OLGU SUNUMU CASE REPORT

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Percutaneous Aortic Stent Implantation in an Adult Patient with Interrupted Aortic Arch

Aortik Ark Kesintisi Olan Erişkin Hastada Perkütan Aort Stent İmplantasyonu

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ABSTRACT An interrupted aortic arch (IAA) is an infrequent congenital or acquired malformation defined as a complete absence or discontinuation of a portion of the aortic arch. In most cases especially in neonates, IAA is accompanied by a cardiac malformation or genetic syndromes. IAA has a very poor prognosis in patients without surgical treatment, so it is rarely detected in adulthood. Nowadays the definitive treatment option is still surgery. However, percutaneous aortic stent implantation may be a life-saving alternative treatment method, especially in patients with high surgical risk. Herein we present a very rare case of Type A IAA in a 55-year-old female patient which was successfully treated with percutaneous aortic stent implantation.

Keywords: Aortic arch syndromes; aorta, thoracic; angioplasty

ÖZET Aortik ark kesintisi (AAK), aort arkının bir kısmının tamamen yokluğu veya kesintiye uğraması olarak tanımlanan, nadir görülen bir konjenital veya kazanılmış malformasyondur. Olguların çoğunda, özellikle yenidoğanlarda, AAK'ye bir kardiyak malformasyon veya genetik sendrom eşlik eder. Cerrahi tedavi yapılmayan hastalarda AAK'nin prognozu çok kötüdür, bu nedenle erişkin yaşlarda nadiren tespit edilir. Günümüzde hâlâ cerrahi altın standart tedavi yöntemidir. Ancak özellikle cerrahi riski yüksek hastalarda, perkütan aort stent implantasyonu hayat kurtaran alternatif bir tedavi yöntemi olabilir. Biz burada, perkütan aortik stent implantasyonu ile başarılı bir şekilde tedavi ettiğimiz, 55 yaşında bir kadın hastada çok nadir görülen Tip A AAK olgusunu sunuyoruz.

Anahtar Kelimeler: Aort kavsi sendromları; aort, torasik; anjiyoplasti

Interrupted aortic arch (IAA), an uncommon malformation of the aortic arch, is defined as a loss of luminal continuity among the ascending and descending portions of the aorta. The incidence of IAA among congenital heart defects is around 1%.[1] There are 3 types of the IAA, and they are classified according to the site of the interruption by Celoria and Patton.[2] Type A is interrupted distal to the left subclavian artery (LSCA); Type B is interrupted between the left subclavian and left carotid artery (LCA), and Type C is interrupted proximal to the LCA. The frequency of occurrence is listed as B, A, and C, respectively. Survival into adulthood with the isolated anomaly is very rare and depends on the development of collateral circulation. Extra anatomic bypass procedures are the most preferred reconstruction method in adult patients with IAAs.[3]

CASE REPORT

A 55 -year- old mild mental retarded woman presented to our cardiology clinic because of uncontrolled hypertension although she was taking 4 antihypertensive drugs, including one diuretic.

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On physical examination, systolic blood pressure was 210 mmHg in both upper extremities in addition to strong pulses, but lower extremity pulses and pressure were weak. High voltage was encountered on the resting electrocardiogram. Transthoracic echocardiography (TTE) examination showed concentric left ventricular hypertrophy and bicuspid aortic valve but no significant gradient was detected on the valve. In Doppler analysis, surprisingly there was no significant pressure gradient distal to the aortic isthmus. We decided to perform aortic computed tomography (CT) due to weak lower extremity arterial pulses. CT angiography showed interruption of the descending aorta just 8 mm distal to LSCA and the interrupted segment was measured approximately 3 mm on CT (Figure 1). The intercostal arteries were enlarged to provide collateral flow to the descending aorta, distal to the interrupted segment. The percutaneous intervention was decided by our heart team after the patient was evaluated as high surgical risk. 6 Fr. sheaths were inserted into the left brachial and right femoral arteries. A right diagnostic catheter was advanced from the left brachial and a multipurpose catheter from the right femoral artery. Trans-stenotic gradient was 70 mmHg. The images were taken from both catheters but there was no anterograde and retrograde dye flow (Figure 2). First, we tried to cross the interrupted segment with a stiff (Hi-Torque Progress 120, a tipping load of 13.6 g, Abbot Vasc, Illinois, USA) guidewire and we successfully crossed the distal segment of the aorta. However, a dissection was detected in the control image after advancing to the interrupted segment of the aorta (Figure 3). So, then we used a relatively soft second hydrophilic guidewire (Hi-Torque Pilot 150, a tip load of 2.7 g, Abbott Vasc, Illinois, USA), which crossed the distal aorta without any problem.

