Coarctation of the aorta in an adult patient case report and review of the literature

Erişkin hastada aort koarktasyonu olgu sunumu ve literatürün gözden geçirilmesi

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Geliş Tarihi / Received : 6 Mayıs 2024 Kabul Tarihi / Accepted: 28 Temmuz 2024

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Hippocrates Medical Journal / Hippocrates Med J 2024, 4(2): 78-82 DOI: https://doi.org/10.58961/hmj.1478872

Abstract

Coarctation of the aorta is a congenital heart disease defined as a segmental narrowing of the aorta distal to the left subclavian artery. The condition is usually diagnosed in childhood and rarely in adulthood. Severe coarctation causes high mortality and heart failure in childhood, whereas adult coarctation is usually asymptomatic. The most common clinical manifestation in adults is systemic hypertension. Coarctation of the aorta is a rare cause of secondary hypertension and can lead to early death if not treated appropriately. In this case report, we present a 39-year-old woman with refractory hypertension who was diagnosed with coarctation of the aorta by echocardiography and computed tomography and underwent successful primary stent implantation.

Keywords Coarctation of aorta, Adult, Hypertension

Özet

Aort koarktasyonu sıklıkla sol subklavian arterin distalinde aortun segmental daralması olarak tanımlanan, konjenital kalp defektlerinde %8-10 oranında görülen, izole veya diğer kardiyak defektlerle birlikte olabilen bir konjenital kalp hastalığıdır. Aortikelastisite anormallikleri ile karakterize geniş yayılımlı bir arteriyopati olarak da tanımlanmaktadır. Durum genellikle çocukluk çağında ve nadiren yetişkinlikte teşhis edilir. Yetişkinlerde en sık görülen klinik bulgu sistemik arteriyel hipertansiyondur. Bu olgu sunumunda, ekokardiyografi ve bilgisayarlı tomografi ile aort koarktasyonu tanısı konulan ve başarılı primer stent implantasyonu uygulanan, refrakter hipertansiyonu olan 39 yaşında bir kadın hastayı sunuyoruz.

Anahtar Kelimeler Aort koarktasyonu, Erişkin, Hipertansiyon

INTRODUCTION

Coarctation of the aorta is a congenital heart disease defined as a segmental narrowing of the aorta distal to the left subclavian artery. The condition is usually diagnosed in childhood and rarely in adulthood. Severe coarctation causes high mortality and heart failure in childhood, whereas adult coarctation is usually asymptomatic. The most common clinical manifestation in adults is systemic hypertension. Coarctation of the aorta is a rare cause of secondary hypertension and can lead to early death if not treated appropriately. In this case report, we present a 39-year-old woman with refractory hypertension who was diagnosed with coarctation of the aorta by echocardiography and computed tomography and underwent successful primary stent implantation.

CASE REPORT

A 39-year-old woman suffering from hypertension for five years admitted to our clinic with resistant hypertension, headache, and claudicatio intermittent.

The patient had a history of smoking, but her family history was unremarkable. Blood pressure in the right and left arms were 170/100 mmHg, and no significant difference was determined. Fingertip pulse oximetry was 99%. Physical examination revealed a 3/6 systolic murmur across the interscapular region, and femoral pulses in the lower extremity were filiform at palpation. No abnormal finding was encountered during other system examinations. Complete blood count and biochemistry values were normal. The cardiothoracic ratio at chest x-ray was normal, and subcostal notches were observed. T negativity was observed at V1, V2, and V3 derivations in the electrocardiography (ECG) sinus rhythm (80 beats/min).

