg/dl and erythrocyte sedimentation of 43 mm/h. Although gram-positive micrococcus was detected in blood cultures in the previous hospital, bacterial blood tests in our hospital were negative. Platelet counts, coagulation studies and serum chemistry analysis were normal. Chest X-ray showed mild cardiomegaly. The cardio thoracic ratio was 0.57. An electrocardiogram revealed left ventricular hypertrophy with secondary ST-T changes. Transthoracic echocardiography demonstrated severe aortic regurgitation (AR) with a bicuspid valve. The aortic annulus was not dilated. The mitral and tricuspid valves appeared normal. Catheterization results and aortography confirmed these findings. He was diagnosed with severe AR due to infective endocarditis with OI.

First operation

After antibiotic therapy (Vancomycin HCl 2 g/day) of six weeks, cardiac operation was performed under usual

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Fig. 1. Transesophageal echocardiogram four weeks after the first operation revealed recurrence of mycotic aneurysm of the left ventricular outflow tract in the mitral-aortic intervalvular fibrosa.



Fig. 2. Aortogram in 30° right anterior oblique projection 20 months after the first operation demonstrating severe aortic regurgitation.

cardiopulmonary bypass. The bileaflet aortic valves were thin and were perforated without vegetation. An aortic annulus showed almost normal findings. After the aortic valve was excised, a mycotic aneurysm (MA) of the left ventricular outflow tract was found in the mitral-aortic intervalvular fibrosa. Because the MA was inactive and the size of its ostium was 8 mm in diameter, it was closed directly by 4 horizontal pledget supported mattress sutures. The aortic valve was replaced with 23-mm Carbo-medics aortic valve prosthesis (Sulzer Carbomedics). All sutures used for valve replacement were reinforced with Teflon felt pledgets. The ascending aorta was carefully closed with 4-0 polypropylene running suture. Hemostasis was achieved during surgery without hemorrhagic complication. The sternum was approximated with usual stainless steel wires.

The patient had an uneventful postoperative recovery. Antibiotic therapy (Vancomycin HCl 2 g/day) was continued four weeks postoperatively. Although he had no symptoms and no inflammatory reaction, the recurrence

of MA of the left ventricular outflow tract without any paravalvular leakage was found by transesophageal echocardiography four weeks after the operation (Fig. 1). Since the patient had no symptoms and paravalvular leakage was not found, he was discharged six weeks after the operation.

He came to our institution reporting chest discomfort 20 months after the first operation. The echocardiography demonstrated paravalvular leakage of the aortic prosthesis at the orifice of the MA. Aortography confirmed severe aortic regurgitation (Fig. 2). The patient underwent cardiac reoperation 21 months after the first operation.

Second operation

Since the right femoral artery was assumed to be very thin due to its venous appearance, the right external iliac artery was used for cardiopulmonary bypass. On opening the ascending aorta, dehiscence of the prosthesis was found at the orifice of the MA. After the prosthesis was resected, recurrent MA of the left ventricular outflow tract

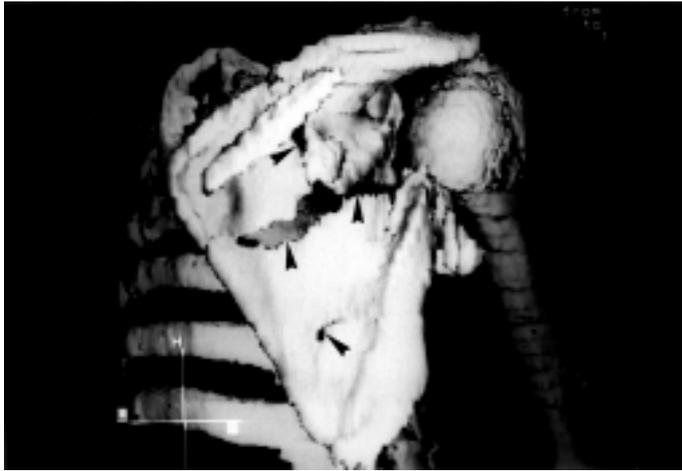


Fig. 3. Computed tomography of the right shoulder 3 days after the second operation revealed multiple fractures of the right scapula (arrows).

was found. Patch closure of the MA was done with two layers of glutaraldehyde-preserved horse pericardium. Re-aortic valve replacement was performed with a 21-mm Carbomedics aortic valve prosthesis. The valve prosthesis was then sutured to the patch and to the remaining native aortic anulus. When the ascending aorta was closed, the aortic wall was torn for its friability. We performed graft replacement of the ascending aorta with collagen-impregnated, woven Dacron graft without any undue tension to the aortic wall. Although 4 units of blood, 20 units of fresh frozen plasma and 20 units of platelet were used intraoperatively, hemostasis could be achieved without critical hemorrhagic complication. The sternum was approximated as the usual way.

General convulsions occurred complicated when he woke up on the bed on the first postoperative day. Although his consciousness was normal after the convulsion, his bilateral shoulder could not be moved. The shoulder roentgenograms showed multiple fractures of bilateral scapula and dislocation of the left shoulder. Computed tomography of the shoulder confirmed these findings (Fig. 3). Fortunately the complication of cardiac, hemorrhagic and wound healing were not found postoperatively. Postoperative transesophageal echocardiography and aortography revealed no evidence of MA of the left ventricular outflow tract and aortic regurgitation (Figs. 4, 5). He is doing well five months after the second operation.

Pathology

Pathologic examination of the aortic valve in the first operation showed slight myxomatous degeneration of the leaflets and inflammation of mainly neutrophilic leukocytes suggestive of endocarditis. Microscopic examina-

tion of the aortic wall in the second operation showed focally myxomatous degeneration without reduction of the elastic fibers.

