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Diagnostic Imaging

Interrupted aortic arch diagnosis by computed tomography angiography and 3-D reconstruction: A case report

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ABSTRACT

Interrupted aortic arch is an extremely rare congenital malformation representing about 1% of congenital heart disease. Early symptoms usually occur early in the neonatal period and clinical deterioration is often rapid and long-term prognosis is limited. Nonetheless, this condition has been identified later in adult life in rare cases. We report a case in an adult male with absence of hypertension history and no further cardiac compromise, with a severe posterior chest pain alongside dyspnea and sweating. Computed tomography angiography revealed interrupted aortic arch type A, bivalve aorta, hemopericardium, aortic dissection Stanford A, and important collateral circulation.

Introduction

Interrupted aortic arch (IAA) is an extremely rare congenital malformation representing about 1% of congenital heart diseases characterized by a complete lack of luminal continuity between the ascending and descending aorta [1,2]. Early symptoms usually occur in the neonatal period, and clinical deterioration is often rapid with limited long-term prognosis. Classification of the compromise is distributed in 3 anatomic types (ABC); type A is located just beyond the left subclavian artery (79%); type B, between the left carotid artery and the left subclavian artery (16%); and type C, between the innominate artery and the left carotid artery (3%) [3].

Clinical presentation of IAA in adults varies from absence of symptoms to hypertension, headache, malaise, differential blood pressure between arms and legs, claudication, limb swelling, and congestive heart failure. Most patients have refractory hypertension since adolescence or early adulthood [4,5]. Here we report a case in an adult male with absence of hypertension history and no further cardiac compromise, who presented with hemopericardium and aortic dissection Stanford A, in whom computed tomography angiography (CTA) aid in the diagnosis of an IAA type A.

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Case report

A 55-year-old male was admitted to the hospital, with 7 hours of anterior and posterior chest pain (visual analog scale score: 8/10), alongside dyspnea and sweating. Past medical history revealed heavy smoking and no history of hypertension and claudication. On physical examination, his blood pressure was 110/64 mm Hg (mean 81 mm Hg), peripheral arteries were palpable and pulses on lower extremities were decreased. Electrocardiogram with a heart rate of 63 bpm was normal at the time. Chest x-ray revealed mediastinal widening of > 8 cm without pleural effusion, and troponin test was positive. Initial management with dual antiplatelet therapy, morphine, atorvastatin, and beta-blocker was established, and aortic dissection was suspected.

CTA revealed IAA type A (Fig. 1), bivalve aorta, hemopericardium (Figs. 2-3), aortic dissection Stanford A (Figs. 3-4), and important collateral circulation (Fig. 5). Patient was referred to the coronary intensive care unit and cardiothoracic surgery service for emergency surgical correction (tube graft) was performed. Patient presented a torpid clinical evolution and died within 5 days.

Discussion

IAA is a rare cardiovascular disease, first described by Stedeile in 1778, accounting for less than 1% of all cases of congenital heart disease [6–8]. IAA is primarily considered to be a diagnosis of infancy. It might be associated with other anomalies including ventricular septal defects, single ventricle, truncus arteriosus, transposition of the great arteries, valvular abnormalities (eg, bicuspid aortic valve, aortic or mitral stenosis, etc.), DiGeorge Syndrome, among others [7–9]. With anomaly it is highly unusual for a patient to survive without surgical interventional, because there would be no path for the blood to leave the heart and enter the systemic circulation [3]. Nonetheless, this condition has been identified later in adult life in

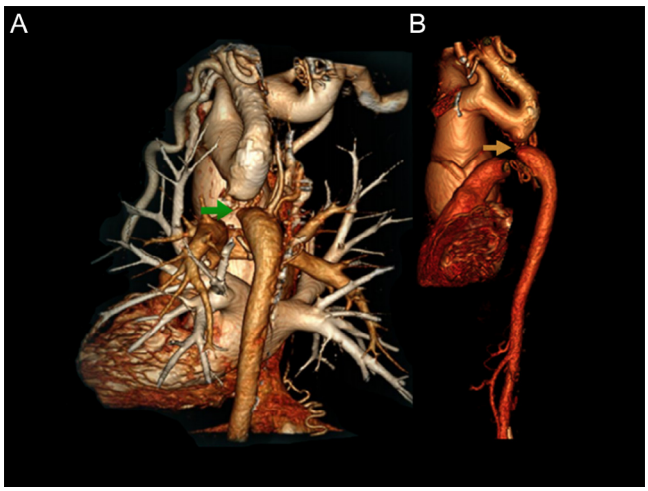

Fig. 1 – (A and B) Interrupted aortic arch type A: 3D reconstruction.

