Preductal Coarctation of the Aorta in Adulthood: Case Report

Erişkin Yaşta Görülen Preduktal Tip Aort Koarktasyonu

Mert DUMANTEPE, MD,[a]
Koray AK, MD,[a]
Ufuk MUNGAN, MD,[a]
Murat UĞUR, MD,[a]
Bilal Kaan İNAN, MD,[a]
Ahmet Turan YILMAZ, MD[a]

[a]Department of Cardiovascular Surgery, GATA Haydarpasa Education Hospital, İstanbul

Geliş Tarihi/*Received:* 20.08.2008
Kabul Tarihi/*Accepted:* 28.10.2008

Yazışma Adresi/*Correspondence:*
Mert DUMANTEPE, MD
GATA Haydarpasa Education Hospital, Department of Cardiovascular Surgery, İstanbul
TÜRKİYE/TURKEY
mdumantepe@gmail.com

ABSTRACT Coarctation of the aorta (CoA) is a congenital narrowing of the thoracic aorta that usually occurs just before or after the insertion of the ductus arteriosus remnant. CoA is a rare congenital pathology and accounts for approximately %6-8 of all live births with congenital heart disease. This lesion, which is seen primarily in the infantile (preductal) form, is the 8th most common congenital heart defect but rarely present in an adult. The management of such cases are still a great challenge to the surgeon. Herein, we reported a 30 year-old man with preductal type CoA, who was treated with patch aortoplasty, and discussed current surgical treatment strategies of this rare pathology.

Key Words: Aortic coarctation, cardiac surgical procedures

ÖZET Aort koarktasyonu (AK) torasik aortun sıklıkla duktus arteriosus kalıntısının yerleştiği yerinin hemen öncesinde ya da sonrasında meydana gelen doğumsal darlığıdır. Doğumsal kalp hastalığına sahip tüm canlı doğumların yaklaşık % 6-8 ini oluşturan nadir bir doğumsal patolojidir. AK nun (preduktal) formu 8. en sık doğumsal kalp anomalisi olup bu lezyona erişkinlerde nadiren rastlanır. Böyle bir olgunun tedavisi cerrahlar için hala büyük zorluklar içermektedir. Bu vakada patch aortoplasti tekniği ile tedavi edilen preduktal tip AK nuna sahip 30-yaşında erkek hasta sunulmuş ve nadir görülen bu patolojinin güncel cerrahi tedavi stratejileri tartışılmıştır.

Anahtar Kelimeler: Aort koarktasyonu, cerrahi girişim

Turkiye Klinikleri J Cardiovasc Sci 2008;20(3):192-6

Coarctation of the aorta (CoA) accounts for approximately %6-8 of all live births with congenital heart disease, which is the 4th common congenital heart defect that necessitates cardiac catheterization and surgical intervention in the first year of life.[1] CoA is defined as a discrete stenosis in the proximal descending thoracic aorta. It is most commonly located just distal to the origin of the left subclavian artery (LSA).[2] Preductal (infantile) type CoA (proximal to the LSA) is pretty rare (about 1% of all cases). The first surgical repair of CoA was performed by Crafoord and Nylin in 1944.[3] This pathology is typically a disease of childhood and early adulthood and can cause reduced life expectancy in patients who have not undergone correction. The natural history of unrepaired CoA includes the development of systemic hypertension and subsequent morbidity and death

from cardiovascular complications such as congestive failure, aortic rupture, stroke and endocarditis. If the condition is uncorrected, most patients die by their 5th decade of life, and very few survive to older ages.[4] Endovascular balloon angioplasty for CoA was introduced in 1982 and supplemented by stent implantation 10 years later.[5]

Surgical therapy is regarded to be a gold standard treatment for these patients but the choice of surgical technique is still a debated issue. In this report, we described an adult patient with infantile type CoA treated successfully with dacron patchplasty.

CASE REPORT

In September 2007, a 30-year-old man attended at our hospital with dizziness, general malaise, frequent headaches, weakness in his legs and hypertension. He had suffered from effort intolerance since childhood. On physical examination, peripheral pulses were all palpable bilaterally over carotid arteries, upper limbs and lower limbs. However, radial artery pulse was stronger in the right arm than in the left. Bounding pulses in the neck were also detected. The blood pressure was 151/69 mmHg in the right arm and 114/71 mmHg in the left arm. However there was no significant difference in blood pressure between upper and lower limbs. The electrocardiogram showed left ventricular hypertrophy and regular rhythm at 64 beats/min. Chest radiography revealed no pathologic abnormality. Transthoracic echocardiography demonstrated significant narrowing of the aortic segment just proximal to the LSA with a maximum gradient of 40 mmHg. Magnetic resonance angiography (MRA) revealed severe hypoplasia of the transverse aortic arch, proximal to the origin of the LSA. The diameter of the aortic arch was 37 mm before coarctation, decreased to 8 mm at the coarctation and 33 mm after coarctation (Figure 1). Both common carotid arteries were dilated. Extensive collateral vessels from the vertebrobasilar system down to the posterior chest wall and the spine supplied the descending thoracic aorta. The patient was scheduled to surgical repair.

