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Case Report

Modified Bentall procedure with mitral valve replacement in a patient with Marfan syndrome: a case report

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ABSTRACT

Marfan syndrome (MFS) is an autosomal dominant connective tissue disorder characterized by fibrillin-1 protein abnormalities, predisposing individuals to cardiovascular complications such as aortic root dilation and mitral valve prolapse (MVP). Management often requires surgical intervention, with the modified Bentall procedure being a standard approach for aortic root replacement. However, managing concomitant mitral valve pathology poses additional challenges due to the complex anatomical relationship between the two valves and the prolonged nature of the procedure. We present a case report of a 35-year-old male with Marfan syndrome who underwent a successful simultaneous modified Bentall procedure and mitral valve replacement for concurrent aortic root aneurysm and mitral valve prolapse. Surgical interventions involved meticulous attention to detail to mitigate risks associated with malpositioned coronary ostial sutures, debridement of the severely calcified mitral valve, and achieving hemostasis. Following surgery, the patient exhibited favorable postoperative outcomes, with evidence of optimal valve function and improved systolic and diastolic parameters, indicative of a successful recovery.

Keywords: Marfan syndrome, Bentall procedure, Mitral valve replacement, Aortic root aneurysm

INTRODUCTION

Marfan syndrome (MFS) is a hereditary connective tissue disorder characterized by mutations in the FBN-1 gene, leading to abnormalities in fibrillin-1 protein synthesis.[1] This results in structural defects in connective tissue, particularly in the cardiovascular system, predisposing individuals to aortic aneurysm formation, aortic regurgitation, aortic dissection, and mitral valve prolapse.[2]

Historically, untreated Marfan syndrome (MFS) had a dire prognosis, with individuals facing a life expectancy of 40–50 years during the 1970s, primarily due to complications such as aortic dissection or heart failure. However, significant advancements in medical and surgical treatments have led to substantial improvements, with the average life expectancy of those with Marfan syndrome approaching nearly 70 years.[3] The Bentall procedure, now established as a standard approach for aortic root replacement, has played a pivotal role in achieving these advancements.[4]

While the primary focus of surgical intervention in MFS has traditionally been on the aortic root, the coexistence of mitral valve pathology poses additional challenges. Mitral valve involvement, although common in MFS, is frequently overlooked during aortic root surgery. However, addressing mitral valve disease concurrently with aortic root replacement is crucial due to the intricate anatomical relationship between the two valves and the

potential difficulties associated with subsequent mitral valve interventions post-aortic surgery.

With limited literature available regarding the management of both aortic root aneurysm and mitral valve prolapse concurrently, the significance of addressing this clinical challenge cannot be ignored. Here we present a case outlining the management of mitral valve prolapse with concomitant aortic root aneurysm in a Marfan syndrome patient. He successfully underwent a modified Bentall procedure with concurrent mitral valve replacement.

CASE REPORT

A 35-year-old male presented with a two-year history of exertional dyspnea and palpitation. On admission, his blood pressure was measured at 120/55 mmHg. He had a collapsing pulse and a heart rate of 92 beats per minute. Additionally, his respiratory rate was 22 breaths per minute. Characteristic features associated with Marfan syndrome were observed, including tall stature, arachnodactyly (long, slender fingers), flat feet, and joint hypermobility (Figure 1). Precordial examination revealed a mid-systolic click, followed by late systolic murmur over the apex, and auscultation of the lungs revealed bilateral basal crepitations. His abdomen was found to be non-tender and distended, with shifting dullness.

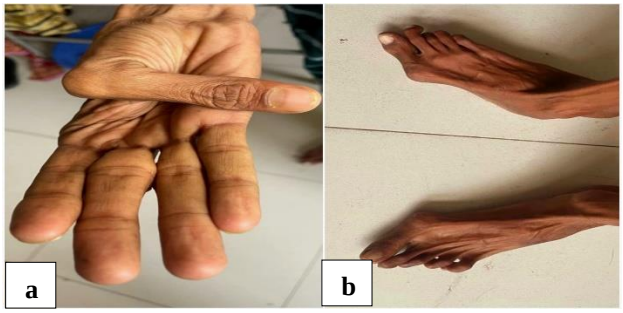

Figure 1: Arachnodactyly (long, slender fingers), joint hypermobility, and flat foot.

