J MEDICINE 2009; 10 (SUPPLEMENT 1) : 32-35

CASE REPORTS

UPPER BODY HYPERTENSION WITH GROSS ATHEROSCLEROTIC NARROWING AND DIFFUSE CALCIFICATION OF THE DESCENDING AND ABDOMINAL AORTA

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Introduction

Of the hypertensive patients presenting to the primary care providers, 95-99% have essential hypertension. Remaining 1-5% have an underlying diagnosis.[1] Narrowing of the aorta distal to the origin of the arteries to the head, neck & upper limbs causes upper body hypertension. Coarctation of the aorta, a congenital narrowing of the aorta just distal to the insertion of ductus arteriosus, is the major cause of upper body hypertension. Gross atherosclerotic narrowing & diffuse calcification of the descending & abdominal aorta rarely emulates the features of coarctation of the aorta.

We report a case of upper body hypertension due to gross atherosclerotic narrowing & diffuse calcification of the descending & abdominal aorta after extensive medline search we have not come across of such reports yet.

Case Report

A 45 year old Bangladeshi lady presented with the complaints of headache, neck pain and chest tightness on exertion for two months. She experienced extreme weakness in her lower limbs upon walking short distances. Walking for even 5-10 minutes brought pain followed by numbness of the lower limbs. She was known to be hypertensive for three years and said that drugs usually made her worse. On one occasion last year she said that she became ill when she took drugs for blood pressure. Her past papers showed that her creatinine climbed to around 2 mg/dl. She discontinued the medication & did not go back for follow-up. She is a mother of two children in their twenties & thirties and the pregnancies were uneventful. She had an abdominal hysterectomy about four years back but no papers are available but she says recovery was uneventful. Examination revealed a blood pressure of 240/120 mmHg in the right arm, 160/120 mmHg in the left arm (all sitting), 120/110 mmHg in the left popliteal (recumbent) (she was taking 5mg amlodipine daily). Her pulse was regular at 76/minute with a radio-femoral delay & subtle radio-radial delay but the pulses below the popliteals were barely palpable. She had gross supra-sternal pulsation. She had a heaving apex. Heart sounds were normal with a harsh ejection systolic murmur (Grade 4/6) best heard over the supra-sternal area with carotid shudder better heard on the left than right. Peculiarly she had an early diastolic murmur (Grade 1/6) only over the supra-sternal area. Lungs were clear. Abdominal bruit was present. At fundoscopy she had Grade 1 hypertensive retinopathy. Her neck veins were flat there was no pedal oedema or thyromegaly & dermatologic examination was unremarkable. Results of lab studies showed normal full blood count, Liver function test, blood sugar 2 hours after breakfast, lipid profile. Serum creatinine was 0.85 mg/dl, serum calcium was 2.9 mmol/L and value of serum sodium, potassium, chloride, calcium were 149.2, 5.82, 105.3, 1.534 respectively. Serum pH (venous) was 7.848 and VDRL was non reactive.

Her ECG met the voltage criteria for LVH with left axis deviation. CTR on chest xray was normal with a LV preponderance of the cardiac silhouette. X-ray of the dorsal spine reveals that the arch, the length of the dorsal aorta and the visible portion of the abdominal aorta to be calcified (figure 1a and 1b).

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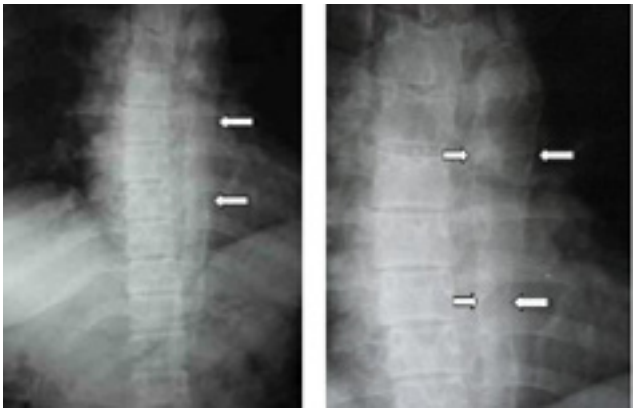

Fig.-1 (a & b) : *Figure 1a(left): calcification through the length of the visible aorta. Figure 1b(right): irregularity of lumen can be appreciated by irregularities of the calcific diameter seen.*

