bypass grafting (CABG) required left ventricular assist device (LVAD) implantation due to deterioration of hemodynamic state. The patient was later transported to specialized center with LVAD support and eventually underwent successful heart transplantation (10). Three patients presented clinically with AMI (2, 9, 10) and two others had history or evidence of peripheral artery thrombosis; one of the common iliac artery and the other of the carotid artery (4, 7). Surprisingly none of the cases have encountered any problems regarding hemorrhage postoperatively. One would expect that the deleterious effects of CPB would aggravate the inherent tendencies of bleeding in ET. Only one of these patients required blood transfusions for hematoma of the groin after a coronary angiogram (7).

The limited surgical experience with ET demonstrates that this entity carries a high rate of morbidity and mortality mainly due to thromboembolic complications despite the meticulous efforts to monitor and manipulate platelet numbers and function.

Reports of successful surgery stress the importance of a combination therapy with cytoreductive agents and antiaggregants to prevent thromboembolic events (6) There is, however a delicate balance of hemorrhage versus thromboembolism to be considered in the case of surgical interventions. Platelet functions are very unpredictable in ET causing hemorrhage and thrombosis in the same patient during the same time frame. Cardiopulmonary bypass causes severe reductions in platelet numbers and function (aggregation) alongside depletion of coagulation factors rendering the patient prone to postoperative bleeding. Theoretically the combination of CPB and an ET patient with sub-normal platelet functions is a potential hematological disaster. Keeping this in mind, we decided to utilize the off-pump beating heart method for revascularization. It is clear in this case that in an attempt to avoid the possible hemorrhagic complications of ET and CPB we have run straight into another devastating (thrombotic) complication. Despite our efforts to normalize platelet numbers preoperatively with Anagrelide and maintain an acceptable level of anticoagulation perioperatively the patient suffered a massive thromboembolic episode. Although an autopsy was not available we postulate that this thromboembolic episode must have happened almost instantaneously because at no stage were there any indications of a problem, such as deteriorating hemodynamics, change in ECG or consciousness.

The lack of response to all resuscitation efforts and the development of cardiac standstill in a matter of minutes alongside convulsions during cardiac arrest suggests a massive thrombosis of not only the bypass vessel but probably all the coronary arteries and major cerebral vessels.

A number of studies have demonstrated that platelet function tests can predict 'major adverse cardiac events' in cardiovascular disease but none of these assays have yet been sufficiently studied in large clinical trials to become part of standard clinical evaluation . Further research is necessary to understand the unpredictable nature of platelet function in patients with ET undergoing major surgical procedures.

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Two-stage successful surgery in an aortic coarctation case operated initially for ascending aortic aneurysm

Öncelikli olarak assandan aort anevrizmasına müdahale edilen aort koarktasyonlu bir olguda iki evreli başarılı cerrahi tedavi

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Introduction

Aortic coarctation is a serious pathology required surgical treatment. About 50% of uncorrected isolated aortic coarctation cases are lost up to the age of 10, only 10% may reach the age of 50 (1). The most common reason for death from untreated aortic coarctation is the aneurysm or rupture of aorta or side branches with a rate of 23% (1). Aortic insufficiency resulting from annuloaortic ectasia and ascending aortic aneurysm together with aortic coarctation rarely occur, and surgical treatment is difficult. It is very important to decide whether surgical operation will be of one and two stage, and to determine intraoperative strategy.

Aortic coarctation is a congenital vessel disease that can cause such complications as myocardial infarction, congestive cardiac failure,

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infective endocarditis, aortic aneurysm, aortic dissection or rupture and intracranial bleeding as a result of present resistant hypertension in adult age (2).

The aneurysm of the ascending aorta or dissecting aneurysm is a life-threatening complication of aortic coarctation. The incidence of aortic dissection and aneurysm in patients with coarctation of the aorta is well known (3); however, the reported cases of surgical correction of this condition are very few (3, 4).

Studies report various operations done when aortic aneurysm (1, 2, 4) or aortic dissection (5) occur together with aortic coarctation. These combined conditions are usually treated by one or two-stage surgery when aortic dissection exists, and the first step involves emergency repair of the aortic dissection. Subsequently, coarctation repair is undertaken at a later date. But if there is ascending aortic aneurysm in addition to aortic coarctation without aortic dissection, the first repair must be performed for aortic coarctation. In our patient, initially the pathology of ascending aorta was operated because of dissection suspicion although aortic aneurysm, and aortic coarctation were repaired later.