Color Doppler echocardiography revealed significant cardiovascular abnormalities, including a grossly dilated aortic root with dissection, severe aortic regurgitation, and concurrent mitral valve prolapse with severe mitral regurgitation. Computed tomography (CT) aortogram (Figure 2) confirmed tricuspid aortic valve with aortic valvular and ascending aortic aneurysmal dilatation. Measurements included an aortic annulus of 39 mm, valvular area of 64.4 mm, sinotubular junction of 73.6 mm, root of the aorta at 66.2 mm, and ascending aorta measuring 30 mm. Abdominal ultrasound showed features suggestive of hepatic congestion and moderate ascites.

Consequently, the patient was diagnosed with Marfan syndrome complicated by aortic root aneurysm, severe aortic regurgitation, and severe mitral regurgitation with features of heart failure. Following the diagnosis, the patient was managed conservatively to address the heart failure, and preparations were made for surgery.

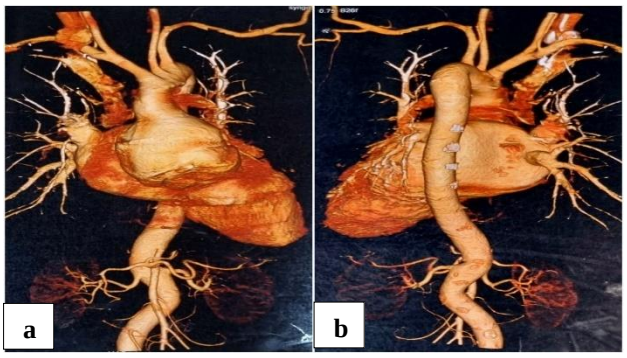

Figure 2: CT aortogram with arch of the aorta and branches showing aortic valvular and ascending aortic aneurysmal dilatation.

During the surgical intervention, the patient's aortic root and ascending aorta were observed to be grossly dilated with dissection at the sinus of the aorta (Figure 3). After ensuring hemodynamic stability following induction of general anesthesia, a median sternotomy was performed. Arterial cannulation was achieved at the arch of the aorta, while venous cannulation was accomplished using a bicaval cannulation strategy. Cardiopulmonary bypass was initiated upon reaching an activated clotting time of >480 seconds, with hypothermia at 28°C maintained throughout the procedure. The heart was subjected to fibrillation using local ice cooling, followed by cross-clamping of the aorta near the origin of the innominate artery. Del Nido cardioplegic solution was then administered antegradely into the coronary ostia for cardioprotection, with repeat doses infused every 80 minutes.

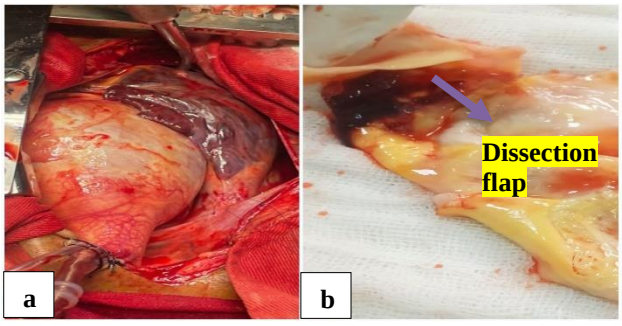

Figure 3: (a) Aortic root and ascending aorta grossly dilated, and (b) dissection at the sinus of aorta.

