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Case Report

Interrupted aortic arch with post-interruption aneurysm and bicuspid aortic valve in an adult: a case report and literature review

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ABSTRACT

Interrupted aortic arch in adults is rare with a limited number of reported cases. We describe a case of a 53-year-old woman with interrupted aortic arch, bicuspid aortic valve, and post-interruption saccular aneurysm of the aorta. To our knowledge, this is only the second report of an adult patient with all 3 abnormalities. We also review the literature on this unusual condition and discuss its relationship with coarctation of the aorta.

Case report

A 53-year-old woman with medical history of hypertension presented to the medical clinic with chief complaints of paroxysmal palpitations and shortness of breath. Associated complaints included decreased exercise tolerance and bilateral lower extremity claudication. She denied chest pain. She was taking metoprolol and furosemide for management of her hypertension. On physical examination, her systolic blood pressure was approximately 145 mm Hg in both upper extremities but diminished to 77 mm Hg and 92 mm Hg at the right and the left dorsalis pedis, respectively. Because of clinical suspicion for coarctation or other cause of obstruction of the aorta, computed tomography (CT) angiography of the chest and the abdomen was performed. The CT study revealed interruption of the descending thoracic aorta just distal to the left subclavian artery (Fig. 1A). The interruption consisted of a thick septum at the aortic isthmus. The internal mammary arteries, intercostal arteries, and other intrathoracic arterial branches were enlarged providing collateral flow to the descending thoracic aorta distal to the interruption (Fig. 1B). A saccular aneurysm of the descending thoracic aorta just distal to the interruption measuring 2.0 cm in diameter was also present (Figs. 1C and D). Although the CT study was not

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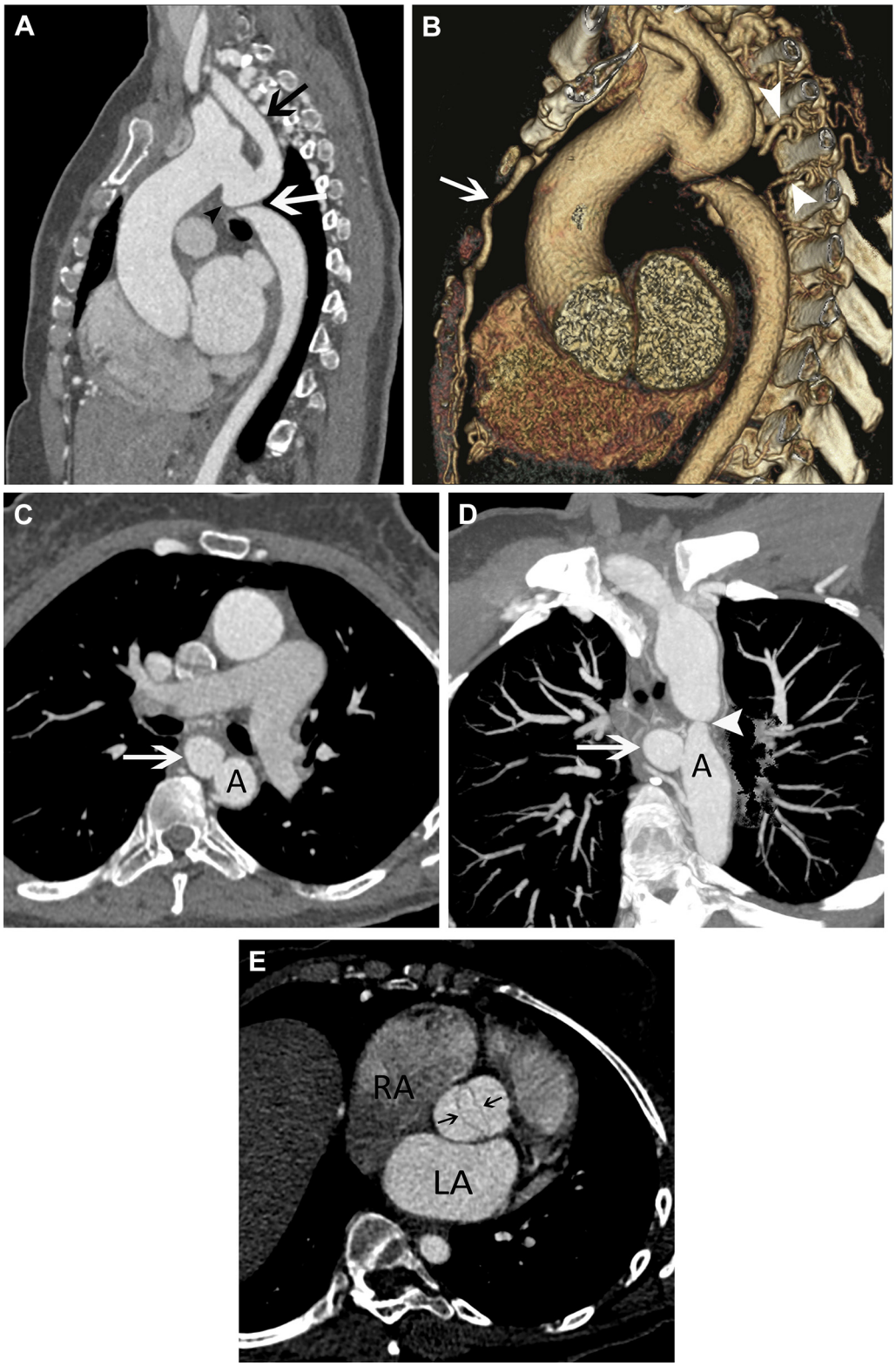

