Coarctation of aorta with aberrant right subclavian artery presenting as Dysphagia lusoria and the impact of Bicuspid Aortic valve

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Abstract:
The coexistence of coarctation of aorta (CoA), Bicuspid aortic valve (BAV), severe aortic stenosis and aberrant subclavian artery is rare. The clinical significance of a bicuspid valve in patients with coarctation is not well established. The aberrant right subclavian artery with a retroesophageal course is the cause of dysphagia lusoria. The clinical examination of both upper and lower limb pulses is important at any age. The lesions are intimately related and the management is a real surgical challenge.

Keyword: Coarctation aorta, aberrant subclavian artery, dysphagia lusoria, Bicuspid aortic valve

Introduction:
Coarctation of the aorta (CoA) was first described by Morgagni in 1760 as a zone of constriction in the descending aorta. The association of a bicuspid aortic valve with coarctation of the aorta is well recognized, but estimates of bicuspid valve in patients with coarctation range from 25–85%. The clinical significance of a bicuspid valve in patients with coarctation is not well established. Hypertension and the gradient over the coarctation region causes more stress on the aortic valve and aortic wall, resulting in aortic valve disease. The presence of aortic problems in CoA predispose to dissection. Both lesions are intimately related and are considered part of a spectrum of diffuse arteriopathy. We report a case of CoA associated with Bicuspid aortic valve and severe aortic stenosis (AS). CoA is an important cause of secondary hypertension in the young age. The presence of dysphagia lusoria in our case was due to the aberrant right subclavian artery. Our case is a real surgical challenge.

Case report Figure 1: 2D Echo obtained from suprasternal position aortic arch view showing the coarctation ridge:shelf lesion distal to LSCA
A 48yr old female, mother of two children presented with dysphagia, exertional dyspnea, presyncope and intermittent claudication. She has been a known case of Hypertension since the age of 23yrs. The hypertension (HT) was diagnosed during her second pregnancy at a primary health care center and she is on regular antihypertensive drugs. She was not evaluated for secondary HT at that age. On clinical examination at admission she was found to have Right radio-radial delay and weak femoral pulses. The left upper limb showed Blood Pressure (BP)-180/110 and right upper limb BP was 110/80mmHg. Blood pressure was not recordable in lower limbs. A systolic thrill and a delayed peaking Ejection Systolic Murmur at Right 2nd Inter Costal Space conducted to carotids were seen. She had no features of Turner syndrome. The patient ECG Showed Left Ventricular Hypertrophy with systolic overload pattern. Echo showed,
Bicuspid Aortic valve, severe AS and coarctation of Aorta. The patient was taken to cardiac catheterization. With the left radial access the catheterization study showed aberrant right subclavian artery originating below the coarctation site. The coronary angiography showed anomalous separate origin of Left Circumflex artery (LCX) from left coronary sinus and patent coronaries. CT angiography confirmed the findings. She is referred to higher Centre for surgical intervention.

Discussion

Dysphagia lusoria (or Bayford-Autenrieth dysphagia) is an abnormal condition characterized by difficulty in swallowing caused by aberrant right subclavian artery. It was discovered by David Bayford in 1761 and first reported in a paper by the same in 1787. Our patient had dysphagia as the prime symptom which gave leads to the diagnosis. The BP recordings are disproportionate in both upper limbs. The left upper limb showed hypertension which was missed earlier, as the measurement was made regularly in the right upper limb. The aberrant subclavian artery was having a retro esophageal course which caused external compression resulting in dysphagia. ‘The gullet of woman has been strangled by this grotesque freak of nature’. The right subclavian artery is the last branch of the aortic arch in ~1% of individuals. It courses to the right behind the esophagus in ~80% of these cases, between the esophagus and trachea in 15%, and anterior to the trachea or mainstem bronchus in 5%. A retroesophageal course may be the cause of so-called dysphagia lusoria.

Figure 4: Cardiac catheterization image showing aberrant RSCA (arrows).

In our case the aberrant subclavian artery had its origin distal to the coarctation segment. There were extensive collaterals formed by left subcavian artery via Left Internal mammary artery (LIMA), axillary artery and their subdivisions to the posterior intercostals and then into the descending aorta. The collaterals caused unilateral rib notching on the left side. The limbs did not show difference in circumference. Right sided notching may occur when Left Subclavian artery is involved in actual coarctation which is not seen in our case. Of those surviving the serious hazards of the first one or two years, 25 per cent die before they reach 20, 50 per cent by 32, 75 per cent by 46, and 90 per cent by 58 years. For coarctation the median is 31 years and the mode is widely spread through the second to fifth decades rather than closely clustered round a point. The patient has spanned her pregnancy uneventful and survived four decades. It is the coexistence of BAV that has progressed to severe aortic stenosis made our patient symptomatic. She underwent cardiac catheterization and aberrant Right Subclavian artery was found distal to CoA. The coronaries showed anomalous origin of LCX and no significant obstructive lesion was found. This complex coarctation is a high risk surgical candidate. In our case the Doppler study showed a diastolic velocity >193 cm/s and Diastolic/systolic velocity (DV/SV) ratio >0.53 which correlates to a Cardiac Magnetic Resonance imaging coarctation aortic aneurysm.
index (CoAi) <0.25 indicative of severe coarctation .9 Surgical approaches to coarctation include resection and end to-end anastomosis, subclavian flap aortoplasty, prosthetic patch aortoplasty, bypass grafts between ascending and descending aorta. The presence of discrete coarctation, aberrant subclavian artery and severe aortic stenosis needs all conditions to be addressed. Combined repair needs an anterior approach but the surgical mortality is high.

**Figure 6: Cardiac catheterization image showing 3 sign and notching of ribs (long arrows) on the left side**
The management of our patient with aortic coarctation and a coexisting cardiac disorder is still a surgical challenge. Single-staged procedures have lower postoperative morbidity and mortality rates than do 2-staged procedures. Rapid development in endovascular interventions allow repair of both lesions in the same surgical session.10

**Conclusion**
The presence of hypertension in the young should be attended seriously and a search for secondary cause should be intensively done. Measurement of four limbs BP should be a mandatory as CoA at earlier stages can be picked up. The association of BAV, severe Aortic stenosis and aberrant subclavian artery had made the surgical management challenging. Early management of CoA could have avoided serious complications.

**References**
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