Critical Reappraisal and Lessons from a Case of Coarctation of Aorta: Focus on basics

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ABSTRACT

Introduction: Coarctation of aorta (CoA) is one of the causes for refractory hypertension, and may remain undiagnosed till adulthood, if associated with obscure symptoms. Generally, such patients are treated for hypertension, and in few investigations may not reveal any specific features. We followed the principles of Enigma code to establish the diagnosis.

Case report: We describe a young patient with a long medical journey, approached multi-disciplinary specialties, before mid-aortic syndrome was diagnosed by us. We noticed a delayed development of a finding (presence of bicuspid valve) on echocardiogram, which were not obvious previously. Computed tomography aortogram showed a long segment coarctation arising 2.5 cm distal to the left subclavian artery, with multiple collaterals arising from bilateral intercostal arteries, left axillary, inferior epigastric artery. Coarctation stenting was performed with good outcome, without any pre- and post-operative complications.

Conclusion: Following every step of basics of clinical examination is essential throughout clinical practice. We emphasize focussing on basics of clinical examination to pick up pulseless diseases to prevent delay in diagnosis. Following the Enigma code helps in decoding the complex unsolved medical case mystery.

Keywords: Coarctation of Aorta; Enigma Code; Midaortic Syndrome; Pulseless Disease.

INTRODUCTION

Coarctation of aorta (CoA), a common congenital defect, categorised as simple and complex based on the absence or presence of other cardiac anomalies, respectively. The latter is diagnosed early, while the former may remain undetected till adolescence and adulthood. It is one of the obscure cause of secondary hypertension in ~0.2% of adults. Early diagnosis is decisive in tendering appropriate management to minimise associated complications and mortality.

We present the journey of a young patient with coarctation of aorta, who remained undiagnosed, misdiagnosed despite multi-specialty consultations.

CASE REPORT

A 17 year young patient presented with a history of (h/o) headache of 5years and diagnosed to be hypertensive since past three years. There was a long medical journey, from family physician to neurologist, nephrologist, cardiologist, psychiatrist and even gynaecologist, was on atenolol and cilnidipine before consulting us with a recent ultrasound (USG) abdomen and renal doppler report.

Previous laboratory reports were within normal limits. Patient was hypertensive (blood pressure (BP) 180/100), similar in both upper limbs, with no difference in bilateral radial pulse volume. Investigations for renal parenchymal disease were within normal range; renal doppler showed monomorphic wave (pulsus parvus et tardus) at the origin of both the renal arteries. With these reports leading to three possible causes for hypertension, (a) Takayasu arteritis (b) fibromuscular dysplasia (FMD), and (c) giant cell arteritis. Bilateral absent femoral pulse, a crucial sign led closer to the diagnosis, inching towards the diagnosis of CoA.

No detectable abnormalities were noted on inspection, palpation and percussion of the chest; there was a loud S2 (aortic component), but no obvious murmur in the aortic area. There was a positive Suzmaan's sign[1] (palpable collaterals in the back left interscapular area). These clinical findings were strongly in favour of CoA and we proceeded with further cardiac evaluation.

X-ray chest showed notching of ribs, a classical finding in favour of CoA. Lead I in electrocardiograph (ECG) had R wave of around 15 mm, so as S in aVR. Echocardiography showed left ventricular hypertrophy (LVH), with doppler evaluation in supra sternal view showed gradient of 88 mm Hg with diastolic tailing and presence of bicuspid valve, a new feature that was not reported in patient's previous reports. Computed tomography (CT) aortogram demonstrated a long segment (47 mm) stenosis of descending thoracic aorta, there was no renal artery stenosis (figs 1-4). There was no Berry aneurysm on CT brain angiogram. There was no oedema of the aortic wall oedema on magnetic resonance imaging.

Clinical signs, and imaging inputs prompted us to considered CoA and Takayasu arteritis as differential diagnosis. When analysed in toto from history to imaging, findings supported the diagnosis of Coarctation of aorta (table 1). Based on the position of coarctation, index case was diagnosed to have middle aortic syndrome.