Fig. 1 – Images from CT angiogram in a 53-year-old woman presenting with paroxysmal palpitations and shortness of breath who was subsequently diagnosed with type A interruption of the aortic arch. (A) Reformatted oblique sagittal image of the thoracic aorta shows interruption of the aorta (white arrow) just distal to the origin of the left subclavian artery (black arrow). Note the maintenance of curvature of the ascending aorta and the presence of a small portion of the distal arch (arrowhead) just distal to the origin of the subclavian artery. (B) Volume-rendered image from a left lateral projection depicts the interruption. Note the dilated internal mammary artery (white arrow) and dilated intercostal arteries (arrowheads). (C) Axial image shows saccular aneurysm (arrow) extending from the descending thoracic aorta (marked A). (D) Reformatted oblique coronal image shows the saccular aneurysm (white arrow) extending from proximal descending thoracic aorta (marked A) just distal to the interruption (white arrowhead). (E) Reformatted view through the plane of the aortic valve shows bicuspid aortic valve morphology (arrows). LA, left atrium; RA, right atrium.

electrocardiogram-gated, reformatted views suggested bicuspid aortic valve morphology (Fig. 1E), and a bicuspid aortic valve was confirmed on subsequent echocardiography. At the time of this report, the patient was uninsured and awaiting approval of enrollment in an insurance plan before further management.

Discussion

Classification and presentation in adults

First classified by Celoria and Patton [1], interrupted aortic arch (IAA) has been divided into 3 types based on the location of the interruption: discontinuity distal to the left subclavian artery is classified as type A, discontinuity between the left common carotid and the left subclavian artery is type B, and discontinuity between the innominate and left common carotid artery is classified as type C.

In neonates, type B is most common representing about 53% of the cases, followed by type A (43%) and type C (4%). Type B IAA is strongly associated with DiGeorge syndrome and chromosome 22q11.2 deletion. More than 98% of infants with IAA have associated congenital heart disease such as patent ductus arteriosus, truncus arteriosus, ventricular septal defects, and aorticopulmonary window [2]. If not surgically treated, infants may die from biventricular heart failure, pulmonary edema, renal failure, metabolic acidosis, and other complications [2].

The presentation of IAA in adulthood differs markedly. In a review of 38 cases of IAA in patients aged older than 18 years, 79% of the patients were found to have type A, followed by 16% with type B and 3% with type C. The average age of diagnosis for the adults with IAA in this review was 39.4 years (range, 18-72 years) and most (74%) patients were men [3]. Most adults with IAA do not have associated cardiovascular shunt lesions and typically have mild symptoms at presentation [4]. Hypertension refractory to medical management is seen in 70% of the patients. Other symptoms include claudication (13%), aortic insufficiency (10%), and congestive heart failure (6%). About 13% of the patients have life-threatening complications at presentation [3]. Almost always, the blood pressure in the lower extremities is markedly diminished compared with that in the upper extremities, and the gradient across the interruption can vary from 50 mm Hg to 130 mm Hg [2,5].

