Mitral Annular Plication and Valve Replacement in a Patient with Marfan's Syndrome: A Case Report

MARFAN SENDROMLU BİR HASTADA MİTRAL ANÜLER PLİKASYON VE MİTRAL KAPAK REPLASMANI: OLGU SUNUMU

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Summary

Objective: Surgical treatment of mitral regurgitation associated with Marfan's syndrome is controversial because of the underlying degenerative process. The purpose of this study was to determine the surgical outcome of a patient with Marfan's syndrome who had had progressively symptomatic mitral regurgitation and annular dilatation.

Case Report: A 31- year old man who had Marfan's syndrome and who had had progressively symptomatic mitral regurgitation was admitted to our clinic. Cardiac catheterization revealed a heavily calcified mitral annular dilatation with severe mitral regurgitation, fair left ventricular function with an ejektion fraction of 45%. We performed mitral anuular plication and also valve replacement under standard cardiopulmonary bypass technique.

Conclusion: Patiend did well postoperatively and was discharged on the 8th. postoperative day. After the operation he continued to do well on digoxin , warfarin and metoprolol tartarat. We suggest that annular plication and valve replacement might be recommended for a better left ventricular function in patients who have severe mitral annular dilatation and calcification unsuitable for repair and if the replacement is only choice.

Key Words: Marfan's syndrome, Mitral annular dilatation, Valve replacement

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Özet

Amaç: Marfan sendromundaki mitral yetmezliğinin cerrahi tedavisi altta yatan dejenaratif hastalıktan dolayı tartışmalıdır. Bu çalışmada ileri derecede miitral anüler dilatasyonu ve semptomatik mitral yetmezliği bulunan marfan sendromlu bir hastada uygulanan cerrahi teknik ve sonucunun araştırılması amaçlanmıştır.

Olgu Sunumu: İleri derecede mitral yetmezliği bulunan marfan sendromlu 31 yaşındaki erkek bir hasta kliniğimize yatırıldı. Kardiyak kataterizasyonda şiddetli mitral anüler kalsifikasyon ve dilatasyonu ile mitral yetmezliği, enjeksiyon fraksiyonu %45 olan kötü sol ventrikül tespit edildi. Standart kardiyopulmoner bypass tekniği altında mitral anüler plikasyon ve mitral kapak replasmanı yaptık.

Sonuç: Hastanın postoperatif seyri iyi geçti ve 8. günde taburcu edildi. Hastanın operasyon sonrası genel durumundaki iyileşme devam etti ve digoxin ,warfarin ve metoprolol başlandı. Kapak replasmanın tek seçenek olduğu ve tamirin mümkün olmadığı ileri derecede mitral anüler dilatasyuonu ve kalsifikasyonu olan olgularda iyi bir sol ventrikül fonksiyonu için mitral anüler plikasyon ve kapak replasmanını öneriyoruz.

Anahtar Kelimeler: Marfan sendromu, Mitral anüler dilatasyon, Kapak replasmanı

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Marfan's syndrome is an autosomal dominant heritable disorder of connective tissue involving the skeletal, ocular and cardiovascular systems caused by alteration in the synthesis of fibrillin (1). Patients with Marfan's syndrome have a shortened life expectancy because of cardiovascular complications of the disease (1,2). Mitral valve dysfunction is present in 80 % of patients with Marfan's syndrome (1,3) and it is the most common cause of morbidity and mortality in infants and children with Marfan's syndrome (4). Recent data demonstrate that surgical intervention significantly prolongs survival in adults with Marfan's syndrome (2). We present herein the surgical intervention of a 31-year old male patient who has a severe mitral annular dilatation and calcification and a fourth degree mitral insufficiency.

Case Report

31-year old male patient with Marfan's syndrome was admitted for the complaints of palpitation and dyspnea. His father had had also Marfan's syndrome and died suddenly. The patient was Class 4 in New York Heart Association. On echocardiography, 4th degree mitral insufficiency was found with the left ventricular enddiastolic diameter as 7.7 cm and the endsystolic diameter as 6 cm. The left atrial diameter was 8.5 cm. Pulmonary arterial pressure was calculated as 86 mmHg. The ejection fraction was 45 %. The aortic root was 3.5 cm. On cardiac catheterization, mitral regurgitation with severe mitral annular dilatation and calcification, left ventricular dilatation, minimal aortic insufficiency with the aortic root as 3.4 cm were found. No pathology was found with coronary angiography. Besides, on lumbal computerized tomography dural ectasia was found in distal lumbal and sacral regions. The eye examination showed ectopia lentis. He also had pectus carinatum.