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Parameters	Coarctation of aorta	Takayasu arteritis
History		
h/o fever, weight loss, night sweats	unlikely	possible
c/o claudication	Uncommon	Common
Positive Suzzmaan's sign	Common & Diagnostic	Unlikely
Echocardiography		
Presence of bicuspid	Likely	Unlikely
Presence of rib notching	Likely	Unlikely
Fundus examination		
Cork-screw appearance	likely	Unlikely
Laboratory investigations		
Normal CRP, ESR , MT test	Possible	Unlikely
Magnetic resonance imaging		
Aortic wall oedema and thickening	unlikely	Seen in Acute phase or arteritis
Parameters describes the findings noted in the index case		

Table-1: Comparison of differences between coarctation of aorta and Takayasu arteritis

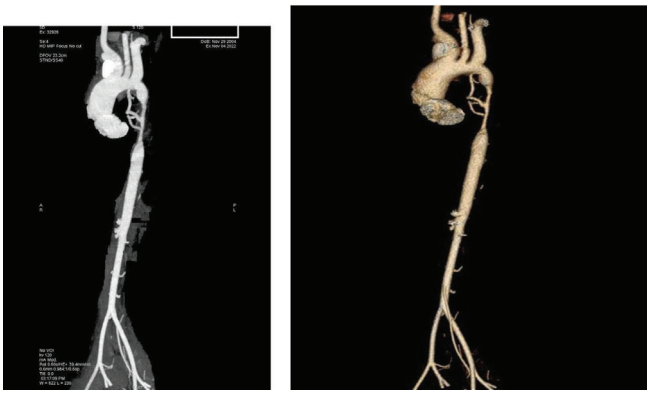


Figure-1: Computed tomography Aortogram showing a long segment, pre-coarctation segment and multiple collaterals.

Fig 1 showing long segment coractation arising 2.5 cm distal to the left subclavian artery, starting from the lower border of D5 vertebra to up to upper border of D8. Total coarctation segment length of 4.6 cm and tightest part with a diameter of 0.5 cm. Pre coarctatation segment descending aorta diameter of 1.2 cm and aorta diaphragm of 1.4 cm. Multiple collaterals are noted arising from bilateral intercostal arteries, left axillary, inferior epigastric artery.

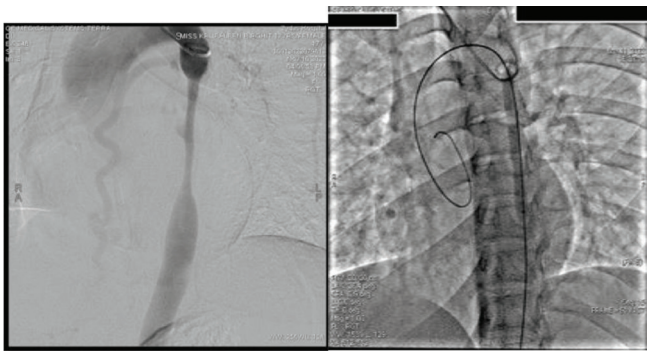
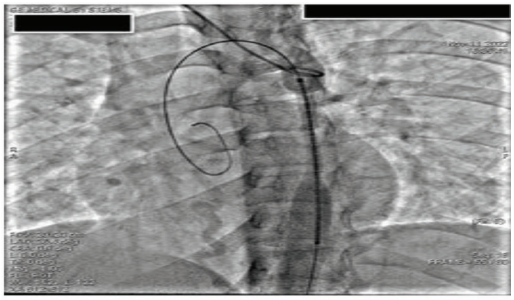


Figure-2: Descending aortogram showing a long segment coarctation and pre dilatation with 8x60 mm balloon.

DISCUSSION

Coarctation of aorta comprises of 6%-8% of congenital heart diseases and has a male preponderance. Clinical presentation depends on the age, with features of congestive heart failure predominating in the neonates and young infants. In



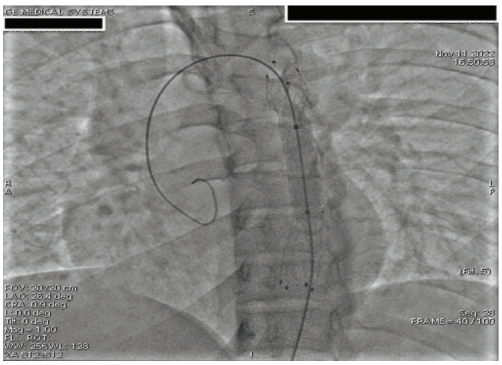


Figure-3: Positioning of 14X80 mm stent, Post dilatation with 12x60 mm balloon

adolescents, it may remain asymptomatic or may present with complaints of leg discomfort or pain while running (claudication type), headache or epistaxis.[2,3] Hypertension and presence of a heart murmur in young adults is the cause for CoA work-up.