Interrupted aortic arch vs severe coarctation

The differences mentioned previously suggest that IAA in infants and adults have differing etiologies. It has been postulated that type B IAA may be caused by defect in formation of the third aortic arch during fetal development and that type C may be caused by defects in the formation of the third and fourth aortic arches. However, type A IAA may be the result of regression or atrophy of a previously existing segment of aortic arch between the ductus arteriosus and left subclavian artery comparable with coarctation of the aorta as this segment can be narrowed, atretic, replaced by a fibrous band, or completely absent [1].

Coarctation of the aorta is thought to occur at the time of ductal closure from contraction and fibrosis of anomalous circumferential fibroductal tissue that pulls the posterior aortic shelf toward the contralateral wall. This process, in its most severe form, could result in focal aortic atresia or IAA [6]. The natural history of isolated IAA and coarctation is also similar with adult survival, late onset of symptoms, and a well-developed collateral circulation bypassing the obstruction [4,6]. Thus, some have postulated that predominance of type A IAA in adults is because of the progression of severe coarctation to complete occlusion [3]. Cases have also been reported of patients with known coarctation, which progressed to IAA [7]. According to Vriend et al. [8], the morphology of the aorta may also help to differentiate aortic atresia due to coarctation from true congenital type A IAA, that is, type A IAA present at birth. In coarctation, the ascending aorta retains its normal curvature and the distal arch extends slightly beyond the origin of the left subclavian artery, both of which are seen in our patient. In congenital type A IAA, the ascending aorta usually has a small caliber and a straight course to its branch vessels, whereas the arch does not extend past the left subclavian artery. Moreover, coarctation is also associated with increased incidence of aneurysms as the increased velocity of blood flow at the region of aortic narrowing could damage the vessel wall predisposing to aneurysm formation [9]. The close apposition of the blind ending segments of the aorta at the site of the occlusion, the maintenance of curvature of the ascending aorta, the extension of the aorta slightly beyond the left subclavian artery, and the presence of postcoarctation aneurysm strongly suggest that our case represents progression of severe coarctation.

Diagnosis

In patients with IAA, chest x-ray may be normal or may show cardiomegaly and/or rib notching [10]. Echocardiogram can be normal but may show left ventricular hypertrophy and/or a bicuspid aortic valve [2,10]. Angiography demonstrates complete occlusion of the aorta with a blind cul-de-sac of the proximal aorta and a blind ending at the superior margin of the descending aorta [2,10]. It also reveals the size and the branching pattern of the aortic arch vessels and the major collateral routes. Multislice CT provides high resolution 3-dimensional imaging of IAA and collateral vessels, which in combination with echocardiography (for detection of intracardiac lesions) is usually sufficient for preoperative planning in most cases [2]. Gadolinium-enhanced magnetic resonance angiography is also effective, but it is more expensive, time consuming, and more susceptible to artifacts than multislice CT [2].

Treatment and prognosis

The primary goal of surgical treatment of IAA is to correct the IAA and repair any other associated cardiovascular lesions preferably in a single-staged procedure [2]. Surgical options include extra-anatomic bypass, end-to-end anastomosis of both the ends of the IAA, graft interposition, and percutaneous stent placement. Extra-anatomic bypass is frequently

the preferred method because of concern of hemorrhage from well-developed collateral vessels [5]. About 8% of patients undergo percutaneous wire perforation through the septum at the IAA followed by placement of a covered stent. This can be attempted when the aortic wall is known to be intact on both sides of septum; however, access to cardiac surgery must be ensured in case of an emergency [7].

Surgery for correction of IAA is successful in most patients and postoperative mortality is rare. In many patients, hypertension gradually resolves after surgery, and antihypertensive medications can be discontinued [5,10]. Other symptoms such as claudication and paresthesia are also ameliorated after surgery [4]. Patients who refuse surgery (about 10% in one series) have been managed medically with antihypertensive drugs [3]. Nonetheless, close follow-up should be recommended.

Conclusion

We present the second case in the literature of an adult patient presenting with the combination of IAA, bicuspid aortic valve, and a post-interruption aneurysm. Although IAA can be fatal when presenting during infancy, patients without shunt lesions may survive to adulthood with adequate development of collateral blood supply to bypass the obstruction. The location of the occlusion, the morphology of the thoracic aorta, and the association with bicuspid aortic valve suggest that some cases of IAA in adults may represent the end result of severe aortic coarctation.

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