Discussion

Osteogenesis imperfecta (OI) is a heritable disorder of connective tissue, transmitted in an autosomal dominant fashion.^{3,6} Patients with OI may have a blue sclerae, triangular head, deafness, numerous spontaneous fractures, skeletal deformities, and a positive family history of blue sclera.^{2,4,5,7} This patient was diagnosed type 1 of OI since he was a man of short stature with blue sclerae, a childhood history of atraumatic bone fracture, and familial history of traits of OI.

OI is categorized in a group of heritable disorders of connective tissue that includes the Ehlers-Danlos syndrome, the Marfan syndrome, the Hurler syndrome and pseudoxanthoma elasticum.^{3,8} Although cardiovascular manifestations in OI are virtually similar to those in the Marfan syndrome, they are rarer than in the Marfan syndrome.⁷⁻⁹ The true prevalence of cardiovascular involvement in OI is not known.

There are 36 reported cases of cardiovascular operation with OI previously. There were 33 male and 3 female patients, ranging in age from 19 to 63 years with a mean of 41.8 years. Nine (25.0%) of 36 reported cases died after operation. Cardiovascular operations in OI have been associated with high morbidity and mortality rates. It appears from all the reported cases that the mortality after cardiovascular operations in patients with OI is mainly due to friability of the tissue and bleeding secondary to platelet dysfunction and capillary fragility.¹⁻⁵ An attempt at surgical correction of severe AR was first



Fig. 4. Transesophageal echocardiogram 4 weeks after the second operation revealed that the mycotic aneurysm of the left ventricular outflow tract had disappeared.

reported in 1965 by Criscitiello and colleagues.¹⁰ Siggers described a patient with OI and AR who survived valve replacement in 1971.¹¹

Fortunately, cardiac operations were successfully performed twice in this patient despite the anticipated difficulties with tissue friability. There are individual differences of tissue friability and bleeding tendency.^{8,12} Moriyama and colleagues stated that the nature of OI is still ill-defined and variable in individuals.⁴ Type I collagen is composed of three polypeptide chains, two alpha-1 chains and one alpha-2 chain, intertwined into a triple helix. The mildest form of OI (type I) appears to result from underproduction of type I collagen due to reduced alpha-1 messenger RNA.³ The severity of the clinical disease is proportional to the quality of the mutation and to the extent of abnormal type I collagen.^{3,13} It is impossible to know tissue friability preoperatively.¹² Since there are cases of abnormal bleeding without abnormality in the preoperative coagulation test,^{3,10,14,15} there are necessary preparations of platelet and fresh frozen plasma intraoperatively.¹²

Dehiscence of the prosthesis with a subsequent paravalvular leak is possible given the weakness of the connective tissue.^{3,6,16} We considered that the cause of the dehiscence of the prosthesis in this patient was the suture dehiscence of the orifice of the mycotic aneurysm (MA) of the left ventricular outflow tract. We used direct closure of the MA because the orifice of the MA was small and there was not any tension. Symbas and colleagues¹⁷ stated that direct closure of MA should be used when the gap is small and the suturing does not result in any undue tension. However, Kamata and

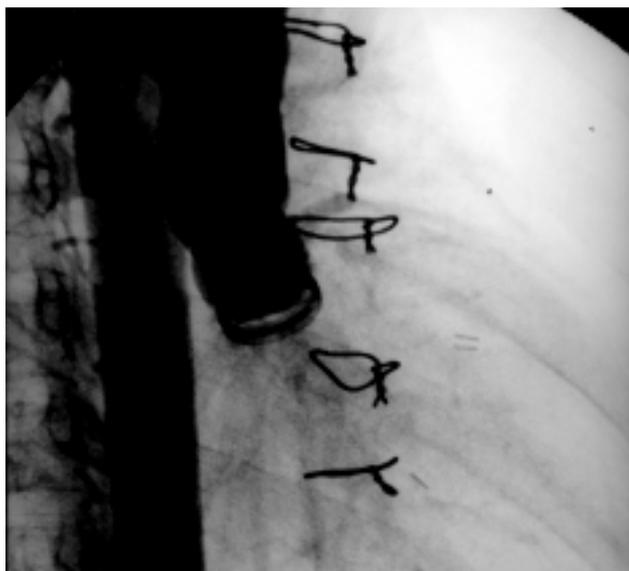


Fig. 5. Aortogram in 30° right anterior oblique projection 6 weeks after the second operation demonstrating disappearance of aortic regurgitation.

colleagues¹⁸ recommended patch closure of the MA so as to avoid recurrence of aneurysm formation even if its orifice proved to be small. Although the recurrence of MA was probably related to tissue friability to some extent, that may be avoided if the procedure of patch closure of the MA was used.

Since the patient had no symptoms and paravalvular leakage was not found, he was discharged six weeks after the first operation. However the patient had chest discomfort and paravalvular leak 20 months after the first operation, and then we performed reoperation. There are no reports of survival cases of reoperation for valvular dysfunction and paravalvular leak. Wong³ stated that valve replacement should be offered to the symptomatic patient with OI. We also recommended that cardiac operation should be made to the symptomatic patient with OI. Although the patient is doing well five months after the second operation, regular follow up for re-dehiscence of the prosthesis should be planned.

Conclusion

A case of a 40-year-old man with dehiscence of the prosthetic aortic valve and recurrence of mycotic aneurysm of the left ventricular outflow tract with osteogenesis imperfecta is presented. Cardiac reoperation was performed successfully in this patient despite anticipated difficulties with tissue friability with osteogenesis imperfecta.

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