Case report

Coarctation of aorta presenting as systemic hypertension in a young adult: A case report

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Abstract

A 33-year-old male who was under investigation for young onset hypertension for nine years had weak lower limb pulse and radiofemoral delay on examination. Transthoracic echocardiogram revealed a bicuspid aortic valve, and a CT aortogram detected coarctation of the aorta at the junction of the aortic arch and descending thoracic aorta.

Coarctation of the aorta is a rare secondary cause of hypertension in young. Symptoms of presentation may vary depending on the site and severity of coarctation. But the commonest presentation is hypertension in the upper extremity of the body. It could be associated with structural heart disease; the commonest is the bicuspid aortic valve. Treatment options for coarctation of the aorta include balloon angioplasty and surgical repair. Lifelong follow-up is needed in both treatment modalities, even after successful correction.

Keywords: Young hypertension, Coarctation of aorta, Radio-femoral pulse delay

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Funding: None

Competing interest: None

Received: 16.11.2023 Accepted revised version: 24.05.2024 Published: 06.08.2024

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Introduction

Coarctation of the aorta is a rare congenital anomaly of the cardiovascular system, which causes narrowing of the aorta at any site along its cause, but the commonest site is immediately after the origin of the left subclavian artery [1,2]. This commonly presents as hypertension in childhood [1]. In addition, it could present as complications of hypertension, such as stroke or heart failure [3].

Here, we present a case of late diagnosis of coarctation of the aorta in a 33-year-old male due to the incidental finding of a weak bilateral femoral pulse.

Case presentation

A 33-year-old male was on treatment for hypertension for 9 years and was investigated for cause since 2015. He was first diagnosed with hypertension in 2015 when he presented with dizziness, on and off chest pain, and exertional dyspnoea. His blood pressure was 200/130
mmHg at first detection. Four groups of antihypertensives, including a diuretic, were needed to control his blood pressure at that time. Since then, he has been investigated for the cause of young hypertension. He did not have a history of episodic headache, blurred vision, palpitations, episodic flushing or heat intolerance, unexplained weight loss, or loose stools. He did not have a recent change of appearance of his body and no joint pains, skin rashes or recurrent oral ulcers. He never complained of any proximal muscle weakness. He had a history of recurrent urinary tract infections. But those were not complicated with pyelonephritis. There was no history of urinary calculi. He started smoking at 13 years of age (one per week) and was a heavy smoker for the last 16 years. He had smoked 1.6 pack years of cigarettes in the first ten years, and then it had been reduced to 0.7 pack years in the last 6 years. He has stopped smoking one month back. He consumes alcohol infrequently but does not use other illicit drugs.

A transthoracic echocardiogram done in 2015 revealed a bicuspid aortic valve with grade 1 aortic regurgitation with Ejection fraction > 60% and no pulmonary hypertension. Coarctation of aorta was not detected during the first echo or follow-up echocardiogram. ESR was 7mm/1st hour. Serum creatinine and Serum sodium were normal(137mmol/l), and serum potassium was low (2.5mmol/l). Urinary potassium excretion was 20mmol/l. An ultrasound scan revealed bilateral normal kidneys. Twenty-four-hour urinary vanillylmandelic acid (VMA) level was normal (3.7mg/24h). Non-contrast computer tomography (NCCT) of the kidney, ureter, and bladder did not reveal any abnormality. Initially, his blood pressure had been controlled with five antihypertensives. Which could be reduced to three antihypertensives later. He was followed up with serial echocardiogram yearly until this admission, but hypertension evaluation had not been continued. He did not have any limitations in activities during exertion except bilateral intermittent claudication when walking around 100 m for the last one to two months duration. He had no past history of ischemic heart disease and strokes and no significant family history other than his mother was diagnosed with hypertension in her late 50s. He was not on any long-term medication other than his routine antihypertensive drugs. Now he is on three antihypertensives, with that his blood pressure is partially controlled. He had two hospital admissions with hypertensive urgency during the last 6 years.

His height was 174 cm, and his weight was 72kg (BMI 23.8). There was no disproportionate growth of the upper and lower segments of the body. He was not pletoric. There were no features of acromegaly, no goitres, and no skin rashes. His pulse rate was 62 beats per minute, which is regular. His upper limb pulse volume was normal, but his bilateral femoral, popliteal, and dorsalis pedis pulse volume was low. There was no radio radial delay. However, there was a significant radio femoral delay. His blood pressure was 150/80mmHg, which was equal in both arms, and his jugular venous pressure was not elevated. The apex beat was normal at the fifth intercostal space midclavicular line. His first heart sound was normal, and his second heart sound was loud. There was an early diastolic murmur, which was best heard in the aortic area and on the left lower sternal border. There was no parasternal heave, and the lungs were clear to auscultate. His abdominal and neurological examination did not reveal any abnormality. The optic fundus shows silver wiring only.

