Pulseless Four Extremities: Atypically Located Coarctation With Concomitant Aberrant Right Subclavian Artery

Nabızsız Dört Ekstremite: Aberran Sağ Subklavian Arter Birlikteliğinde Atipik Yerleşimli Koarktasyon

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ABSTRACT

An unusual case of pulseless four extremities caused by atypically located critical coarctation of aorta with aberrant right subclavian artery in a 21 year old patient without any symptom for years such as syncope is presented. The patient's aortography demonstrated severe coarctation proximal to the left subclavian artery, accompanied by aneurysm formation and aberrant right subclavian artery. This case report describes the successful diagnosis of the interesting patient mentioned above and the surgical complete correction procedure. Patient was discharged uneventfully after 12 days of follow-up with palpable pulses and no neurological complications.

Keywords: Pulseless, Coarctation of aorta, Aberrant right subclavian artery

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ÖZET

Yıllardır senkop gibi herhangi bir semptomu olmayan 21 yaşında bir hastada aberran sağ subklavian arter ile atipik yerleşimli kritik aort koarktasyonu sonucu oluşan nabızsız dört ekstremitenin olağandışı bir olgusu sunulmaktadır. Hastanın aortografisinde anevrizma formasyonu ve aberran sağ subklavian arterin eşlik ettiği, sol subklavian arter proksimalinde yerleşmiş olan şiddetli koarktasyon görüldü. Bu olgu sunumu, yukarıda bahsedilen ilginç hastanın başarılı teşhisini ve cerrahi tam düzeltme prosedürünü anlatmaktadır. Herhangi bir nörolojik komplikasyon gelişmeyen ve dört ekstremite nabızları palpabl olan hasta postoperatif 12. günde sorunsuz bir şekilde taburcu edildi.

Anahtar Sözcükler : Nabızsız, Aort koarktasyonu, Aberran sağ subklavian arter

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INTRODUCTION

Coarctation of aorta (CoA) is a common defect with a prevalence of 4%-8% among all congenital heart defects (1). It can be described as a constricted aortic segment, usually located in juxtaductal plane, with histological properties such as shelf-like medial thickening and superimposed neointimal tissue. Although being the most encountered location is aortic isthmus, it can be located in arcus, ascending-descending thoracic or abdominal aorta. Such locations are rare and should cause suspicion on inflammatory or autoimmune diseases including Takayasu arteritis, fibromusculary dysplasia or neurofibromatosis. It can be occur as an isolated defect or accompanied by other lesions, most commonly bicuspid aorta(2) and ventricular septal defect. Clinical presentation varies on the degree and location of coarctation, size and number of the collateral vessels and state of the ductus arteriosus. While severe stenosis causes congestive heart failure and shock in neonates, a milder stenosis may remain asymptomatic until adulthood (3). The aim of this case presentation is to introduce the successful repair of a rare clinical condition in which the arterial circulation of the four extremities originates from the distal part of the stenosis due to aberrant right subclavian artery (ARSA) accompanying atypically located coarctation of the aorta.

CASE REPORT

A 21-year-old woman with a 3-month history of chest pain, cough, leg pain and weight loss, underwent thoracic CT, resulting in diagnosis of CoA and aneurysm formations at descending aorta. She was admitted to our hospital with CT results at July 2019. At physical examination, pulseless four extremities, pansystolic murmur at left sternal edge with loud S2 heart sound noted. Her blood pressure was normal and no pressure or saturation difference was established between four extremities. Besides rightward axis, her ECG was normal. Chest X-ray revealed a widened superior mediastinum and prominent aortic knob, no cardiomegaly was detected. Echocardiographic examination revealed no concomitant cardiac anomaly except mild mitral and tricuspid regurgitation. For further evaluation, we obtained arcus aortography, CT and MRI angiography. Rheumatology Department evaluated the patient for Takayasu arteritis and after imaging and blood antibody testings, this prediagnosis was ruled out.

Her aortography revealed >95% focal stenotic segment between left carotid and left subclavian artery (LSCA), 2 aneurysmatic dilatation detected at proximal descending aorta, and left subclavian artery (LSCA) was originating from aneurysmal dilatation. Thoracoabdominal BT angiography showed coarctation distally to left vertebral artery ostium, 6cm × 4cm (craniocaudal × transvers) multilobulated aneurysmal dilatation, and presence of retroesophageal ARSA originating from descending aorta (fig.1). After imaging results, the patient was diagnosed with atypical CoA and ARSA. Owing to significant and symptomatic coarctation, concomitant aneurysmal dilatation and ARSA, we planned operation of total correction.

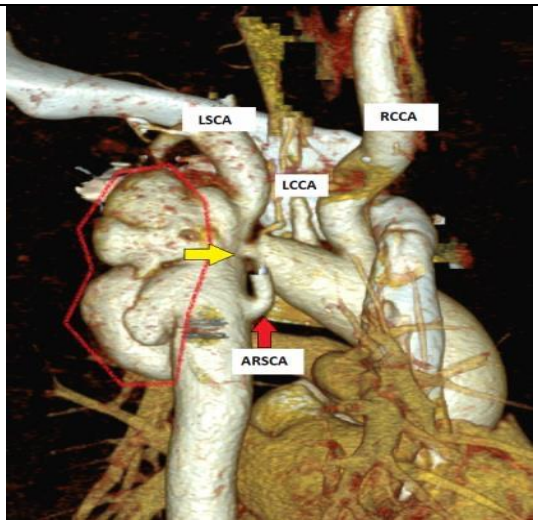

Figure 1: Three – dimensional CTA image shows an aberrant right subclavian artery(ARSA)(red arrow),severe aortic coarctation (yellow arrow) and post – stenotic dilatation (red border).(RCCA, right common carotid artery; LCCA, left common carotid artery; LSCA, left subclavian artery)