Index case was young (17 years) with a diagnosis of secondary hypertension; presented with h/o headache of long duration (five years). Patient's previous multiple consultations did not establish the underlying pathology, hence, was subjected to a thorough basic general and systemic examination.

Pulse holds the key in diagnosis of CoA, which includes a characteristic clear-cut disparity between upper and lower extremity pulses and BP, which may be unobtainable in the legs, particularly when it is <20mm Hg, absent distal lower extremity pulses and diminished femoral pulse that lag behind the brachial pulse (radio-femoral delay). Our patient had absent bilateral femoral pulse, which Vigorous right

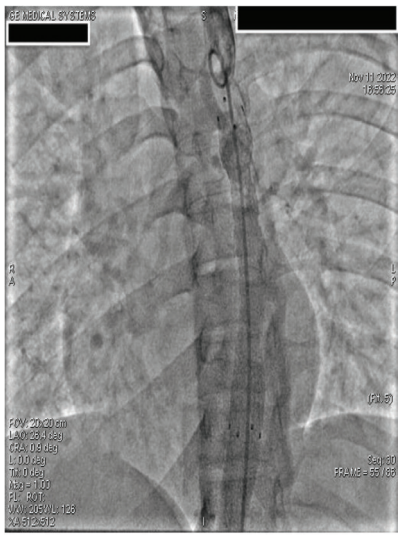


Figure-4: Post-stenting angiogram
Post-angiogram showing well expanded stent with no residual stenosis and no dissection distall

upper extremity pulse, with an exception if the coarctation involves the origin of the left subclavian artery, wherein diminished left arm pulse is possible. Both the femoral pulse were not palpable in the index case, hence, we cannot comment on the radio-femoral delay.

Presence of a bicuspid aortic valve (as noted in the presented case) yields an ejection click between the lower left sternal border and the apex. Audible systolic bruit over the middle left back and the upper left sternal border also a sign of CoA. Presence of a systolic ejection murmur in the upper right sternal border denotes aortic stenosis.

Identify, Break and Solve are the three basic principles of deciphering the enigma code, which the British-French intelligence used to decipher Morse coded radio messages of Germany. By and large the same rules can be applied to solve complex clinical situations in medical science. As a cardiologist, I followed the Enigma code, which made my job simple, just to decipher the codes and establish a final diagnosis.

1. **Identify** - young patient presenting with hypertension, indicating that the working diagnosis was secondary hypertension. Renal pathology was one of the cause in the list, but available reports ruled out chronic kidney disease (polycystic kidney), and adrenal pathology. Presence of monophonic waves an interesting finding on renal doppler signalled renal artery stenosis, Taksyasus arteritis,[4] FMD, or giant cell arteritis as possible differential diagnosis, and with echocardiograph showing only LVH, CoA was also considered a probable diagnosis.

2. **Break and regroup** : With all previous investigations in hand, which led to nowhere, we went back to the basics, following the Enigma model. Reworking the case for every possible clue to diagnosis was planned and executed. Blood pressure in right arm was 180/100 and almost the same in left arm in sitting position, indicating no significant difference between the two upper limbs.

We followed the very first basic of clinical examination, feeling the pulses; both carotid pulses were equal in volume. Clinching clue was absent bilateral femoral pulsations, which almost lead to the final diagnosis, yet a step away from final diagnosis.

Continued clinical examination of systemic examination of cardiovascular system, did not reveal much on inspection, palpation and percussion. A loud S^2 (aortic component) without any obvious murmur was a conspicuous sign. A murmur heard in left interscapular area and a positive Suzmaan's sign were in favour of a probable diagnosis of coarctation of aorta. Non-invasive BP monitoring was not possible due to absent pulse and when pulse pressure is < 20 mm hg (as in femoral artery) as in the indexed case, tactile feeling of pressure is not possible. BP was 220/110 in the ascending aorta and 120/100 Hg in femoral as recorded in catheterization laboratory .