The ascending aorta was transected approximately 2 cm above the coronary ostia, while distally, it was transected just above the aneurysmal sac. The left and right coronary arteries were carefully dissected free from the surrounding tissue in preparation for the creation of right and left coronary artery buttons, which would be later implanted. A left atriotomy was performed to excise the severely calcified mitral valve, followed by the implantation of a 31/33mm On-X bileaflet mechanical valve (Figure 4).

Subsequently, the left atriotomy was closed. The aortic valve leaflets were excised, and the annulus was debrided in the presence of calcification. After sizing the aortic annulus, a composite valve-graft comprising a 23 mm On-X bileaflet mechanical valve and a Dacron tube graft (30 mm in size) was utilized to replace the diseased portion of the aorta. The diameter of the Dacron tube graft was matched to the outer diameter of the mechanical valve's sewing cuff and secured with interrupted 4-0 propylene sutures. Following this, 2-0 pledgeted polyester horizontal mattress sutures were employed to affix the Dacron tube graft to the aortic valve annulus. Subsequently, coronary ostia were created within the tube graft, with round openings slightly larger than the coronary ostia's diameter. Coronary arteries were then re-implanted to the graft end-to-side using continuous 6-0 polypropylene sutures (Figure 5). Pericardial felt strips were used to reinforce the anastomoses and ensure hemostasis. Caution was exercised during coronary artery implantation to avoid ostia dissection or tension. Finally, the distal end of the conduit was connected to the distal ascending aorta using continuous 4-0 polypropylene sutures (Figure 6). Deairing was performed, and the patient was gradually taken off cardiopulmonary bypass once adequate hemostasis was confirmed.

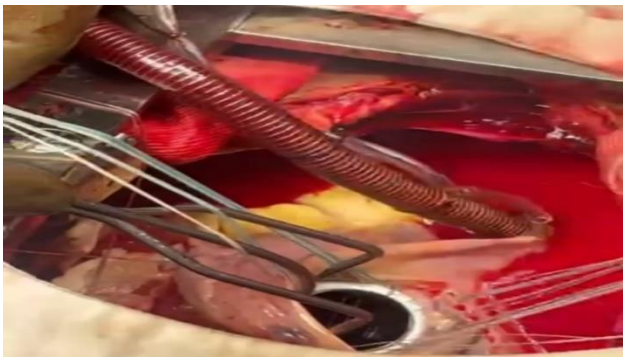

Figure 4: Implantation of 31/33 mm on-X bi-leaflet mechanical mitral valve.

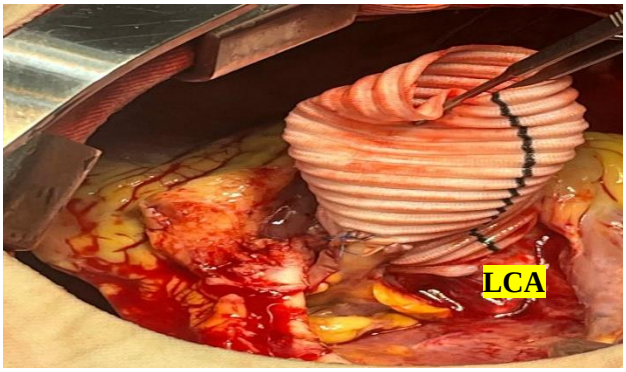

Figure 5: Left coronary artery (LCA) sutured to the Dacron tube graft in an end-to-side manner using polypropylene sutures.

Postoperatively, the patient recovered well without any immediate complications (Figure 7). Follow-up echocardiography showed satisfactory valve function and mild left and right ventricular systolic dysfunction. Serum creatinine levels remained stable at 1.3 mg/dl.