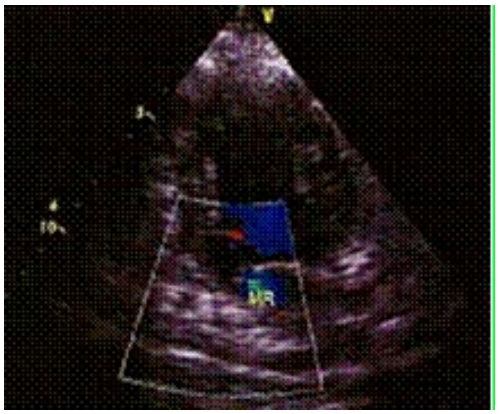

Fig-2: *Trivial mitral regurgitation.*

Echocardiogram showed mild LVH & trivial MR (Fig.-2). Ejection fraction. was 60% & lumen of the descending aorta was narrow.

Next a CT angiogram was done. Extensive calcification of the aortic wall was seen starting from the distal aortic arch to the upper abdominal aorta at the level of L2 vertebra. Gross calcification & narrowing, upto 1cm in diameter, was seen at the mid dorsal level. Distal dorsal aortic diameter was about 1.7cm & about 1.5cm at the level of the isthmus. There was gross calcification & narrowing, upto 1.5cm in diameter, of the proximal abdominal aorta. Visceral filling at the coeliac trunk, superior & inferior mesenteric and the renal arteries were unremarkable (Fig.-3).

Two weeks after she was put on a combination of 80mg Valsartan, 12.5mg Hydrochlorthiazide, 50mg Atenolol & 5mg Amlodipine daily her blood pressures were as follows 200/90 mmHg (right arm, sitting), 130/80 mmHg (left arm, sitting) and 100/70 mmHg (left popliteal, recumbent). A further reduction in blood pressure was not contemplated due to risks of renal hypoperfusion. After 4 weeks of above treatment her followup investigtions revealed, serum creatinine 1.1 and serum sodium, potassium, chloride were 143.1, 4.13 and 104.8 respectively.

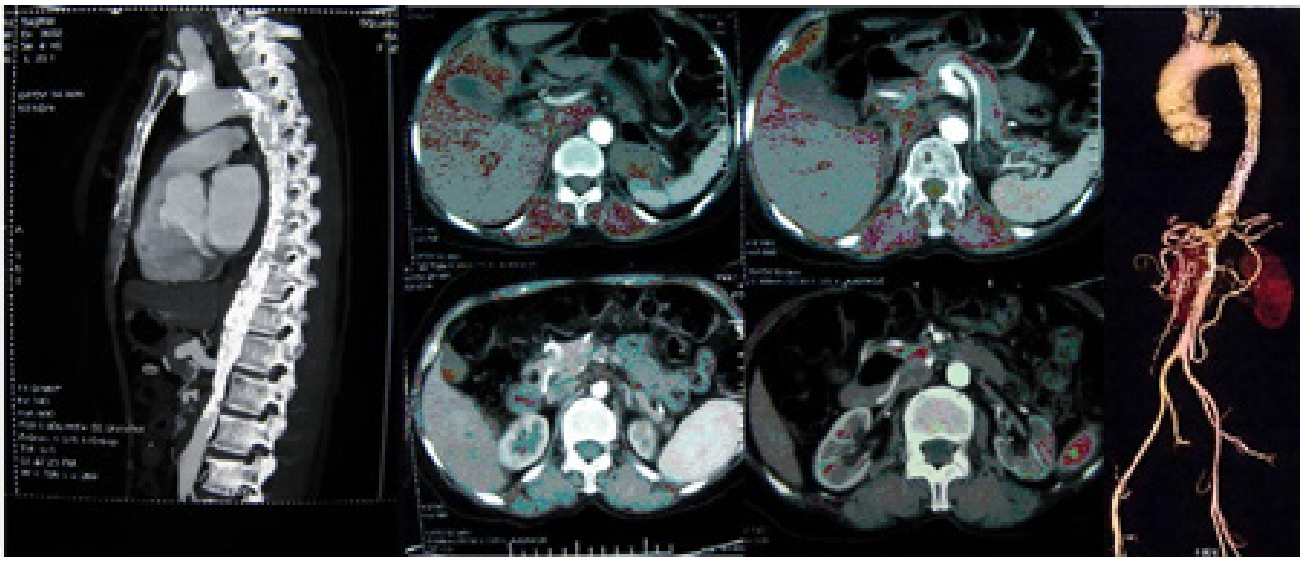

Fig.-3: *Visceral filling not remarkably affected despite the gross atherosclerotic narrowing and diffuse calcification. Gross segmental narrowing from below the origin of celiac trunk to infrarenal aorta.*