In this admission, his full blood count, blood urea, serum creatinine, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), thyroid stimulating hormone (TSH), aspartate transaminase (AST), alanine transaminase (ALT), serum magnesium, serum calcium, serum sodium and chloride levels were normal. However, serum potassium was at a lower margin (3.5mmol/l). Metabolic alkalosis was found in arterial blood gas analysis. His serum aldosterone level was 14.04 ng/dl (3-16), and his serum renin level was 76.17mIU/l (3.11-41.2). Renin aldosterone ratio was 0.18 (< 3.7). His renal artery duplex did not reveal any renal artery stenosis, and his bilateral kidneys appeared normal. A chest X-ray was done (Figure 1). That revealed bilateral rib notching. In CT aortogram (Figure 2), there was coarctation of the aorta at the junction of the aortic arch and descending thoracic aorta with multiple dilated intercostal collateral vessels. Repeat 2D echo cardiogram revealed ejection fraction > 60% with moderate left ventricular hypertrophy and bicuspid aortic valve with aortic regurgitation. There was post ductal coarctation. The peak aortic pressure gradient at coarctation was 62 mmHg, and the mean pressure gradient at coarctation was 32 mmHg. The patient was diagnosed to have moderate juxta ductal coarctation with sluggish flow in the descending aorta and bicuspid aortic valve with mild aortic regurgitation. He was referred to an interventional cardiologist at the National Hospital of Sri Lanka, Colombo, for balloon dilatation and is awaiting intervention.

Discussion

This young male with resistant young hypertension was found to have low volume femoral pulse, hypokalemia with metabolic alkalosis, high renin, marginally high aldosterone level and bilateral rib notching in chest x-ray. Coarctation of the aorta was identified in the 2D Echo and confirmed with a CT aortogram. the patient was referred for balloon angioplasty after assessing the aortic pressure gradient.
Hypertension in less than forty years of age is considered young hypertension, which could be secondary to a correctable cause most of the time. Coarctation of the aorta is one of the rare causes of young hypertension and resistant hypertension [4,5]. Usually, it is detected during childhood [1]. However, depending on the severity of narrowing and formation of collaterals, presenting age and symptoms may differ [5,6]. Even though narrowing can occur at different locations of the aorta including aortic arch, thoracic aorta, or abdominal aorta the commonest site of narrowing in coarctation is just after the origin of the left subclavian artery [2]. In our patient, coarctation was at the arch of the aorta and the junction of descending thoracic aorta. The commonest presentation of coarctation is hypertension in the upper part of the body, but it also can present as absent or weak femoral pulse, intermittent claudication, and left ventricular hypertrophy [1,3]. Our patient presented with resistant hypertension at the age of 24 years and later he developed reduced bilateral femoral pulse and intermittent claudication. When there is reduced blood flow to bilateral kidneys due to coarctation, that stimulates hypersecretion of renin. Our patient also had a high renin level. His aldosterone level was at upper margin of normal, which could be because he was on angiotensin receptor blocker. He had metabolic alkalosis with hypokalemia as well, which is a common finding in coarctation of aorta.

Coarctation of the aorta is associated with congenital heart diseases in 6-8% of patients [2]. It is commonly associated with the bicuspid aortic valve as in our patient. Our patient was detected to have a bicuspid aortic valve at first presentation with hypertension. But coarctation had not been detected at that time. In the presence of a bicuspid aortic valve, there could be systolic ejection click and an ejection systolic murmur at left upper sternal edge [2]. But our patient did not have that as he had aortic regurgitation. If the patient is female, she needs karyotyping because Turner’s syndrome can be associated with isolated coarctation of aorta [2]. Coarctation of aorta itself has a risk of infective endocarditis. In the presence of bicuspid aortic valve, infective endocarditis risk is further increased [7]. Unrepaired coarctation of aorta can be complicated by premature coronary artery disease, aortic dissection, or aortic aneurysm and strokes, commonly at 3rd and 4th decade of life. All those complications are due to uncontrolled hypertension [2].

Treatment of coarctation of the aorta could be either surgical or transcatheter-directed therapy. Surgical treatment includes excision of the coarctation segment and end-to-end anastomosis, commonly practised in small children and neonates [2]. Transcatheter-directed therapy includes balloon angioplasty and stent angioplasty, which are commonly done in adults [2]. Our patient was detected with coarctation of the aorta at the age of 33. So, he was referred for balloon angioplasty. There is a risk of re-coarctation in 10% of cases after surgical correction. That needs balloon angioplasty later. Even after balloon angioplasty, there is a high risk of aortic aneurysm formation. So, regular follow-up is needed for a lifetime [2].

Figure 1: Arrows indicate rib notching in chest X-ray

Figure 2: Arrow indicates the coarctation of aorta in CT Aortogram

Conclusion

All patients under 40 years of age with hypertension should be evaluated for secondary causes as most of them are treatable. Radio-femoral pulse delay is a simple but important physical sign that helps to suspect aortic coarctation.

Consent

Informed written consent was taken from the patient for writing his story and publishing photographs taken of his body parts, photographs of his X-ray and CT angiogram.
Timeline

- 2015/04: Diagnosis of Hypertension and 1st 2D Echo
- 2018/05: Hypertensive urgency
- 2023/05: Hypertensive Urgency
- 2023/07/14: Present Admission
- 2023/7/18: Chest X-Ray (Bilateral rib notching)
- 2023/7/21: CT Aortogram (coarctation of aorta detected)
- 2023/08/04: Renin Angiotensin Aldosterone ratio
- 2023/08/05: 2D Echo
- 2023/08/07: Measure aortic pressure gradient, Referred to interventional cardiologist
- 2023/9/3: Follow up arranged at NHSL
- 2023/9/19: Rental Artery duplex

References