Type B Interrupted Aortic Arch With a Very Large Right Subclavian Artery Aneurysm in an Adult

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ABSTRACT: Interruption of the aortic arch and right subclavian artery aneurysm is a rare congenital malformation. Survival in adults depends on the formation of collaterals to supply the descending aorta. The interruption of the aortic arch must be taken into account, particularly in patients with hypertension and weak pulses in the lower extremities. We present a case of aortic arch interruption and a right subclavian artery aneurysm in a woman who survived to adulthood.

KEYWORDS: Aortic arch syndromes, subclavian artery, aneurysm, adult

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Introduction

The interrupted aortic arch (IAA) is a rare cardiac malformation that accounts for 1.5% of congenital cardiac anomalies.[1] IAA associated with a right subclavian artery aneurysm is a very rare vascular malformation. The presentation of IAA in adulthood is extremely rare. In adulthood, the common complaint of patients with isolated IAA is hypertension,[2] but this is a rare cause of hypertension in adults. We describe the rare case of a 66-year-old woman who presented dyspnea and was later diagnosed with IAA and a right subclavian artery aneurysm. To our knowledge, no cases have been previously reported in the literature of such a huge right subclavian artery aneurysm and interrupted aortic arch.

Case Report

A 66-year-old woman with the main complaint of exertion dyspnea for 6 months. She became increasingly symptomatic about 30 days ago. She had a history of hypertension for several years and had been treated with Captopril. She was a nonsmoker with no family history of cardiac disease. However, she had not undergone a complete evaluation of the etiology of hypertension. On physical examination, her arterial blood pressure was 160/90 mmHg in the right upper extremity and 110/60 mmHg in the left upper extremity. Femoral and popliteal pulses were weak. The breath sounds were clear in auscultation. Systemic examination did not reveal other abnormalities. Chest radiography showed a huge intrathoracic mass in the upper lobe of the right lung (Figure 1). Routine laboratory tests were within normal limits. Transthoracic echocardiography showed concentric left ventricle hypertrophy and mild aortic regurgitation. Computed tomography angiography (CTA) showed an interruption of the aortic arch between the left subclavian artery and the left common carotid artery (Figure 2) and intense collateral circulation and a saccular aneurysm in the subclavian artery. The aneurysm measured 76 × 64 mm with atherosclerosis change and mural calcification (Figure 3). The patient was scheduled for surgical correction but refused surgery due to a COVID-19 infection. She underwent conservative therapy with medications to reduce hypertension. Her condition has been stable for 12 months of follow-up.

Discussion

IAA is extremely rare in adults and only a limited number of cases have been reported in the literature. IAA is a rare congenital malformation that occurs with a frequency of 3 cases per million live births.[3] IAA is defined by an interruption in anatomical continuity between the ascending and descending aorta. If IAA patients have an established effective collateral circulation, they can survive into adulthood. However, IAA in adults is extremely rare, with few cases reported.[4] It is typically found shortly after birth, and without surgery, it is likely to be fatal. Unless treated, the average survival of 90% of infants is 4 days.[5] In most cases, IAA is associated with other congenital cardiac malformations, such as patent ductus arteriosus, ventricular septal defect, and bicuspid aortic valve. Rarely is IAA an isolated finding. The classification made by Celoria and Patton[6] is the most widely used, based on the position of interruption that can be distal to the left subclavian artery (type A), between the left common carotid and left subclavian arteries (type B), and between the innominate and left common carotid arteries (type C). Type B is the most common presentation in infants and type A is the most common in adults.[7] The most common presentation in adults is hypertension refractory to medical therapy (70%), claudication (13%), and aortic insufficiency (10%).[8] In adults, the symptom ranges from asymptomatic to heart failure. Most patients had refractory hypertension since adolescence. Less symptomatic patients are often diagnosed incidentally. The most important

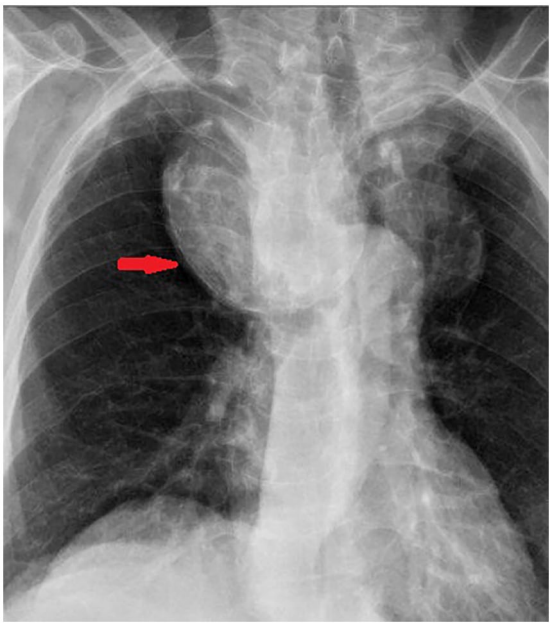

Figure 1. Chest radiogram showing a huge mass (red arrow).

