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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 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

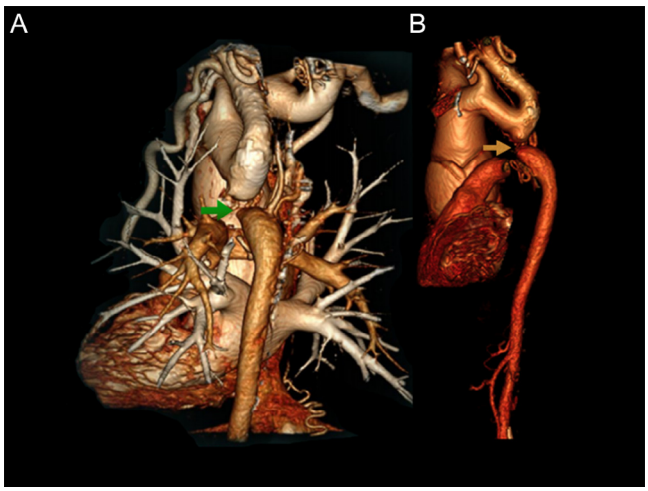

Fig. 1 – (A and B) Interrupted aortic arch type A: 3D reconstruction.

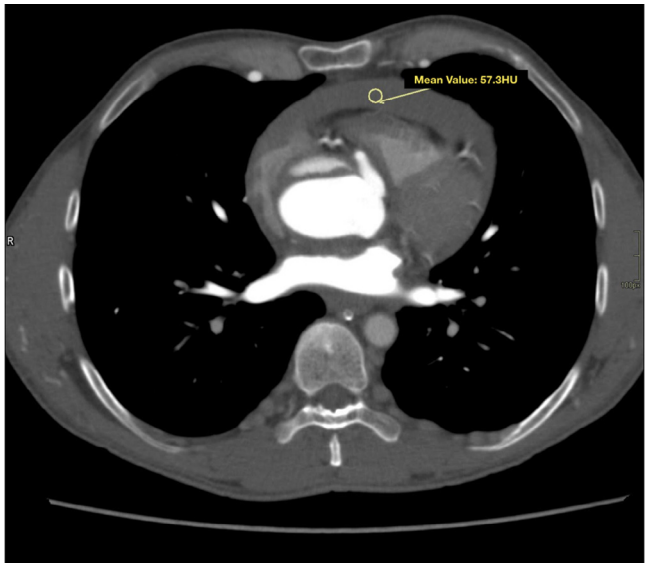

Fig. 2 – Region of interest on the hemopericardium.

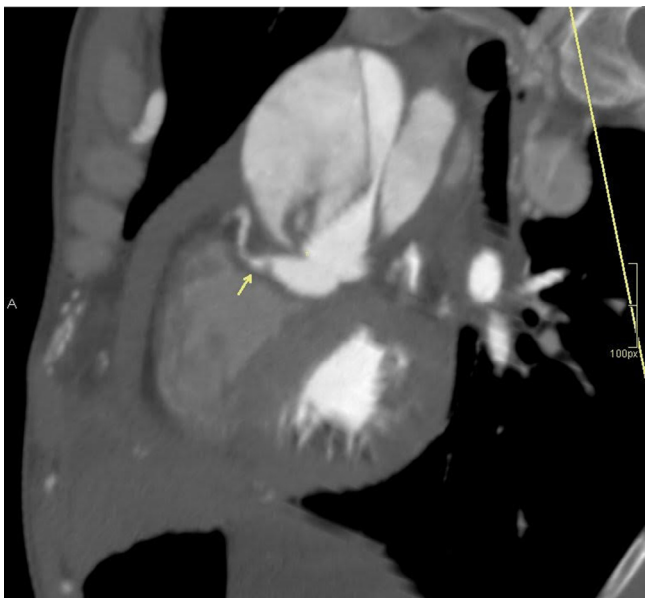

Fig. 3 – Modified 3-chamber thick slab maximum intensity projection showing the origin of the dissection flap immediately above to the origin of the right coronary artery without compromising it (arrow) and hemopericardium.