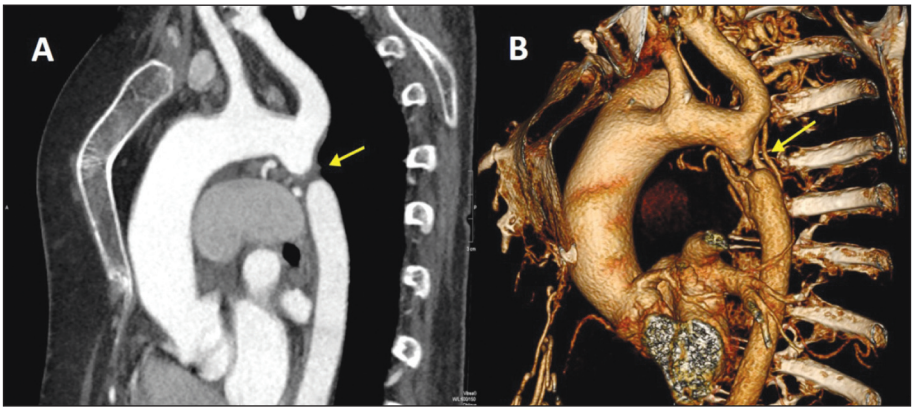

FIGURE 1: A-B) 2D and 3D computed tomography angiography showed interruption segment of the aorta.

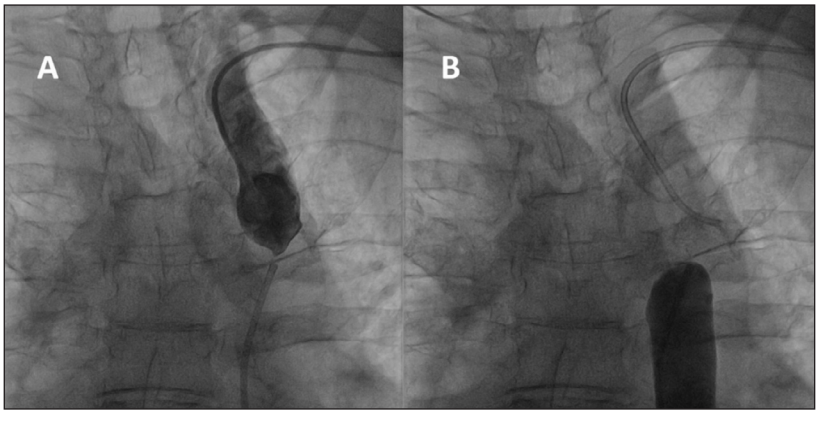

FIGURE 2: A-B) The angiographic images were taken from both catheters' interrupted segments of the aorta..

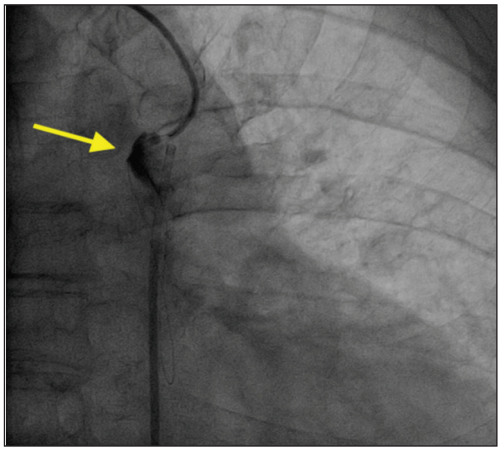

FIGURE 3: A dissection was detected in the control image after advancing to the interrupted segment of the aorta.

Then a 6F right diagnostic catheter passed to the distal aorta. We exchanged the wire with 0,035 inches amplatzer 300 cm super-stiff wire (Abbott Vascular, Santa Clara, California). The 0,035 wire was snared from the right femoral artery and externalized from the right femoral sheath. A 45×18 mm covered Cheatham Platinum (CCP, Numed Inc., Hopkinton, NY, USA) stent was pre-mounted on NuMED Balloon-in-Balloon (NuMED, Inc.; Hopkinton, NY) catheters. The stent was advanced over the wire into an interrupted segment and implantation without a problem via 14 Fr. Mullins long sheaths (Cook Medical, Bloomington, Indiana, USA) (Figure 4). After stent placement, control angiography showed a good result and the pressure gradient between proximal and distal aorta was 5 mmHg. There were no complications after the procedure and the patient's blood pressure has dropped to normal values. Informed consent was obtained from the patient and her legal guardian.