The patient was operated in August 2019. The operation was performed through a median sternotomy. Ascending and arcus aorta was dissected and it was observed that the right subclavian artery did not originate from truncus brachiocephalicus. ARSA was found by dissection towards the axilla and mobilized proximally as much as possible, it was transected from this section. Since the length of ARSA was not sufficient, the connection between right carotid artery and ARSA was provided by a 7 mm dacron graft. Another a 7 mm dacron graft was anastomosed to this graft with end-to-side technique, and a 8 mm dacron graft to right femoral artery similarly for the connection of arterial access and CPB circuits. Cardiopulmonary by-pass initiated after double arterial and right atrial cannulation, followed by aortic cross clamping and cardioplegia infusion. Proximal LSCA, which was originating from aneurysm, was detached, the proximal part was sutured and a 7 mm dacron graft anastomosis was performed to the distal part by end-to-end technique. Using 22 mm dacron graft, extra-anatomical by-pass from the arcus to the descending aorta was performed (fig.2). The aortic segment between distal transverse arch and proximal descending aorta, which contains stenotic and aneurysmatic sections, was ocluded by suturing continuously both ends. After end of CPB, the graft continued LSCA anastomosed to ascending aorta and the surgery was completed successfully (fig.2). The patient was discharged after 12 days of follow-up with significant relief of symptoms and palpable peripheral pulses. The patient continues to be followed up without any problems. (fig.3)

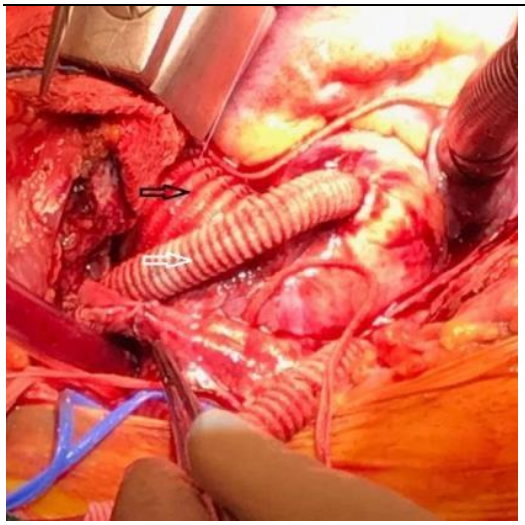

Figure 2: Intraoperative images shows the graft between arcus and descending aorta (black arrow) and other graft between left subclavian artery and ascending aorta (white arrow).

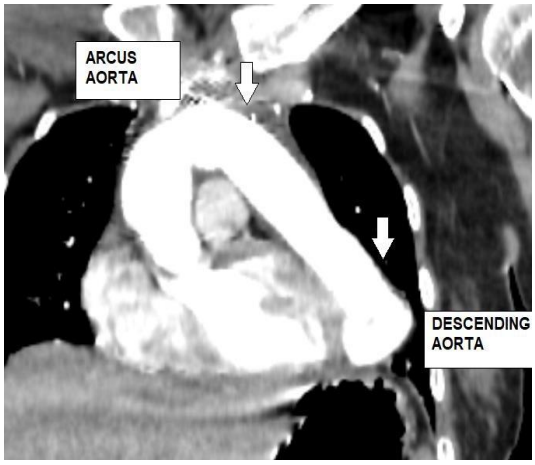

Figure 3: Postoperative CTA images shows the the graft between arcus and descending aorta (white arrow).

DISCUSSION

It is a rare condition for a patient with such severe aortic coarctation to be asymptomatic until the last three months. As it is known, subclavian steal syndrome may develop in subclavian artery stenosis due to retrograde flow in the vertebral artery (4). However, in this patient both subclavian arteries originated distal to the stenotic segment. Therefore, we think that subklavian steal syndrome and syncope did not occur in this patient since both vertebral artery flows were supplied from the arterial system distal to the stenosis zone.

Although CoA is a common anomaly among congenital heart defects and ARSA is the most common congenital aortic arch anomaly (with a prevelence of 0,5%-2%)(1), coexistence of this two anomaly is rare clinical entity, which accounts for 1% in cases of ARSA (5). This rare coexistance and atypical localization of CoA was complicating our patients clinic. In such patients, vasculitis, especially Takayasu arteritis, connective tissue diseases and some genetic syndromes associated with great arteries, should be considered in terms of etiology. In our patient, the disorders mentioned above were not found in the laboratory tests and multidisciplinary evaluations performed for differential diagnosis.

As is known, aortic coarctation surgeries are usually performed with thoracotomy approach. However, we considered that sternotomy is more appropriate in this patient, since access to the arch and ascending aorta is required for anastomosis of the distal aorta and LSCA grafts. We performed selective cannulation and anterograde perfusion of carotid artery, with femoral cannulation and retrograde perfusion of distal thoracic-abdominal aorta,visceral organs and lower extremities for avoidance of such complications. Extra-anatomical by-pass techniques are effective in patients with complex coarctation (3), and allows us to abstain from further mobilization of the aortic arch, preventing injury of surrounding structures as we performed here. As it is known, one of the most important complications in surgical procedures related to the aortic arch is neurological events. We evaluated neurological functions with NIRS monitoring throughout the surgery and we did not observe any neurological disorders. After surgery, the patient showed an uneventful recovery with no neurologic complications.

In conclusion, coarctation of the aorta located in the distal transverse arch and the coexistence of ARSA is an extremely rare condition. Despite such a critical stenosis in the aorta, it is not expected for the patient to be asymptomatic for a long time. For such a complex anomaly patient specific surgical approach should be planned to avoid neurological complications.

Conflict of interest

No conflict of interest was declared by the authors.

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