Discontinuity of the arch beyond the origin of the left subclavian artery in an adult: Interruption or coarctation?

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ABSTRACT

Congenital aortic anomalies are uncommon causes of secondary hypertension and are seldom suspected in the adult age group. We present a case of aortic interruption unexpectedly diagnosed on autopsy in a 38-year-old male who presented with cardiovascular collapse. Apart from interruption, a finding unique to our case was aneurysmal dilation of the proximal descending aorta just before the obstruction with thrombosis. We also attempt to review the literature for interrupted aortic arch in adults and clarify the nomenclature of interruption versus coarctation.

Keywords: Adult congenital heart disease, aortic coarctation, interrupted aortic arch, secondary hypertension, sudden cardiac death

INTRODUCTION

Among cardiovascular disorders, ischemic heart disease and stroke are the most important entities,¹ both of which have hypertension as the underlying modifiable risk factor. In most instances, the hypertension is essential although on occasion, it can be secondary. Congenital or acquired aortic anomalies are extremely uncommon causes of secondary hypertension. If these anomalies are not considered as potential causes of hypertension in young adults, then it is very easy to miss the diagnosis. We report the findings in a 38-year-old male who presented with cardiovascular collapse. At autopsy, we found discontinuity between the aortic arch and the descending thoracic aorta with aneurysmal dilation of the proximal descending aorta just before the obstruction with thrombosis.

CASE REPORT

A 38-year-old male became unconscious following sudden onset of giddiness. He was admitted to the emergency services of our tertiary care center “in extremis” and died despite resuscitation. He had a history of hypertension, diabetes mellitus, and left hemiparesis although no specific details were available. Apart from a random blood glucose level of 409 mg/dL, routine hematological and biochemical investigations were normal.

At autopsy, there was mild cardiomegaly (heart weight 300 gm) with moderate enlargement of the left ventricle. The aorta and pulmonary trunk were normally related although both the trunk and its branches were dilated and thin walled. The ascending aorta was aneurysmally dilated, having a maximum internal diameter of 2.8 cm. The aortic arch was left sided and contained a few calcified atheromatous plaques. The brachiocephalic arteries were similarly mildly atherosclerotic but showed no significant luminal stenosis. Just distal to the origin of the left subclavian artery [LSA, Figure 1], however, there was fusiform aneurysmal dilation of

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the proximal 4 cm of the descending thoracic aorta, which had a diameter of 3.5 cm. Opening the segment revealed occlusive laminated thrombus, with the wall being atherosclerotic and calcified. A distinct constriction was found just beyond the aneurysm, with serial longitudinal sections at this site showing lack of communication with the descending thoracic aorta, a finding confirmed by subsequent microscopy [Figure 2]. The distal segment was supplied by a sprout of four to five thin-walled arteries, one of which was of a large caliber. The origin of these collaterals, however, was unfortunately not noted at autopsy. Our diagnosis was interruption of the aortic arch (IAA) “Type A” interruption.

Further examination of the heart revealed bicuspid aortic valve (anterior to posterior type) [Figure 3a], moderate concentric left ventricular hypertrophy, and evidence of spontaneous closure of a preexisting trabecular muscular ventricular septal defect [Figure 3b]. The left coronary artery was dominant. All the coronary arteries were diffusely atherosclerotic, with multifocal critical stenosis [Figure 3c and d]. The left anterior descending artery, 3 cm from its origin, showed luminal occlusion for a length of 0.8 cm produced by a fresh organizing thrombus which had developed over an eroded atherosclerotic plaque [Figure 3e]. We presume this to be the cause of his sudden cardiovascular collapse.

Examination of the brain revealed mild frontotemporal atrophy, atheromatous changes in the vertebrobasilar system and lacunar infarcts in the basal ganglia, thalamus, hippocampus, andpons. There were histological features of hypertensive angioopathy, occasional microaneurysmal formation, and organized thrombus.