In transthoracic echocardiography stenosis in the descending aorta was observed. A maximum gradient of 51 mmHg was measured in the descending aorta after the subclavian artery. Thoracoabdominal CT angiography was performed, and postductal aortic coarctation was diagnosed **(Fig. 1).**

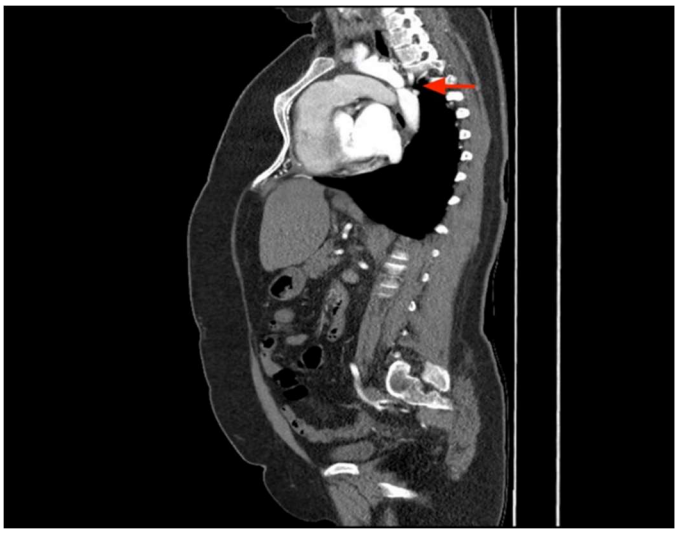

Figure 1. Thoracoabdominal CT angiography

Informed consent was obtained from the patient. Endovascular intervention was planned on the base of the patient's age and body surface area, the coarctation morphology, and the non-recurrent nature of the coarctation. The patient was taken to the catheter laboratory **(Fig.2).**

Aortography was performed under local anesthesia with a pigtail catheter from the right radial artery. A 14F 85mm sheath was localized to the right femoral artery. The lesion was passed with a 0.035 hydrophilic guidewire.

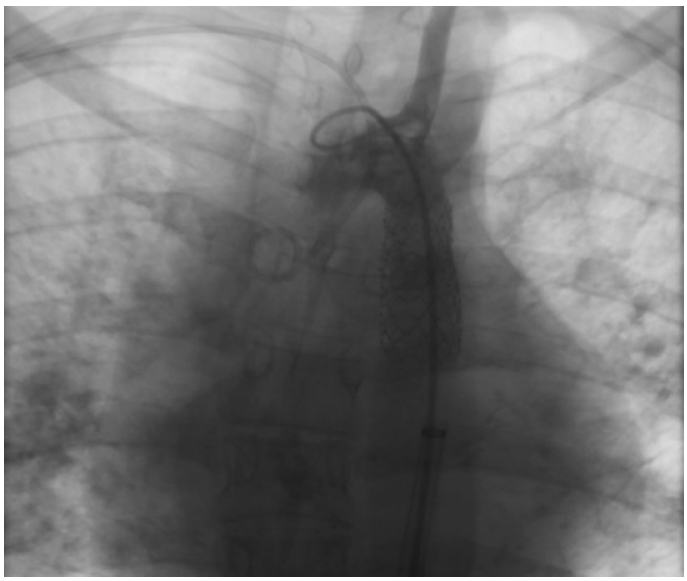

Figure 2. Angiography image after percutaneous

A 43-mm stent was inserted into the stenotic segment by loading a 18X40mm balloon catheter over the guidewire. Control aortography showed that 95% patency had been achieved in the coarctation region.After the procedure, the patient was followed up in the intensive care unit for 1 day and in the cardiovascular surgery service for 1 day. No complications were observed.

Echocardiography control was performed postoperative 1st and 3rd-monththe gradient was between 5-10 mmHg. Regression was observed in cladicatio complaint. Antihypertensive treatment was discontinued.

DISCUSSION

Aortic coarctation is a congenital heart disease characterized by narrowing of the aorta below the left subclavian artery. It affects 8-10% of congenital heart defect case sand can occural one or with other cardiac issues (1) . There are two morphological (preductal/infantile and postductal/adult) and four anatomical (isthmus, arcus, descending, and abdominal) classifications (2). It's more prevalent in males, with the preductal type common in children and postductal type in adults. Untreated adults have a life expectancy of about 35 years, with an 80% mortality rate in the fifth decade (3). Common complications include heartfailure, myocardiali nfarction, and intracranial hemorrhage. It often coexists with bicuspid aorta and other cardiac conditions like Shone's complex, ventricular septal defect, patent ductus arteriosus (PDA), and intracranial aneurysm (4).