At surgery, median sternotomy, standard cardiopulmonary bypass, mild degree hypothermia (28° C), antegrade isothermic blood cardioplegia were used. In the operation, he was found to have a huge left atrium and left ventricle, and the mitral annulus was approximately 9 cm in diameter. Both of the mitral leaflets and the annulus were severely calcificated. For this reason, it was not possible for mitral repair and mitral valve replacement was preferred. However, the mitral annulus was so wide that 33 mm valve was small for it. For the appropriation of the annulus to the valve, we did annular plication towards posterior at the level of both commisures and mitral valve was replaced with 33-mm St. Jude bileaflet mitral valve (Figure-1 and 2). The aortic cross-clamp time was 78 minutes and total bypass time was 98 minutes. The patient was weaned from cardiopulmonary bypass without difficulty. The patient did well postoperatively and discharged on the 8th day with digoxin, warfarin and metprolol tartarat. He was class 2 in New York Heart Association 2 months after the operation. Left ventricular enddiastolic diameter was 65 mm, endsystolic diameter was 52 mm, and left atrium was 64 mm echocardiographically. Pulmonary arterial pressure was calculated as 56 mmHg, aortic root was 36 mm and the ejection fraction was 49 % (Tablo 1).

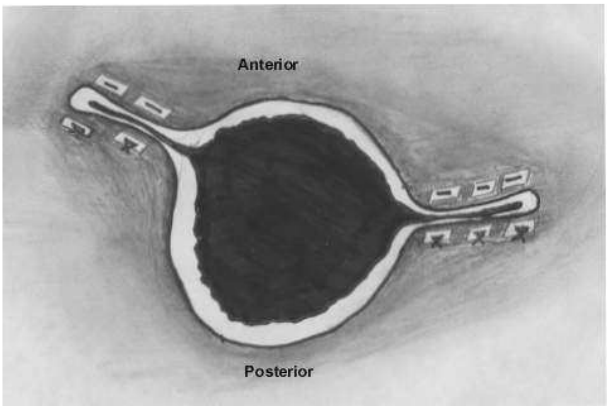

Figure 1. Leaflets have been removed and commisures plicated.

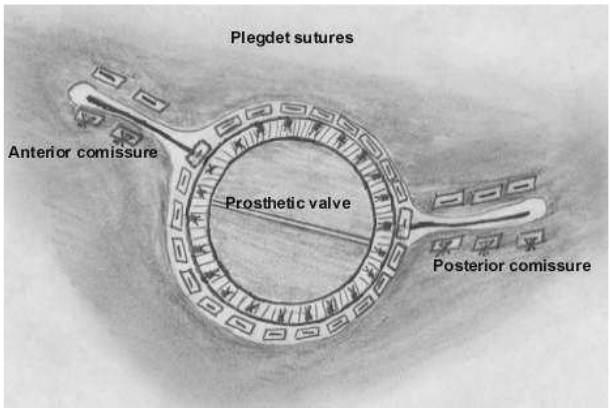

Figure 2. Prosthetic valve have been accommodated into narrowed annulus.

Comment

Marfan's syndrome is the most common heritable disorder of connective tissue. If untreated, cardiovascular manifestations of the Marfan's syndrome cause death in one half of patients during the first four decades of life (2,3). Cardiovascular complications including dilatation of the ascending aorta, aortic dissection, aortic regurgitation, and mitral valve dysfunction determine life expectancy in patients with Marfan's syndrome. Pathologic

Tablo 1. Preoperative and postoperative echocardiographic parameters

	Preoperative	Postoperative
Left ventricle enddiastolic diameter	77 mm	65 mm
Left ventricle endsystolic diameter	60 mm	52 mm
Left atrial diameter	85 mm	64 mm
Pulmonary artery pressure	86 mmHg	56 mmHg
Ejection fraction	%45	%49

involvement of the mitral valve is frequently seen in patients with Marfan's syndrome. Mitral valve dysfunction often precedes aortic involvement and progresses in childhood, and 1 patient out of 8 will develope moderate or severe mitral regurgitation and about 10 percent of patients will have calcification of the mitral annulus (5).

There is a consensus regarding the surgical management of the aortic lesions in Marfan's syndrome, but controversy continues to exist about the surgical management of the mitral valve dysfunction (6). The indications and the technique of operation vary according to the presence of lesions (7). Although successful series of valve repair in cases having Marfan's syndrome and mitral regurgitation were reported recently, repair cannot be done for the cases having severe annulus and leaflet calcifications (8). In this case, annulus and leaflet calcification was present which made the repair impossible. The mitral annulus was so wide that 33mm valve small for it. For this reason, we made annular plication to the anterior and posterior commisures to make the annulus smaller. We brought the annulus to 33 mm valve length and made valve replacement. The patient got better in the postoperative period and his complaints were minimal as he was seen on several controls. Our case is interesting as he had a rarely seen wide annulus with calcification. We suggest that annular plication and valve replacement might be recommended for a better left ventricular function in patients who have severe mitral annular dilatation and calcification unsuitable for repair and if the replacement is only choice.

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