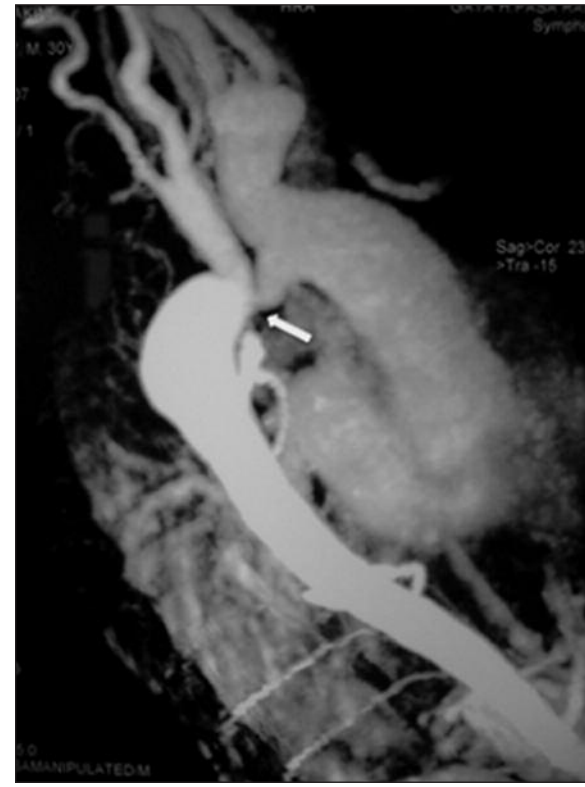

FIGURE 1: Magnetic resonance angiography (left anterior oblique projection) indicates infantile (preductal) coarctation of the aorta

After induction of general anesthesia and double-lumen intubation of the trachea, right radial artery catheter was placed for continuous arterial pressure. A left posterolateral thoracotomy was performed through the 4th intercostal space. After opening of mediastinal pleura, a discrete ring-like coarctation was observed in the aorta just proximal to the level of the LSA orifice. The phrenic and vagus nerves were carefully dissected and preserved. The distal aortic arch beyond the origin of the left carotid artery was found to be tortuous and elongated. The aortic arch proximal to the left carotid artery, the LSA and the aorta distal to the LSA were dissected and encircled. The ligamentum arteriosum was divided. The aortic arch was cross-clamped just distal to the left common carotid artery proximally and at the level of the mid-descending thoracic aorta distally. The LSA was occluded with a snare. Upon application of aortic cross-clamps, the distal pressure in the left femoral artery was 55 mmHg; thereby none of the preventive measures

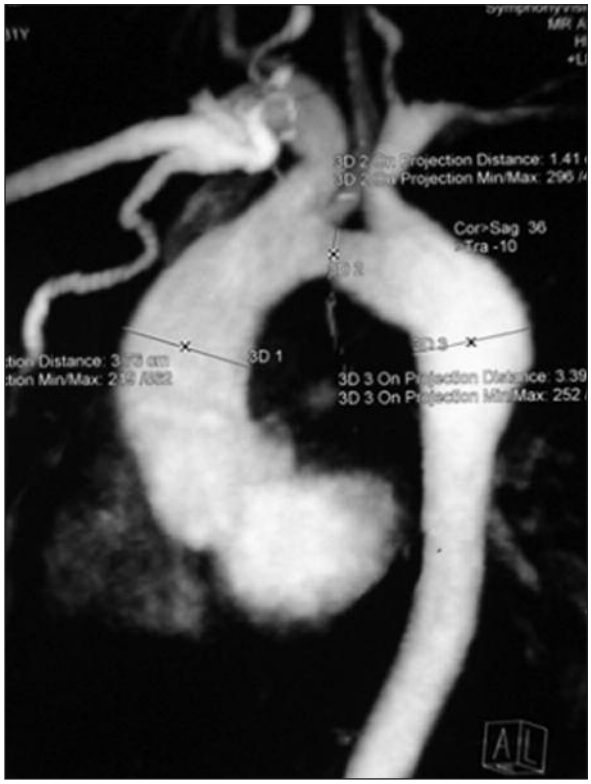

FIGURE 2: Magnetic resonance angiography shows postoperative condition with preductal aortic arch and coarctation treated with dacron patch.

described for spinal cord protection were utilized. The region of aortic coarctation was partially excised and aortic continuity was restored with a Dacron patch (Figure 2).