DISCUSSION

IAA is a rare anomaly that is defined simply as the complete disconnection of 2 parts of the aorta. If IAA is not treated in the neonatal period, it is very unlikely to be compatible with life, so isolated IAA rarely reaches adulthood.[1] Neonatal and adult forms of IAA should be evaluated differently. Because infants with IAA typically have associated other congenital heart defects while adults have not; Type B interruption is common in infants, whereas Type A interruption is most frequently reported in adults.[3-5] Substantial collateral circulation must be present to maintain flow and make possible survival, as happened with our patient. In some cases, especially in adult patients, the initial affection would be a coarctation of the aorta, which evolves to the progressive closing of the lumen or patent ductus arteriosus closes over time as collateral circulation increases. There are various methods for diagnosing IAA. First of all, especially in daily practice, when performing the physical examination of resistant hypertension patients, we should measure the blood pressure in both upper and lower extremities so as not to disregard aortic pathologies. Even though TTE is the first method at the diagnostic stage, angiographic reconstructions of the images obtained with multi-slice CT units are reliable non-invasive diagnostic modalities for the precise diag-

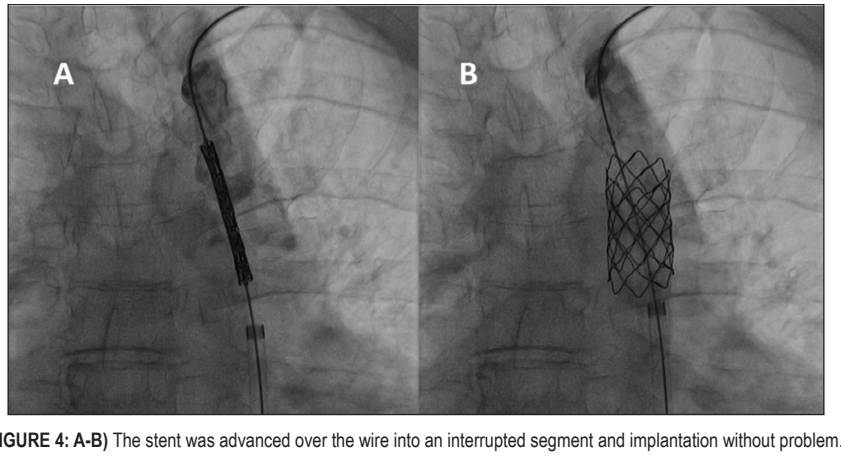

FIGURE 4: A-B) The stent was advanced over the wire into an interrupted segment and implantation without problem.

nosis of the IAA.[6] The choice of treatment depends on; age, anatomy and location, aortic alignment, and whether there is a saccular aneurysm. Nowadays surgery is still the definitive treatment of choice. Extra-anatomic bypass procedures are the most frequently preferred method of reconstruction in adult patients with IAAs.[3,5,7-9] However percutaneous aortic stent implantation is an increasing treatment option in selected patients with IAA, accompanied by advances in invasive cardiology. Crossing the atretic segment percutaneously is technically the most difficult part of the cases. Invasive cardiologists overcame this challenge with different methods such as radiofrequency perforation, transseptal needle, or stiff guidewire techniques in the literature. We overcame this anomaly using two exclusive guidewires Hi-Torque Progress 120 and Hi-Torque Pilot 150. Even with successful stenting in IAA or coarctation of aorta (CoA) patients, long-term complications such as stent fracture, stent migration, aneurysm development, and hypertension may develop in their long-term follow-up. Therefore, patients should be followed closely in the postoperative period. The results of the COAST study, which is the most important study on this subject showed us during the two-year follow-up no deaths, serious adverse events, or surgical interventions were reported. All patients experienced satisfactory post-procedural results with low rates of complications including aneurysms (5.7%) and stent fracture (11% at 2 years) without loss of stent integrity, stent embolization, aortic wall injury, or re-obstruction.[10] Chronic hypertension remains present in approximately 50% of the patients with CoA or IAA even in the presence of an anatomically satisfactory repair, especially in adult patients. The exact pathophysiology of late-onset hypertension after CoA or IAA repair remains to be intricated. Reduced aortic compliance and an abnormal baroreceptor function may explain this. Post-treatment hypertension is a risk factor for premature death and requires aggressive treatment. Follow-up blood pressure measurements and exercise testing are important, as exercise-induced hypertension can predict future systemic hypertension. Timely intervention is crucial, as early treatment may prevent cardiovascular complications. According to recent studies, percutaneous aortic stent implantation is an alternative, easy and safe procedure that reestablished the flow with no significant gradient across the aorta.[10,11] Especially in patients with high surgical risk, percutaneous stent implantation should be considered as an important life-saving alternative treatment.