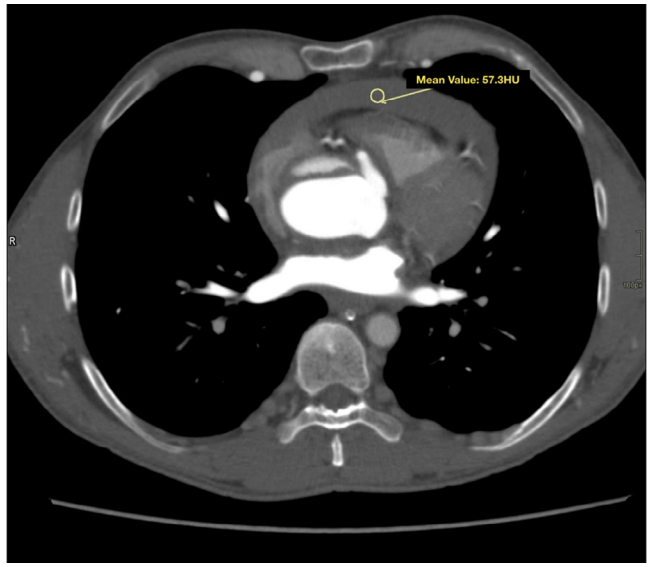

Fig. 2 – Region of interest on the hemopericardium.

rare cases [5,10–14]. Patients with isolated IAA may survive until adulthood due to the development of significant collateral circulation ensuring the maintenance of a blood flow to the distal aorta [7,15,16], as in our patient, who had important collateral circulation and had an associated bivalve aorta observed on the CTA.

About 37 cases of IAA in adults have been reported over the past 40 years [3], which has led to a better identification and description of this condition in adulthood. There have been some differences identified between neonatal IAA and adult IAA: in infants, IAA is usually associated with other congenital

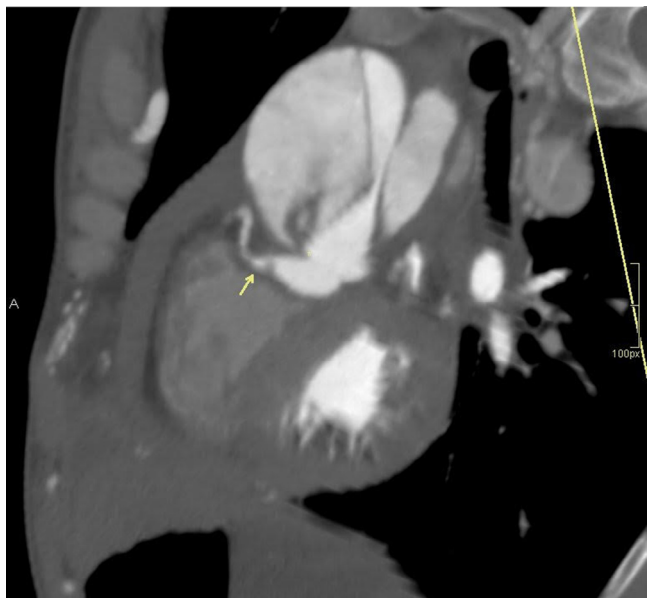

Fig. 3 – Modified 3-chamber thick slab maximum intensity projection showing the origin of the dissection flap immediately above to the origin of the right coronary artery without compromising it (arrow) and hemopericardium.

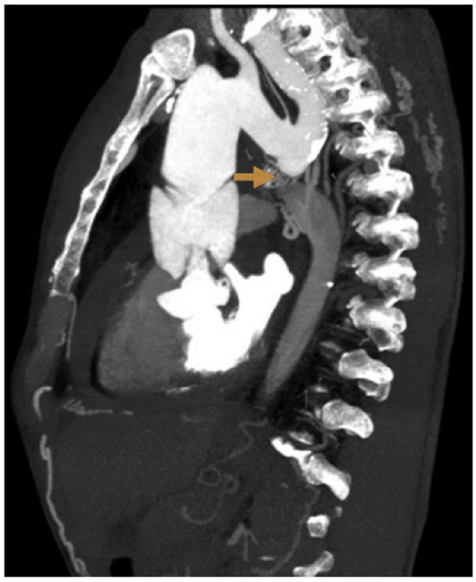

Fig. 4 – Projection of maximum intensity in the multidetector technique computed tomography showing type A interruption of aortic arch and aortic dissection comprising the ascending aorta.

cardiovascular abnormalities, whereas in adults it is usually isolated; furthermore, in adults, type A IAA appears to be much more common in comparison with neonatal IAA, were type B and type C are more prevalent [3,10,15]. Some hypotheses have been proposed for these differences. First, patients with type A interruptions are more likely to have adequate collateral flow to prevent severe symptoms. Second, patients with asymptomatic type B and C IAA have disparate upper extremities blood pressures, making diagnosis in childhood more common; and third, a subset of type A interruption may in fact represent the end point of a progressively narrowed aortic coarctation [3]. This latter point could potentially mean that the adult form of IAA is altogether different from the neonatal form [3,10].