ECG may show high left ventricular voltage (HLVV) indicating left ventricular hypertrophy in CoA. ECG of the index case demonstrated LVH as per voltage criteria; R wave in lead I (4 mm) and s wave in aVR (15 mm). ECG may also demonstrate left ventricular "strain" pattern (variable, but may be noticeable in the lateral leads (I, aVL, V5-6)), asymmetric T-wave inversion originating from a depressed ST segment and right ventricular hypertrophy (less common), which however not reported in the index case.

Transthoracic echocardiography is diagnostic while cardiac magnetic resonance imaging (MRI) and cardiac computed tomography help in further evaluation. Echocardiography, revealed gradient with diastolic tailing, which led me another step ahead. The diagnosis pathway suggested strongly and supported the diagnosis of CoA. There was no presence of neither renal artery stenosis nor Berry aneurysm CT aortogram with CT brain angiogram but a long segment stenosis of descending thoracic aorta was noted, very unusual for adult coarctation, but can be expected with Takayasu arteritis (type 3b variant), very similar to mid aortic syndrome.[4, 5] Similar finding of long segment stenosis of descending thoracic aorta was reported by transthoracic echocardiogram in middle aortic syndrome in a 14 year old female with shorter h/o dyspnoea, fatigue (2 months) and refractory hypertension (6 months).[6] Similar observation was reported in a 9 year old child by Truong and Fonseca.[7] Mural enhancement and wall thickening are a possible feature on cardiac MRI in Acute Takayasu arteritis, unlikely in CoA, which was not seen in our patient as well.[10]

Two possible diagnosis of coarctation of aorta and Takayasu arteritis (high middle aortic or upper thoracic aorta syndrome) were considered for further workup. Fibromuscular dysplasia (FMD) and giant cell arteritis were ruled out as age was not in favour of the diagnosis.

3. **Solve** – Narrowing down to two differential diagnosis, it was the time to solve the enigma. Putting together the soft clues and reanalysing helped in arriving at final diagnosis of CoA.

Based on the position of coarctation and symptoms collectively, a diagnosis of middle aortic syndrome was made. Additional involvement of renal other abdominal

vessels are as frequent as in 2/3rd of cases[8] and can result in secondary hypertension[5]; however, we ruled out renal artery pathology in the index case. Middle aortic syndrome can be congenital or acquired, and our patient had simple CoA as there was no intra cardiac lesion except bicuspid aortic valve. Treatment options include endovascular balloon angioplasty, stenting, surgery, we considered the latte two and both were explained to the family, allowing them to select one.

Endovascular-stenting of coarctation is the most followed technique with good results. But with the index case, there were problems such as long lesion requiring long and sturdy stent, the cost of the procedure, which put the question on affordability. Atrium stent and CP stent are expensive and not covered under the state sponsored health insurance plan used by the patient to avail the treatment, a common situation with most of these patients, a hindrance in availing the access to the much needed treatment. However, covered stents are not free from disadvantages i.e. large access sheath in small sized patients and risk of spinal cord ischemia with long stents, which makes careful selection of patient for better outcome.

Surgery is a feasible option but cosmetically, no young female would prefer to have scar on any part of her chest. Additionally, hypoplastic arch and juxta subclavian ductus arteriosus (DA), carries a high risk and bleeding from collateral circulation is a challenge, but is a possible option.

Coarctation stenting was done with 14 x80 mm stent during November 2022, yielded good end result without any pre-and post-operative complications. Pressure was well controlled and all peripheral pulse were palpable without delay.

Pre-procedure gradient was 100 mm hg across coarctation, which reduced to ~ 20 mm Hg after the procedure. Aggressive balloon dilatation with 1:1 balloon was avoided; as covered stent was not available we stooped at this point. At month 1 follow-up, a pressure gradient of 10 mm Hg was recorded on echocardiogram. After three months of post-stenting, the patient is stable clinically, with a BP of 140/90 mm Hg on two drugs (telmisartan and amlodipine) and eight mm Hg gradient on echocardiogram .As available reports are promising with no mortality in long-term follow-up,[9]we suggested long term follow-up, and the patient is being monitored periodically.

CONCLUSION

This case reminds us not to forget our basics of clinical examination. Good clinical examination and history taking are vital to solve such enigmatic cases. Pulseless diseases may be missed, if all palpable pulses are not palpated. A careful clinical correlation of pathological and radiological findings are needed. Following the Enigma code helps in decoding the complex unsolved medical case mystery.

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