After completion of aortic repair, arterial pulses in all extremities were found to be equal. The rest of the procedure was completed in a standard fashion. Early postoperative course was uneventful. Postoperatively, the pressure gradients between the right and left arms dropped to below 10 mmHg in intensive care unit (132/83 vs 124/74 mmHg, respectively). Transoesophageal echocardiography revealed 7 mmhg residuel aortic gradient in the post operative 3th month The patient was discharged from the hospital on postoperative day 7 and is still doing well.

DISCUSSION

Coarctation of the aorta, a congenital narrowing of the aorta that usually occurs near the site of insertion of the ductus arteriosus, is classified anatomically as preductal (infantile) or postductal (adult). This lesion, which is seen primarily in the infantile (preductal) form, is the 8th most common congenital heart defect but rarely present in adults.[6] Adult patients with CoA have a range of clinical presentations. These include the presence of additional cardiovascular anomalies (predominantly aortic valve abnormality) and presentation with complications after coarctation repair in childhood (such as recurrent CoA or aneurysm formation). Most adult patients with coarctation are completely asymptomatic and the diagnosis are only made by investigation for the cause of hypertension, even though some symptoms occur which as matter of fact can be life threatening such as heart failure, infective endocarditis, aortic dissection or rupture, cerebrovasculer accidents.[7]

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. Surgery has still an important part to play in the management of the adult patient with CoA despite the recent advances of endovascular technology. End-to-end anastomosis, prosthetic interposition tube grafts, subclavian flap repair, and extra-anatomical corrections can be preferred for surgical treatment.[8-10] Balloon angioplasty, endovascular repair and hybrid procedures can be used as alternative treatments.[11] Angioplasty with or without stenting has been used successfully to treat lesions in both children and adults, primarily in those patients with recurrent coarctation.[5] Each Technique has specific advantages, disadvanteges and long-term outcome profiles. The choice of procedure depends on several variables, including the specific anatomy of the CoA, the patient's age and the surgeon's preference.

In this report, we used the prosthetic patch aortoplasty method. Our technique's superiority against the other methods is a single incision without graft related problems, no necessity to cardiopulmonary bypass, hypotermic circulatuar arrest and shorter operation time. Even though

some authors suggested different extra-anatomic routes for the bypass conduit, this technique can be used for interrupted aortic arch and in combined procedures such as with valve and coronary artery surgery. Adhesions from previous surgery may make a left thoracotomy approach difficult and extra-anatomical correction facilitates blood through an alternate route without disturbing the adhesions.[9]

A subclavian flap procedure utilizes the LSA to enlarge the aortic lumen and is thought to be associated with a reduced incidence of restenosis when undertaken in early life.[8] Good long term results in childhood have been reported by using this technique but it is not recommended for adults because of the risk of upper limb ischaemia despite the presence of collateral blood supply to the upper limb. End-to-end anastomosis, in which the aorta is extensively mobilized and the descending aorta is anastomozed to the underside of the arch, is especially useful in the setting of a hypoplastic arch. In teenaged children or adults this approach is often technically difficult owing to the excessive tension created at the anastomotic site. The technique of subclavian flap aortoplasty is considered to be associated with lower rates of re-coarctation than resection with end-to-end anostomosis, because it avoids the need for a circumferential aortic anostomosis which fails to grow adequately. The sacrifice of the left subclavian artery, which this technique entails, results in diminished pulses, minimal shortening and often claudication during exercise in the left arm.

Complications of the surgical treatment, including postoperative hemorrhage, recurrent laryngeal nerve injury, paraplegia, Post-coarctation syndrome (mesenteric adenitis), hypertension, recoarctation, aortic aneurysm formation and rupture, sudden death, ischemic heart disease, heart failure and cerebrovascular accidents have been reported[9]. Postoperative Paradoxical hypertension, if present, is usually secondary to increased sympathetic activity in the first 24 hours,[12] and to increased renin-angiotensin activity there after. Persistent hypertension is a substantial problem after surgical repair of aortic coarctation and is more frequent in patients who undergo surgery at a more advanced age. Bouchart and colleagues reported long-term follow-up results in 35 patients who underwent coarctation repair at an age older than 20 years. In their series, all 5 of the patients who were operated at age 40 or older remained hypertensive and needed antihypertensive medication at late follow up.[13] Despite good long-term results after surgical repair of coarctation of the aorta, patients should be followed on a regular basis primarily in order to recognize systemic hypertension as early as possible and to improve the long-term outcome by antihypertensive treatment.

In the presentation of infantile aortic coarctation, we performed successful surgical repair in a 30-year-old man who had severe symptoms and uncontrolled hypertension caused by infantile coarctation of the aorta. We conclude that repair of CoA without CPB can be performed safely and effectively using this technique without the risk of graft related problems. Beside this the operation time is shorter and surgery is easier than other surgical procedures.

Aknowledgement

We thank Mrs. Maya Celik for English corrections.

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