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Conflict of Interest

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, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Mustafa Demir, Ömer Faruk Keskin;* ***Design:*** *Atila İyisoy, Bilgehan Savaş;* ***Control/Supervision:*** *Mustafa Demir, Ömer Faruk Keskin;* ***Data Collection and/or Processing:*** *Mustafa Demir, Atila İyisoy;* ***Analysis and/or Interpretation:*** *Atila İyisoy, Bilgehan Savaş;* ***Literature Review:*** *Ömer Faruk Keskin, Bilgehan Savaş Öz;* ***Writing the Article:*** *Mustafa Demir, Ömer Faruk Keskin;* ***Critical Review:*** *Atila İyisoy, Bilgehan Savaş.*

REFERENCES

1. Fiore AC, Peterson RE, Huddleston CB. Repair of interrupted aortic arch with ventricular septal defect. In: Ungerleider RM, Meliones JN, McMillan KN, Cooper DS, Jacobs JP, eds. Critical Heart Disease in Infants and Children. 3rd ed. Philadelphia: Elsevier; 2019. p.565-71. [Crossref]
2. Celoria GC, Patton RB. Congenital absence of the aortic arch. Am Heart J. 1959;58:407-13. [Crossref] [PubMed]
3. Sai Krishna C, Bhan A, Sharma S, Kiran U, Venugopal P. Interruption of aortic arch in adults: surgical experience with extra-anatomic bypass. Tex Heart Inst J. 2005;32(2):147-50. [PubMed] [PMC]
4. Atallah J, Robertson M, Dyck J, Ross DB. The survival of a newborn with an interrupted aortic arch and a closed ductus arteriosus. Congenit Heart Dis. 2008;3(2):144-5. [Crossref] [PubMed]
5. Borgohain S, Gupta A, Grover V, Gupta VK. Isolated interrupted aortic arch in an 18-year-old man. Tex Heart Inst J. 2013;40(1):79-81. [PubMed] [PMC]
6. Gran F, Barber I, Betrián P. Interrupción del arco aórtico [Interrupted aortic arch]. Rev Esp Cardiol. 2008;61(6):629. Spanish. [Crossref] [PubMed]
7. Shinkawa T, Jaquiss RD, Imamura M. Single institutional experience of interrupted aortic arch repair over 28 years. Interact Cardiovasc Thorac Surg. 2012;14(5):551-5. [Crossref] [PubMed] [PMC]
8. Özeren M, Aytaçoğlu B, Karaca K, Vezir Ö, Sucu N. The single-stage extra-anatomic repair of an isolated interrupted aortic arch in an adult and a review of the literature. Turk Gogus Kalp Damar. 2012;20(3):615-8. [Crossref]
9. Özkara A, Çetin G, Köner Ö, Saltık L, Kansız E. Transposition of the great arteries with ventricular septal defect and interrupted aortic arch: a successful surgical correction with one stage and two separate incisions. Turk Gogus Kalp Damar. 2006;14(3):230-2. [Link]
10. Meadows J, Minahan M, McElhinney DB, McEnaney K, Ringel R; COAST Investigators*. Intermediate Outcomes in the Prospective, Multicenter Coarctation of the Aorta Stent Trial (COAST). Circulation. 2015;131(19):1656-64. [Crossref] [PubMed]
11. Suárez de Lezo J, Romero M, Pan M, Suárez de Lezo J, Segura J, Ojeda S, et al. Stent repair for complex coarctation of aorta. JACC Cardiovasc Interv. 2015;8(10):1368-79. [Crossref] [PubMed]