Discussion

Blood pressure (BP) recordings often differ between arms, but the extent to which these differences are reproducible and whether the differences have prognostic importance is unknown.[2] Yet the practice in clinical medicine should routinely include bilateral BP measurements in sitting position and in case of any major difference an inquisitive clinical examination will usually pay off. Atherosclerotic changes in the left subclavian artery was a major cause of significant BP difference between the arms in this case. CoA of the descending thoracic aorta generally presents in childhood. The aortic CoA in adult patients is extremely rare; only a few cases where it is the sole congenital malformation or where it is combined with other defects in the same patient have been reported.[3] Atherosclerotic aortic disease mimicking CoA is also rare. Such diffuse calcification of the aorta as in the present case has been rarely reported.

Presence of atherosclerotic aortic plaques is a strong independent predictor of coronary artery disease and of embolic events.[4] Moreover, calcification of the aortic arch is significantly and independently related to increased risk of CHD in both sexes and with increased risk of ischemic stroke among women.[5] Our patient had a normal coronary angiogram.

Medical management is to lower the BP without causing renal hypoperfusion. Lower limb BP, urine output and followup creatinine & electrolyte would serve as a guide.Surgical management will be a debate amongst CVT surgeons. The aortic arch has long been considered an "off-limits" area for the majority of cardiothoracic surgeons, mainly due to the adverse neurologic sequelae following such a complex surgical reconstruction.[6] Pethig et al. pointed out severe hemodynamic instability after relief of the aortic CoA with ascending-descending aorta bypass.[7] None the less a logical surgical plan for this patient would be ascending aorta to infrarenal bypass with synthetic tube graft followed by renal revascularisation – auto-transplantation of kidneys or bypass from synthetic aorta to renal artery. This would save the kidneys, reduce the other complications of uncontrolled hypertension & improve her lower limb symptoms.

Vascular calcification, long thought to result from passive degeneration, involves a complex, regulated process of biomineralization resembling osteogenesis. Evidence indicates that proteins controlling bone mineralization are also involved in the regulation of vascular calcification. Vascular calcification may include both osteogenic and chondrogenic differentiation. In human, it is primarily osteogenic with bone tissue formation. Understanding its mechanism has implications for management of vascular disease, particularly in the context of bone disease.[8] Atherosclerosis in the thoracic segment of the aorta is closely associated with fat deposition within the plaques, resulting in positive remodeling of the vessel. On the other hand, in the abdominal segment, atherosclerosis may or may not be associated with fat deposition. In the case where atherosclerosis is associated with fat deposition, there would be positive remodeling of the vessel with arterial dilation, which could be related to the genesis of aneurysm; conversely, in the case where it is not associated with fat deposition, plaques would have more fibrosis and calcification, and the increased rigidity of the wall would prevent compensatory dilation, thus originating the obliterative form of the disease.[9]

Conclusion

Vascular calcification is an important manifestation of atherosclerosis. For more than half a century, it has been associated with a poor prognosis attributable to vascular disease.[10] However, the process reduces the vascular resilience required for hemodynamic function and introduces high mechanical failure stress in the artery wall & hence the importance of calcium score in CAD. Diffuse calcification of the length of dorsal aorta continuously upto a significant part of the abdominal aorta is rare. To make it more complicated the patient has segments of gross atherosclerotic narrowing in the dorsal aorta & abdominal aorta. This has given a clinical picture of upper body hypertension, a clinical mimicry of adult coarctation of aorta. At the same time suprarenal aortic narrowing is possibly having a positive feedback effect on hypertension simulating renal artery stenosis .Therefore, it is a rare diagnostic & therapeutic challenge.

Acknowledgements

Mr Abu Yousuf Rana, Mr Safikul Alam Sujan.

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