Aortic coarctation, a congenital condition, refers to segmental narrowing in the aorta. Dr. Bonnet classified it into preductal and postductal types in the early 20th century (2). Aortic coarctation is common in infants (preductal) and adults (postductal), affecting about 4 in 10,000 births, with a higher prevalence in males. Its exact cause is unknown, but theories like ductal tissue migration have been proposed.

The flow theory suggests hypoplasia in the transverse arch or isthmus due to decreased blood flow caused by intracardiac anomalies. Genetic factors, including NOTCH1 gene mutations, have been linked to coarctation, with increased incidence seen in conditions like Turner syndrome (15-35%). Family members of affected individuals also have a higher likelihood of the anomaly (5,6).

Echocardiography and CT angiography are the main diagnostic tests for coarctation of the aorta. Echocardiography, especially Doppler examination from the suprasternal region, confirms the diagnosis by revealing turbulent flow and high gradients in the coarctated area. Cardiac catheterization is used to locate the coarctation, measure pressure, assess the distance from the subclavian artery and plan endovascular interventions.

Severe aortic coarctation in newborns can cause high mortality from heart failure. In adults, it leads to hypertension and possible claudication. Diagnosis considers high upper limb blood pressure and low lower limb pressure. The exact cause is uncertain, possibly related to nervous system imbalance and vascular issues (7,8).

The recommended treatment for aortic coarctation involves addressing systemic arterial hypertension and a ≥50% anatomical stenosis with a >20 mmHg difference in upper and lower extremity systolic blood pressure. Early intervention is crucial to prevent coarctation-related cardiovascular events.

Treatment options include percutaneous interventions or surgery. The choice between them is debated. According to the American College of Cardiology (ACC)/American Heart Association (AHA) guideline, interventions should be decided by a cardiac team based on patient specifics. Surgica lintervention is preferred for patients with accompanying cardiac anomalies, heart valve disease, long coarctation segment, or isthmus hypoplasia (1).

According to ESC guidelines, interventions (surgical or endovascular) are recommended for adult patients with a >20 mmHg blood pressure difference between upper and lower extremities, upper extremity hypertension (>140/90 mmHg), or severe left ventricular hypertrophy (9). Intervention may be considered in cases with ≥50% aortic stenosis, irrespective of pressure gradient and presence of hypertension.

The first surgical repair was performed by Crafoord in 1944. Balloon angioplasty was performed by Lock et al. in 1982, followed by the first stenting procedure in 1991(10,11). Surgical techniques for aortic coarctation depend on segment length and collateral circulation. Approach varies based on cardiac issues, often using left posterolateral thoracotomy.

In endovascular treatment, adults with aortic coarctation often undergo percutaneous balloon angioplasty or stent placement for recoarctations and aneurysms post-surgery. Balloon angioplasty may lead to post-procedural aneurysms due to controlled tearing, and recoarctation risk is higher compared to surgery (12,13). Stents are preferred in adults for their appropriate aortic lumen diameter, low residual gradient, and lower complication rates. Coated stents are popular, but consensus on their use is lacking (14).

Despite successful surgical or endovascular treatments, coarctation of the aorta persists in 25-30% of patients during follow-ups. Post-repair hypertension is more common in those treated late, especially in patients over nine years old, as shown in a study by Morgan et al. No specificage-related timing for treatment has been established (15).

Aortic coarctation, a congenital heart issue, requires surgery or endovascular procedures for correction. Ongoing monitoring is vital due to possible complications. Endovascular methods, such as stenting, reduce blood pressure, lower drug reliance, and minimize complications, offering a promising alternative with shorter hospital stays.

Considering that our patient was a 39-year-old woman with a tight native aortic coarctation (51 mmHg gradient) and poststenotic dilatation of the descending aorta, closed stent implantation was the best option in this case because of the high risk of dissection or rupture with conventional stenting.

Ethical Declerations

Since the text is a case report, ethical permission was not obtained, but a written consent form was obtained from the patient.

Conflict of Interest Statement:

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, shareholding and similar situations in any firm.

Financial Disclosure:

There is no supporting organization

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