OLGU SUNUMU

Type A aortic interruption in a 22 years old male

22 yaşında erkek hastada Tip A aort interüpsiyonu

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SUMMARY

The aortic interruption is a rare congenital malformation. It is defined as a loss of luminal contiunity between the ascending and descending portion of aorta. This entity was reported 3 per million in live births in the literacy and highly fatal if left untreated. We presented type A aortic interruption in a hypertensive, symptomatic 22 years old man.

Keywords: aortic interruption, congenital, malformation, CT angiography

ÖZET

Aortik interüpsiyon nadir rastlanan bir konjenital malformasyondur. Asendan ve desendan aorta arasındaki luminal devamlılığın yokluğu olarak tariflenir. Literatürde üç milyon canlı doğumda bir bildirilmiştir ve tedavi edilmezse oldukça ölümcüldür. Burada 22 yaşında hipertansif semptomatik erkek hastada tip a aortik interüpsiyon olgumuzu sunduk.

Anahtar kelimeler : aortik interruption, konjenital, malformasyon, BT anjiografi

İntroduction

Aortic interruption is a rare congenital malformation and was first described in 1778 by Steidele(1). This condition is defined as a loss of luminal continuty between the ascending and descending portion of aorta.(2) Interrupted aortic arch is usually diagnosed and repaired during the neonatal period. It has three morphological type. Type A has luminal discountity distal to the left subclavian artery, type B has between left carotid artery and left subclavian artery, type C has between left carotid artery and innominate artery.(3)The most common type is B (53%), followed by A (43%) and C (4%) (4). In these patients, the distal circulation was provided by aortic collateral arteries. Prognosis is very poor in untreated cases. There are a lot of kind of surgical procedure for treatment purpose.We presented a successful surgical repair, performed with a single-stage extra-anatomic technique on left posterolateral thoracotomy.

Case Report

In March 2013, a 22 years old man admitted to our center with palpitation and cold lower extremities around one year. History revealed different blood pressure more than 20 mm Hg between upper and lower extremities. On examination, lower extremities pulses were feeble, whereas arm pulses were very stronger. Diastolic murmur was heard on the left side of the sternum. Chest X-ray was normal. Bicuspid aorta was determined with echocardiography but structure and function of the leaflets were normal. Interrupted aortic arch could not be showed with aortography so Magnetic Resonance Imaging (MRI) was performed to describe the lesion. On the MRI, coarctation was starting approximately 2 cm beyond the origin of the left subclavian artery. The ascending and descending aortic segments were otherwise normal. Extensive collateral circulation was seen. We performed single stage extra-anatomic bypass through left posterolateral thoracotomy. Dacron graft was placed between ascending and descending portion of the thoracic aorta, under the proximal and distal simplex clamping(Fig.1). Perioperataive complication was not seen and postoperative bleeding was minimal. The patient was taken into uneventfully intensive care unit at postoperative time. He was discharged out of our center on postoperative fifth day. He was controlled on postoperative first month, he had not palpitation and his lower extremities pulses were very well. postoperative MRI revealed patent grafts and good distal flow.

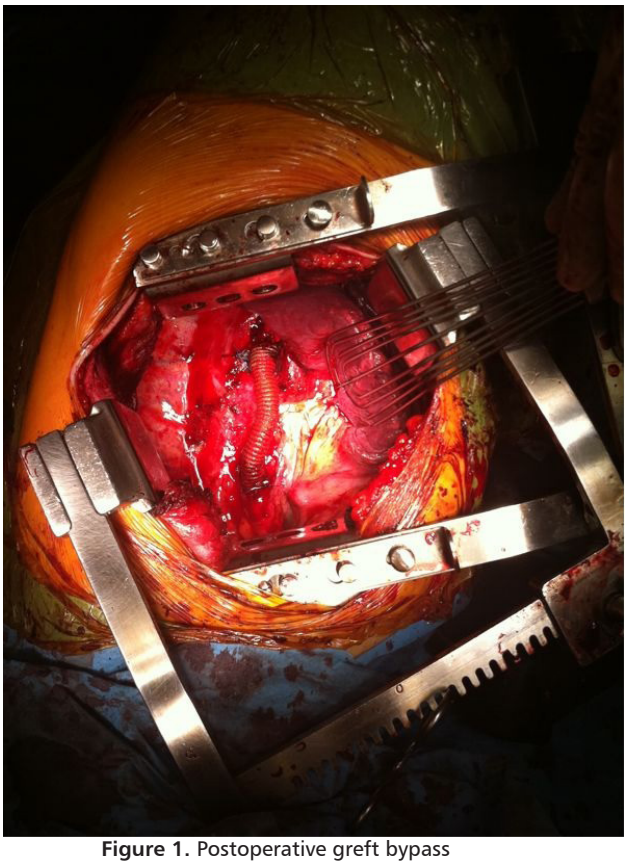

Figure 1. Postoperative greft bypass

Discussion

Interruption of aortic arch is a rare congenital malformation and is defined as a loss of luminal continuty between ascending and descending portion of thoracic aorta.(5) It was reported 3 per million live births. Only a few cases of IAA in adults have been reported in the medical literature.(6)The mortality rate of patients with untreated interrupted aortic arch can exceed 90% from birth to age 1. Interrupted aortic arch was classified into 3 types depending upon the site of discontinuty by Celoria and Patton in 1959. Distal to the left subclavian artery (type A), between the left common carotid and left subclavian arteries (type B), or between the brachiocephalic and left common carotid arteries (type C). Type B interruption was the most common (53%), followed by type A (43%) and type C (4%). Other cardiac anomalies (ventricular septal defect, patent ductus arteriosus, bicuspid aortic valve, left ventricular outflow tract obstruction, aortopulmonary window, and truncus arteriosus) may occur simultaneously. Infrequently, interrupted aortic aorta can be present in adulthood if it has adequately collateral circulation to provide other systems. The anomaly is identified with echocardiography, computer tomography angiography and magnetic resonance imagination. In adults, IAA has typically been repaired in a single stage by means of an extra-anatomic approach. There are no neurological, renal and gastrointesinal systems complications, secondary interventions, rehospitalization and deaths. We performed single-stage extra-anatomic bypass with dacron graft in 22 yeras old male's type A of interrupted aortic arch. We didn't observe postoperative drainage. The patient recovered without any sequela in postoperative period. On follow-up, the graft was patent at postoperative 1 month and blood flow was very well. In conclusion, IAA is a rare congenital abnormality and rarely reported in adults. MRI and CT angiography are useful diagnostic tools for interrupted aortic arch.(2) Early diagnosis and appropriate surgical technique is very important for the patitnet's survival. Thus we consider that single-stage extraanatomic bypass technique is very safe surgical method in the treatment of adult patient.

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