CASE REPORT

Primary coarctation-related isthmus aneurysm in an adult

Bir erişkinde primer aort koarktasyonuyla ilişkili istmus anevrizması

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Summary– A 56-year-old female patient was transferred due to the presence of a left supraclavicular pulsatile mass. Further work-up was performed to confirm diagnosis. Angiography and computed tomography were also performed. The anatomy of the thoracic aorta suggested an unknown and untreated aortic coarctation. A second aortic narrowing was identified at the aortic arch, a result of elongation of the aorta. Endovascular approach was not possible, due to complexity of the aortic anatomy, and tortuosity. Open surgical repair was successfully performed. A primary coarctation-related isthmus aneurysm is an exceptional finding in adults, and is a life-threatening condition when diagnosis is delayed. Management and treatment are controversial and challenging.

Özet– Elli altı yaşındaki bir kadın hasta sol supraklaviküler nabazanlı kitle varlığı nedeniyle sevk edildi. Tanıdan emin olmak için ileri araştırmalar yapıldı. Anjiyografi ve bilgisayarlı tomografi de çekildi. Torasik aorta görüntüsü bilinmeyen ve tedavi edilmemiş aort koarktasyonunu akla getirdi. Aort kavsinde aort elongasyonuna bağlı ikinci bir aort darlığı saptandı. Karmaşık aort anatomisi ve tortiozite nedeniyle endavasküler yaklaşım mümkün olmadı. Başarılı açık cerrahi onarım gerçekleştirildi. Primer aort koarktasyonuyla ilişkili istmus anevrizması erişkinlerde istisnai bir bulgudur. Tanı geciktiğinde yaşamı tehdit edici bir durumdur. Yönetimi, tedavisi çelişkili ve zordur.

Presently reported is the case of a patient with a left supraclavicular pulsatile mass. Angiography and computed tomography identified an aortic aneurysm located at the aortic isthmus. The anatomy of the aorta suggested an unknown and untreated aortic coarctation as the origin of this primary isthmus aneurysm. Endovascular approach was not possible, due to aortic tortuosity. Open surgical repair was successfully performed.

CASE REPORT

A 56-year-old female patient was admitted to our institution after she had noticed a left supraclavicular pulsatile mass. During childhood, she had been diagnosed with a heart murmur but had undergone no further work-up. In addition, she had undergone surgery and radiotherapy for breast cancer 2 years prior. Physical examination confirmed the presence of a supraclavicular pulsatile tumor with a loud systolic murmur. No pressure gradient between the left arm and leg was detected. Aortography and computed tomography scan were performed. The aortic arch was markedly elongated and kinked. A 41x36-mm saccular aneurysm emerged from the end of the kinked segment, growing into the left thoracic inlet, followed by a 36x33-mm fusiform aortic dilatation. Two areas of aortic narrowing were identified; the narrowest was 13x18-mm, just before the saccular aneurysm. The second was a 22x16-mm area located after the fusiform dilatation, at the level of the ligamentum arteriosus (Figure 1a-c). An ectasia of the origin of the left subclavian artery was also identified. Both mammary arteries were dilated. Echocardiography ruled out the presence of bicuspid aortic valve and aortic root pathology. Due to aortic tortuosity and small aortic di-

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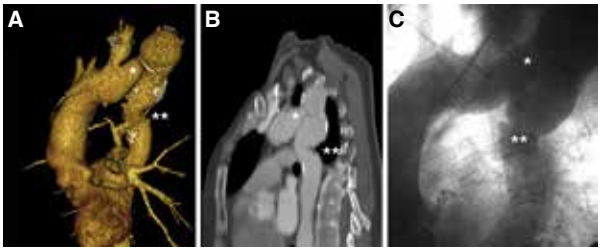

Figure 1. **(A)** Left-sided 3-dimensional computed tomographic scan reconstruction showing the aortic coarctation. Isthmic aneurysm, delimited by 2 aortic narrows (proximal* and distal** coarctation). **(B)** Computed tomographic angiography, with sagittal section showing correlation with prior 3-dimensional computed tomographic reconstruction. **(C)** Aortography of the aortic arch, showing elongated aortic aneurysm extended to the neck.