**DISCUSSION**

The atherosclerotic arterial disease noted in our patient can be explained on the basis of upper body hypertension, existing because of lack of communication between the brachiocephalic arteries arising from the aorta arch and the descending aortic segment. Such a finding in a young adult raises a question as to whether the lesion afflicting the aortic arch should be described as an aortic coarctation or IAA, a conundrum which has previously received attention.[2,3] In reality, these lesions exist as part of a continuum. Coarctation of the aorta is usually characterized by a discrete stenosis in the region of the insertion of the arterial duct to the aortic arch. On the other hand, aortic interruption represents the absence of anatomic and luminal continuity between adjacent segments of the aortic arch. Among the three types of interruption,[4] so-called “Type A”, or interruption at the isthmus, bears the closest resemblance anatomically to coarctation, since the discontinuity occurs immediately distal to the LSA. We have labeled our case as an example of interruption, since we found no evidence of continuity, either macroscopically or microscopically, between the proximal and distal segments of the aortic pathway. The thinned out aneurysmal wall was in close approximation to the media of the distal thoracic aorta. There was no poststenotic dilatation and no atherosclerosis in the distal segment. A discordant note, however, was our observation that the site of total obstruction was located well distal to the LSA with an aneurysmal segment of descending thoracic aorta placed in between, thus simulating coarctation.[2] This could be explained, nonetheless, on the basis of aneurysmal dilatation of the distal end of the interrupted arch.

Irrespective of whether the lesion in our patient represents coarctation or interruption, it is likely to have been present from birth. Despite no evidence of persistent patency of the arterial duct, the patient survived into adulthood. This is explained by the flow to the descending aortic segment provided by the collateral arteries observed during autopsy. This prompted us to review the 10-year data from 2007 to 2016 [Table 1] for such lesions in adults aged 18 years of age or greater. During our review, we came to appreciate from the imaging studies conducted that the anatomic mimicry between coarctation and interruption is further accentuated when there are complete obstruction and apparent loss of luminal continuity in the setting of coarctation. Unfortunately, the term “interruption” has also been used for these lesions, and these patients have undergone endovascular stenting.[5] The gold standard with which unequivocally to definitely identify interruption at the isthmus is direct visualization during surgery or examination of the pathological specimens. We did not, therefore, include unoperated cases of Type A interruption in our review, nor cases in which the type of interruption had not been specified in our
Among the published literature, we found 10 cases from 2007 to 2010 reviewed by John et al. and Gordon et al. when these authors reported their own cases, and these have been summarized in the table.

Among 29 confirmed cases of interruption identified in adults reported in the past 10 years, there were 21 males and 8 females, with ages ranging from 18 to 62 years, with a mean of 37.7 years. The interruption at the isthmus is more common in adults, being found in three-quarters of instances, in comparison to interruption beyond the origin of the left common carotid artery, or “Type B,” in children. The common presentations had been upper body hypertension, exertional dyspnea, and lower limb claudication. Neurological events, as seen in our case, were identified in four patients. Although our patient died of an acute coronary syndrome, angina had been described in only 2 additional patients. Noteworthy, associations had been bicuspid aortic valve in 6 patients, absent LSA in two patients, and the combination of a ventricular septal defect, patent arterial duct, and common arterial trunk in one patient. Looking at the nature of the septal defect in our case, it is possible that defects may have closed in several other cases, and hence, this would not have been detected by echocardiography or other imaging modalities. An unusual finding in our patient was an atherosclerotic aneurysm in the segment of the arch proximal to the interruption, which could be explained as a consequence of hypertension. Aneurysmal disease has been reported in six other patients with interruption, affecting the ascending aorta, related in all probability to bicuspid aortopathy, the arch segments distal to the interruption, the collateral arteries, and the LSA.