Case report

A 33-year-old man was admitted to our unit for dizziness and sudden onset of retrosternal, constrictive pain that suggested an acute cardiac problem. On clinical examination, a 3/6 systolic murmur was heard in the aortic area. He have not been diagnosed cardiac disease, reporting only a recent diagnosis of hypertension.

The patient was conscious, pale, sweaty, restless, and agitated. His jugular veins were distended; the blood pressure in his upper extremities was 220/100 mmHg. The carotid and radial pulses existed and were symmetrical, whereas femoral pulses were absent. Chest radiography, performed at the bedside, demonstrated middle mediastinal widening, and evidence of bilateral rib notching, and electrocardiography showed sinus bradycardia with left ventricular hypertrophy. Transthoracic and transesophageal echocardiography demonstrated minimal presence of fluid in the pericardial cavity, dilated ascending and aortic root (approximately 7 cm), advanced aortic valvular incompetence, and signs suspicious for intimal flap in the ascending aorta. However, this image in echocardiography also revealed artifact image. Computerized tomography (CT) of the chest revealed an aneurysm of the ascending aorta, and a coarctation of the descending aorta (Fig. 1). The interscapular and chest pain increased after one hour after arrival to hospital. Because ascending aorta was very large, and although the image in echocardiography resembled artifact, there was a probability of dissection and accordingly a higher risk of rupture, and the patient's clinical condition deteriorated so dramatically that he was taken immediately to the operating room. After median sternotomy, the pericardium was opened. It was stretched because of very large ascending aorta and small amount of fluid. The aneurysmal ascending aorta was thin but not discolored. The femoral and axillary artery cannulations were made to protect spinal and cerebral regions. During operation, perfusion pressure related with femoral and axillary cannulation was monitorized and the pressure was kept above 50 mmHg. Venous drainage was established with a single "two -stage" venous cannula placed into the right atrium, and cardiopulmonary bypass (CPB) was routinely instituted. The aneurysm was limited to the ascending aorta. The ascending aorta was cross-clamped and incised transversely. There was no dissection in ascending aorta. The sinuses of Valsalva were abnormal, they were suspended, and the coronary ostia were displaced. The aortic valve was bicuspid, with partially fused, thickened, and calcified leaflets. The aortic valve was excised, and the segment of the ascending aorta comprising the aneurysmal part was resected. The patient was performed modified Bentall procedure as described by Yakut et al (6). The anastomoses were reinforced with gelatin resorcinol glue and Teflon strips. At the end of the procedure the patient was easily weaned off CPB; CPB, and cross-clamp times were 217 and 173 min, respectively.

The patient was extubated the next morning. Intensive care unit stay was 3 days. No cerebral, respiratory or renal complications occurred. The remaining postoperative stay was also uneventful. In postoperative period, because the clinical findings and preoperative CT were highly suggestive of a coarctation of the thoracic aorta, 3-dimensional and multislice CT was made at the thoracal region. This confirmed the presence of the thoracal coarctation, which revealed a severe coarctation of the aorta, just distal to the take-off of the left subclavian artery (Fig. 2, Video 1. See corresponding video/movie images at www.anakarder.com). The patient was discharged on the 15th postoperative day with anti-hypertensive medications.

A month after the repair of the aneurysm, the patient underwent elective correction of the aortic coarctation. A left posterolateral

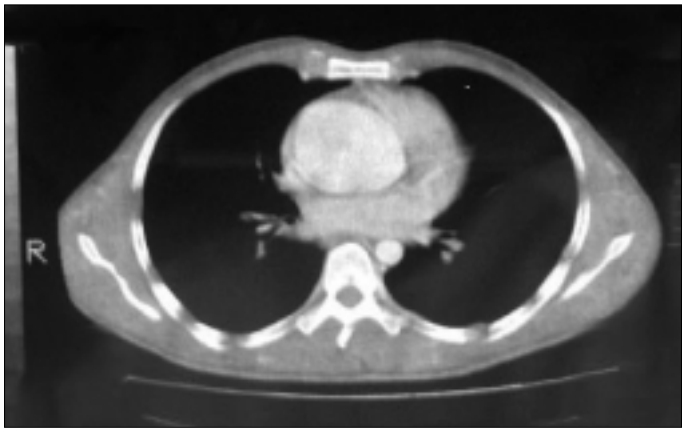

Figure 1. Preoperative computed tomographic scan shows aneurysm of the ascending aorta and coarctation of the descending aorta