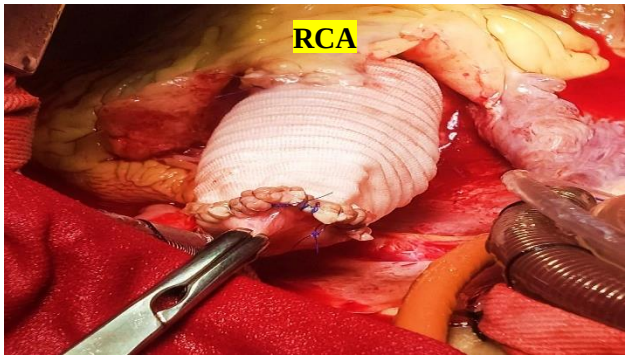

Figure 6: Distal end of the Dacron tube graft anastomosed to the distal ascending aorta with right coronary artery (RCA) re-implantation.

Figure 7: Postoperative follow-up of the patient.

DISCUSSION

Around 1 in 5,000 individuals are affected by Marfan syndrome, characterized by multi-systemic involvement, including skeletal, ocular, and cardiovascular manifestations.[5] Cardiovascular abnormalities, notably aortic root dilatation, and mitral valve prolapse, significantly contribute to morbidity and mortality in affected individuals.[6]

In our case, the patient exhibited classic features of Marfan syndrome, such as tall stature, arachnodactyly, and flat foot alongside notable cardiovascular anomalies including aortic root dilatation, aortic dissection, severe aortic regurgitation, and severe mitral regurgitation.[7,8] The diagnosis was confirmed through echocardiography and CT imaging, which highlighted the characteristic findings.

Surgical intervention remains a cornerstone in the management of Marfan syndrome-related cardiovascular complications, aiming to prevent life-threatening events such as aortic dissection and rupture.[1,9,10] In this case, the patient presented with aortic dissection, necessitating

prompt surgical intervention. However, optimizing the patient's cardiac condition before surgery proved to be exceptionally challenging due to the severity of heart failure. Intensive efforts, including the use of diuretics, were required to stabilize the patient's cardiac function and correct the heart failure. The procedure involved a modified Bentall procedure with mechanical prosthetic valve replacement for both the aortic and mitral valves. This decision was influenced by factors including patient age, durability of mechanical valves, and the necessity for long-term anticoagulation therapy.[11]

During the surgical procedure, meticulous attention was paid to minimize risks related to malpositioned coronary ostial sutures, which could lead to serious complications like myocardial infarction or cardiac dysfunction. Techniques such as partial mobilization of both ostia to prevent postoperative kinking and the use of colored markers for accurate suture orientation were employed to ensure precise surgical execution.[11] To address anastomotic bleeding, careful anastomosis techniques were utilized using polypropylene sutures and ensuring proper alignment of coronary ostia with Dacron tube graft. Additionally, the distal anastomosis was performed using 4-0 polypropylene sutures, with pericardial felt strips reinforcing the suture line. Furthermore, fibrin glue was applied to seal potential sources of oozing along the suture line.

Patients diagnosed with Marfan syndrome and mitral regurgitation exhibit improved survival rates with repair rather than replacement of the mitral valve. Studies indicate that the survival outcomes and risk of reoperation for Marfan syndrome patients are comparable to those with myxomatous mitral disease.[12] However, in this case, severe calcification rendered mitral valve repair unfeasible.

Regular follow-up is crucial to monitor valve function, detect signs of prosthetic valve dysfunction, and effectively manage anticoagulation therapy. Long-term surveillance is necessary to identify potential complications related to aortic grafts and endocarditis. Genetic counselling plays a vital role in guiding inheritance patterns, family planning, and the implications of genetic testing for both patients and their families.

Our patient's favorable outcome aligns with the findings of previous studies, which have indicated that utilizing a combined surgical approach to manage aortic root and mitral valve pathology carries a low risk of mortality, thus highlighting the efficacy of this approach.[12,13]

CONCLUSION

In conclusion, our case highlights the successful utilization of simultaneous modified Bentall procedure and mitral valve replacement for managing Marfan syndrome-related cardiovascular complexities. This approach signifies a significant advancement in treatment. Given the limited number of similar case reports, our contribution adds valuable insights, warranting further research to refine and validate this combined procedure, ultimately improving patient outcomes.

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