The treatment for IAA is the surgical restoration of aortic luminal continuity, facilitated by the use of several extra-anatomic techniques to bypass the lesion. It should always be remembered that lesions such as interruption or coarctation, especially in young hypertensive individuals, can often be diagnosed by careful palpation of the brachial and femoral pulses, and subsequent measurement of the blood pressures in the upper and lower limbs. It remains the case, nonetheless, that some patients continue to be missed, and as in our example, end with disastrous consequences. To clarify nomenclature, the distinguishing feature between interruption and coarctation is absence and the presence of luminal continuity, respectively. Cases where guide wires can be passed across through endovascular means cannot be aortic interruption. Lack of luminal continuity and presence of fibrous strand can be labeled as aortic arch atresia; however, since this term is ambiguous from an embryological and clinical stand point, its use can be avoided.

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Conflicts of interest
There are no conflicts of interest.
Table 1: Interruption of aortic arch in adults - review from 2007 to 2016

| Author and year of publication | Age/sex | Presenting symptoms | Interruption type | Associated anomalies |
|--------------------------------|---------|---------------------|-------------------|----------------------|
| John et al.[6] 2011 and Gordon et al.[7] 2011 | Age range of 19-62 years (mean age 37.2 years) 7 males, 3 females | Upper body hypertension (10); lower limb claudication (3); dyspnea (3), chest pain (2); palpitation (1); syncope (1); pedal edema (1); childhood detection of heart disease (2) | A (9); C (1) | BAV-2; VSD-1; PDA-1; absent LSA-1 |
| Henaine et al.[8] 2010 | 18 years, male | Upper body hypertension; loss of consciousness after consultation; childhood detection of interruption but did not come back for consultation | Nil | Nil |
| Verhaert et al.[9] 2010 | 28 years, female | Diagnosed as a complex congenital heart disease at the age of three years | B | Type I truncus arteriosus, PDA |
| John et al.[6] 2011 | 34 years, male | Upper body hypertension | A | BAV, ascending aortic aneurysm |
| John et al.[6] 2011 | 52 years, male | Upper body hypertension; Murmur since childhood | A | Nil |
| Gordon et al.[7] 2011 | 35 years, female | Upper body hypertension; lower limb claudication | A | Nil |
| Teskin et al.[10] 2011 | 44 years, male | Upper body hypertension; exertional dyspnea; palpitations; lower limb claudication | A | Nil |
| Yu et al.[11] 2011 | 53 years, male | Upper body hypertension, exertional dyspnea. polyarteritis | A | Aneurysm distal to interruption |
| Lafci et al.[12] 2012 | 34 years, male, smoker | Exertional dyspnea; palpitation; diastolic murmur detected at the age of 20 years | B | Aortic regurgitation |
| Erkanli et al.[13] 2012 | 29 years, female | Upper body hypertension; headache | B | Nil |
| Borgohain et al.[14] 2013 | 18 years, male | Upper body hypertension; lower limb claudication | A | BAV |
| Vural et al.[15] 2013 | 52 years, male | Upper body hypertension; headache; dizziness; neurological deficits | A | Nil |
| Davis et al.[16] 2013 | 57 years, male | Upper body hypertension; exertional dyspnea | A | BAV, aneurysm distal to interruption |
| Gowda et al.[17] 2014 | 21 years, male | Lower limb claudication | B | Aneurysm distal to interruption |
| Rybicka et al.[18] 2014 | 52 years, male | Congestive heart failure | B | BAV with AS |
| Mehrpooya et al.[19] 2014 | 59 years, male | Upper body hypertension; respiratory distress | A | AS (valve nature not mentioned) |
| Sasaki et al.[20] 2014 | 49 years, male | Upper body hypertension; past cerebrovascular accident | A | Aneurysms of thoracic and abdominal collaterals |
| Bai et al.[21] 2015 | 29 years, female | Upper body hypertension; subarachnoid hemorrhage; acute paraplegia | A | Right bundle branch block; thrombus proximal to interrupted segment |
| Oz et al.[22] 2016 | 24 years, male | Upper body hypertension | A | Left subclavian aneurysm |
| Abdoli et al.[23] 2016 | 34 years, male | Upper body hypertension | B | Absent proximal LSA |

BAV: Bicuspid aortic valve, VSD: Ventricular septal defect, PDA: Patent ductus arteriosus, LSA: Left subclavian artery, AS: Aortic stenosis

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