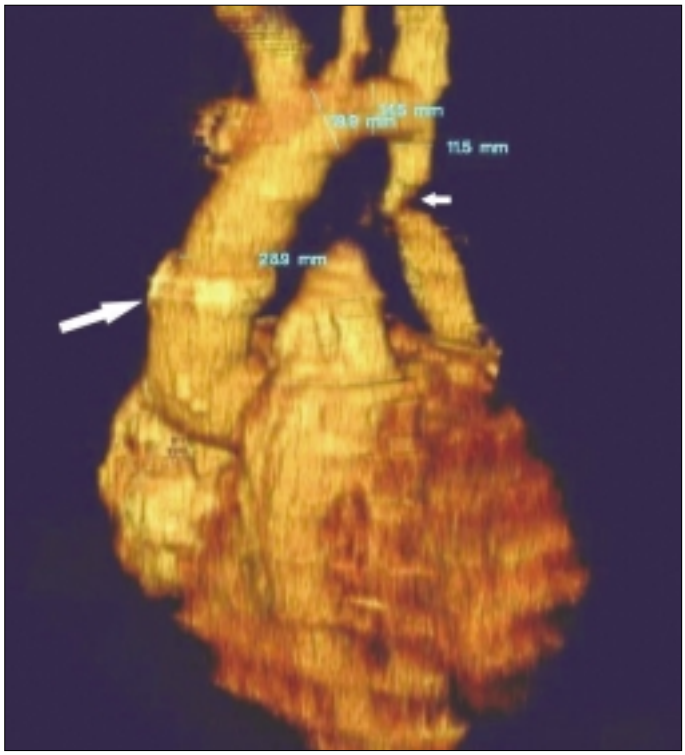

Figure 2. Multislice computed tomography shows aortic coarctation just the below the left subclavian artery after ascending aorta replacement in sagittal cross-section view

thoracotomy was carried out at the fourth intercostal space, and we observed marked dilatation of the descending aorta. Cross-clamps were then placed proximally and distally to the coarctation. Due to the fact, that in descending aorta perfusion pressure was measured 60 mmHg, we did not need atrio-femoral shunt for spinal protection. The narrow isthmic segment was entirely resected and the patchplasty was performed between the distal aortic arch and the thoracic aorta with piece Dacron graft with a continuous 5/0 prolene suture.

The patient was discharged on the 7th postoperative day. On routine follow-up, the patient was in excellent condition. He returned to full-time work. On the 2nd month of follow-up, he was free of complications and two-dimensional echocardiography revealed normal heart contractility, good function of the aortic valvular prosthesis, a regular ascending aorta, and no evidence of aneurysm.

Discussion

Vascular complications and arterial hypertension are major risk factors in the evolution of disease connected with coarctation of the aorta. The miscellaneous studies pointed out that aortic rupture was the cause of death in 19% to 23% of patients with coarctation of the aorta; in 75% of the instances, rupture take place in the ascending aorta (2).

In patients with ascending aortic aneurysm together with aortic coarctation, initially aortic coarctation must be repaired in one or two-stage procedure, in order to lessen proximal hypertension, decrease the chances of progressive dissection or rupture, and enable safe perfusion during correction of the aortic aneurysm (3). Surgical repair may be performed through two different incisions by thoracotomy and sternotomy in cases treated with surgery in one stage (4) or coarctation treatment may be carried out with extra-anatomical bypass following ascending aortic aneurysm repair by sternotomy only in one stage (1). In two-stage surgery, first coarctation is repaired by thoracotomy and then ascending aortic aneurysm is repaired at a later stage (2).

In cases having aortic coarctation and aortic dissection together, ascending aortic dissection is repaired and coarctation is repaired by extra-anatomical bypass in one stage or first dissection and then coarctation is repaired in two stages (3, 5). In our case, both ascending aortic aneurysm and aortic dissection suspicion occurred besides aortic coarctation. We could not treat coarctation initially because of dissection suspicion and rupture risk resulting from ascending aortic aneurysm (almost 7 cm). For this reason, ascending aortic pathology was corrected initially by sternotomy urgently.