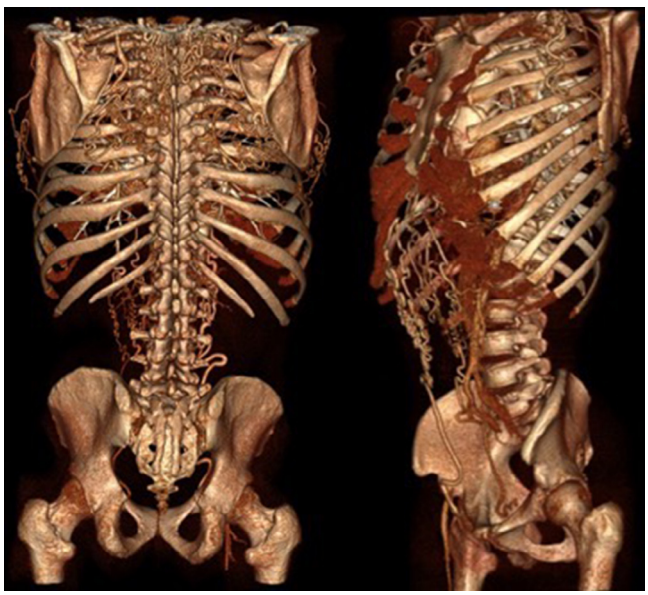

Fig. 5 – 3D Reconstruction of the computed tomography showing important collateral circulation.

Gordon et al. [3] published a review of IAA in the adults, describing the 37 cases reported in the literature up until 2010. The average age of the cases reviewed was 39.4 years (range 18-72 years), and it was more commonly diagnosed among men (74%) than women (26%); this information is consistent with the patient from our case report. Regarding clinical presentation, most patients had refractory hypertension as their presenting symptom, followed by claudication, congestive heart failure, and aortic insufficiency [3]. Chest pain in our patient corresponded to aortic dissection and hemopericardium; associations of these serious complications have been reported in patients with bivalve aorta (>50 years; 17.4% [95% confidence interval 2.9%-53.6%]) with or without IAA [17]. Chest pain has also been reported in patients with IAA and hypertensive crisis [11,14], a different type of IAA [18], or acute coronary syndrome [19].

Diagnosis assessment of IAA by CT or magnetic resonance imaging can easily demonstrate morphologic features of IAA and the potential complex associated findings because of their multiplanar capabilities, which facilitates the understanding of the anomaly and its anatomic relationships [7,20,21]. Furthermore, the multidetector CT have additional advantages over echocardiogram and magnetic resonance imaging, including short scanning timing, resulting in diminished sedation requirements, higher spatial resolution, and the simultaneous evaluation of the airway and lungs [7,20]. Additionally, the use of diagnostic imaging can also aid in the differentiation of IAA from coarctation of the aorta [9]. In the case of our patient, the use of CTA assisted not only in the visualization of the morphologic features of the IAA in the patient, but could also help in the surgical plan and approach used.

The main treatment for IAA in the adult is the same as in infants, which is the reconstruction of the aortic continuity to enable appropriate blood flow, usually by surgical means (eg, end-to-end anastomosis, graft interposition, or extra-anastomotic bypass) or by percutaneous approach in selected patients [3,9]. The main objective of the surgical intervention is to improve the patient's symptomatology and to prevent potentially fatal sequelae [3]. However, conventional surgical repair is typically a challenge because of the extent of collateral circulation in adult patients [10]; therefore, the mortality rates can range from 15% to 20% [22]. Unfortunately, even though our patient had a surgical intervention to correct the aortic arch, he died during the postoperative period within a week. Mortality in our case report may be secondary to the life-threatening complications present at initial presentation (aortic dissection plus hemopericardium), which might indicate that the initial clinical presentation of IAA in adults is an important factor in patient survival.

Conclusions

Although exceedingly rare, we report a 55-year-old male with IAA type A with no relevant past medical history. The CTA was

crucial to confirm the diagnosis of IAA and in illustrating the collaterals.

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