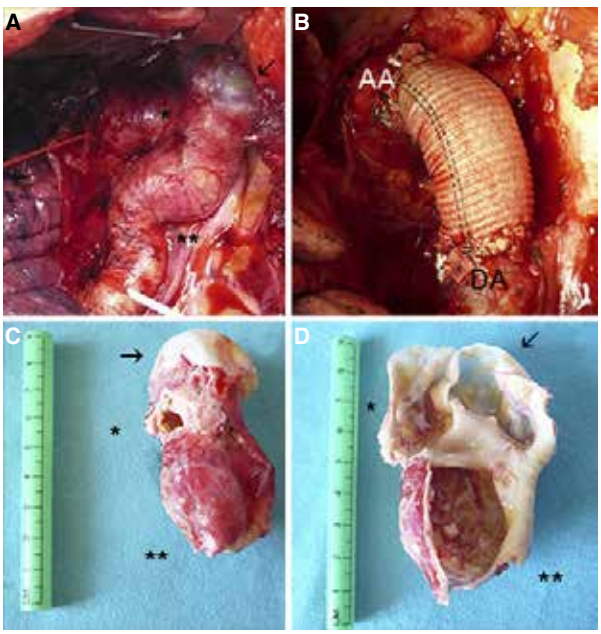

Figure 2. **(A)** Left thoracotomy incision made for the open surgical repair of aortic coarctation and aneurysm. Aortic wall weakening is visible at the upper side of the aneurysm (arrow). **(B)** Aortic bypass grafting after resection of aortic coarctation and saccular aneurysm (arrow). **(C, D)** Aortic wall specimen obtained after resection. Aortic wall fibrosis and calcification of the aorta is remarkable in comparison to the extreme weakening of the aortic wall, observed at the level of the isthmic aneurysm, rendering it nearly transparent (arrow). Aortic narrowing is patent at the proximal (*) and distal (**) sides of the aneurysm. AA: Aortic arch; DA: Descending aorta.

ameter at the level of the narrowed segments, open surgical approach was selected.

Supported by femoro-femoral cardiopulmonary bypass, the thoracic aorta was accessed via left posterolateral thoracotomy. The wall of the saccular aneurysm was found to be extremely thin and firmly adherent to the thoracic wall. In addition, the aorta was found to be narrower than expected (Figure 2a, c, d). It was replaced, immediately after the origin of the left subclavian artery to the onset of the first intercostal artery, by a 24-mm polyester graft (Hemashield Platinum; Maquet Cardiovascular LLC, Wayne, NJ, USA) (Figure 2b). Surgery was successfully performed. However, during the postoperative period, recurrent chylothorax was observed. Conservative management was initially attempted, and complete parenteral nutrition and reiterative thoracic drainage were applied. However, favorable evolution not was observed, and thoracic duct ligation by video-assisted thoracoscopy was required.

DISCUSSION

Aortic coarctation is the third most prevalent form of congenital heart disease. Accounting for 5–8% of all congenital heart defects, it is a pathology usually diagnosed during infancy.[1] However, approximately 20% of coarctations may not be diagnosed until adulthood. Less severe degrees of aortic narrowing allow for collaterals to establish around the coarctation, resulting in less severe presentations, and explaining the absence of significant transcoarctation gradients.[1–3] Adults tend to be incidentally diagnosed with hypertension, or aortic rupture or dissection, infective endocarditis, or stroke, due to signs of heart failure. Untreated thoracic aortic coarctation leads to morbidity and early demise. Mean age of death is 35 years for untreated patients, and an estimated 90% of all patients will die by their 6th decade.[1,3]

Thoracic aorta coarctation is frequently combined with other congenital defects, such as aortic arch hypoplasia or bicuspid aortic valve, occurring in 20–85% of cases. However, aortic coarctation is rarely associated with aortic aneurysms.[3–5] The presence of primary coarctation-related aortic aneurysms is exceptional, and reports have been scarce. Thus, the natural evolution of these aneurysms is unknown. When detected, prompt correction should be indicated.[1,2–5] Likely, in our patient, presence of severe aortic coarctation resulted in increased shear stress on an already pathologic aortic wall, leading to an isthmus aneurysm.

Coarctation management includes open surgical

approach or endovascular therapy, dependent on age at diagnosis.[3–5] In addition, concomitant congenital defects may complicate patient management. Open surgery is the first option for infants and children. Aortic stenting, when feasible, is particularly indicated in older children, adults, or in cases of recurrent coarctation. Endovascular treatment may be particularly advantageous in cases of non-discrete coarctation, and appears to be associated with low rates of re-coarctation.[3–5] However, the aortic anatomy may render this option unfeasible. When the aorta is tortuous or extremely narrow, or when calcification or stenotic aortic dilation is present, thoracic aortic endografting becomes a challenge. The presence of concomitant aortic aneurysm may confirm the selection of open surgical approach.[3–5]

Early or late development of aortic dilatation following coarctation repair has been described. However, to the best of our knowledge, primary coarctation-related isthmic aneurysm in an adult has never before been reported. The high risk of rupture and early death may explain the lack of reported instances of primary isthmic aneurysm, in the context of aortic coarctation, in adults.

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