We thought that it would not be appropriate to perform extra-anatomical bypass following aortic pathology repair in one stage and coarctation repair by thoracotomy carrying out a second incision in the same step. Because, in one-session surgery, to reach coarctated segment is difficult due to collateral circulation in case to which only sternotomy is performed. Besides, bleedings related to wide and profuse collaterals prevent reaching surgical area (2). Moreover, in the same step for coarctation treatment, surgical operation and anesthesia period increases through a second thoracotomy, infection tendency and thus surgical risk increases. That is, no attempt was performed to correct the aneurysm of the ascending aorta and the coarctation on the same occasion, by means of a compounded approach, because we considered it would extremely increase the operative time and risk, a view indicated by others in a similar situation (7). However, extra-anatomical procedures, frequently can be used for the interrupted aortic arch, recoarctation and in combined procedures such as with valve and coronary artery surgery (8). For this reason, we did not prefer extra-anatomical bypass procedure that we thought to be a time-consuming and palliative procedure.

The aim of repair of cases with aortic coarctation is to allow proximal blood flow to pass distally without obstruction. This can be achieved by either widening the narrowed region or by creating an alternate path for blood flow. Surgical or endovascular techniques can be used as an alternative treatment. End-to-end anastomosis, prosthetic interposition tube grafts, subclavian flap repair, and extra-anatomical corrections (9) can be preferred for surgical treatment. We preferred to perform patchplasty to this patient of old age. In addition to the surgical procedures, there are procedures other than surgical treatment. Balloon angioplasty, endovascular repair and hybrid procedures (10) can be used as alternative treatments. Developments in endovascular technology over the past decade may potentially reduce the morbidity from open surgical repair. Endovascular devices are deployed through peripheral arteries and fluoroscopy is used to confirm accurate placement. Complex anatomical variations may require endovascular devices to be deployed directly into the aorta. Both surgeon and interventional radiologist must work closely to ensure accurate placement (9). However, such procedures were not preferred for our patient because of high cost and lack of endovascular procedures in our region. Generally, surgery still has an important role in the management of the adult patient with coarctation despite the recent advances of endovascular technology. Clinical trials for these patients are difficult to carry out because of the small patient number and the variety of complicating factors. The multidisciplinary team can optimize the management of these patients and combined procedures may further improve results.

The topic of how to replace the aortic valve in the presence of coarctation was not difficult to solve, because the valve was grossly abnormal: it was therefore excised and replaced with a mechanical prosthesis. Such surgical procedures as, classical and modified Bentall applications, separated surgical treatment, David and Ross procedures can be applied in cases of ascending aortic aneurysm and root replacement could be applied. We performed a Bentall replacement of the ascending aorta and aortic valve with a composite prosthesis as described by Yakut et al. (6), because the aortic root was large, the sinuses of Valsalva were enlarged, and the coronary ostia were displaced in our patient. The flanged composite graft offers excellent long-term results, with very low prevalence of prosthetic-related complications. Because the new created sinuses and the flange are especially helpful to continue physiologic function of the aortic root (6), we preferred Bentall procedure with flanged technique.

Selecting the place of arterial cannulation under these circumstances, to insure sufficient blood flow to both the upper and inferior body parts, was a matter of concern. We decided that femoral cannulation alone would not ensure adequate cerebral perfusion. We cannulated the femoral and the right axillary arteries separately, in order to perfuse cerebral and spinal regions simultaneously due to coarctation. In additional, the adequacy of the perfusion of the lower body was evaluated by continuous monitoring of urine output, which was satisfactory throughout the perfusion. Postoperatively, the patient showed evidence neither of impaired renal function nor of ischemic injury to the spinal chord. We presume that the satisfactory perfusion of the aorta distal to the coarctation was the result of a well-developed collateral circulation and because of axillary and femoral perfusion during procedure.

Conclusion

Generally, initially distal pathology related to aortic coarctation should be corrected to eliminate cardiac load in cases of aortic coarctation together with ascending aortic aneurysm. However, as in our case, because of the suspicion of dissection and rupture risk due to a very large ascending aorta, first ascending aortic aneurysm with proximally lesion was operated to treat life-threatening pathology that was thought to be urgent. A second surgical operation was not carried out in the same incision in order not to increase anesthesia period, to decrease surgical risk and thus to eliminate malperfusion risk, and

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coarctation treatment was left to a later period electively. Extra-anatomical bypass was not performed in the same step considering the correction of coarctation with patchplasty would be appropriate and curative, without facing any clinical problems. Besides, although antegrade perfusion with axillary cannulation may be enough in the presence of a well-developed collateral circulation, more appropriate perfusion to the upper and lower parts of the body is presumably best accomplished by separate antegrade and retrograde cannulation.

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