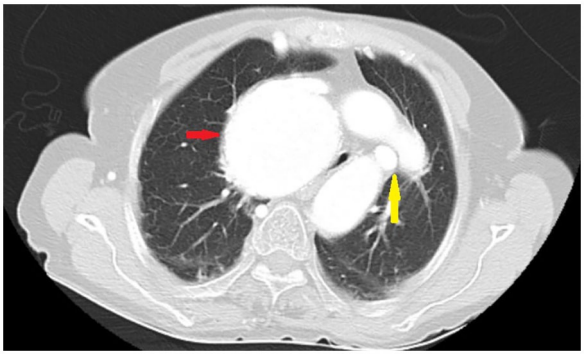

Figure 2. CTA showing type B interrupted aortic arch (yellow arrow) and right subclavian artery aneurysm (Red arrow).

aspect of the diagnosis is a careful physical examination, such as a measurement of blood pressure in the 4 limbs. CTA and echocardiography are recommended for diagnosis.[9] Angiography is necessary if CTA cannot distinguish between IAA and coarctation. Magnetic resonance angiography has been described in the evaluation of cardiovascular anatomy and coexisting cardiac anomalies, so it can play a complementary role in diagnosis. The main treatment for IAA in adults is surgical correction. When collateral circulation is abundant, surgery can be difficult due to a high risk of bleeding.[3] The prognosis is excellent after surgery. For adult patients with IAA and without symptoms, in addition to hypertension with sufficient collateral circulation, surgical intervention may not be the best option and antihypertensive drugs appears to be adequate and safe.[10] The average age of IAA diagnosis in adults is 39.4 years and most of them are men (74%).[11]

Subclavian artery aneurysm (SAA) is very rare and occurs in <1% of the population. The most common cause of SAA is

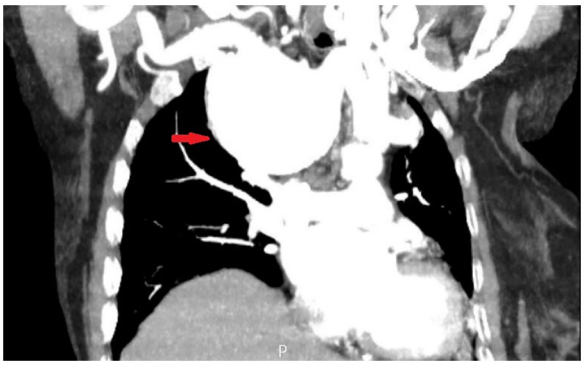

Figure 3. CTA showing huge right subclavian artery aneurysm (red arrow).

atherosclerosis. The other cause is trauma, infection, aortitis syndrome,[12] and congenital arterial disease.[13] Most patients with SAA are asymptomatic. If they became symptomatic, they may be related to distal embolization, rupture, and thrombosis. Other symptoms include dysphagia, dyspnea, and pulsatile mass. Subclavian artery aneurysms should be considered in the differential diagnosis of any chest mass noted on a chest radiograph. Doppler ultrasound and thoracic CTA scan are useful for diagnosis. According to the anatomical conditions, size of the aneurysm, and etiology of the aneurysm, open surgery, or endovascular repair can be used to treat SAA. Spontaneous rupture, thrombosis, and embolization are common risks associated with subclavian artery aneurysms, so surgical repair is highly recommended. Surgery technique is very important. During the operation, it is generally recommended to expose proximal and distal ends of the subclavian artery first, and then exposure of aneurysm, proximal and distal tract control, incise the anterior wall of the aneurysm, ligate the bleeding artery, and transect proximal and distal arteries. Finally, that perform artificial vessel graft in comparative diameter. Surgical complications consist of laryngeal nerve palsy, upper limb ischemia, chylothorax, cardiac complications, graft occlusion, and brachial plexus injuries.[14]

Conclusion

IAA is very uncommon in adults. It usually occurs along with other heart problems. Hypertension is an important public health issue. IAA may have been missed in older individuals with hypertension. Appropriate diagnosis is important in reducing complications related to hypertension. A careful physical examination plays a central role in the diagnosis of hypertension. This case report highlights the importance of considering IAA in the etiology of hypertension.

Author Contributions

All authors reviewed and approved the final manuscript.

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Informed Consent

This patient gave their written consent for the publication of this report.

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