Case Report

Two Cases of Secondary Hypertension with Rare Aetiology

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Secondary Hypertension comprises approximately 5% of Systemic Hypertension[1]. Renal parenchymal, Renovascular and Endocrine Diseases are amongst the common causes of Secondary Hypertension. Takayasu's Arteritis is a rare form of Primary Systemic Vasculitis that appears to be commoner in Asia than Europe or North America[2] and in contrast to Japanese patients, who have a higher incidence of aortic arch involvement, the series from India reports higher incidences of thoracic and abdominal involvement. In general, patients from Indian subcontinent tend to have greater prevalence of Pan-aortic Disease (both above and below the diaphragm) when compared with the west. In India the women : men ratio is around 1.5:1. Takayasu's Arteritis most frequently affects young women[3]. Therapeutic intervention like Percutaneous Transluminal Angioplasty (PTA) and Stenting, By-pass Surgeries or surgical reconstruction should be performed when disease is made inactive by the use of effective immunosuppressive Therapy[4].

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Key words **: Takayasu's Arteritis (TA), Descending Thoracic Aorta (DTA), Diethylenetriaminepentacetate (DTPA).**

Case 1

A twenty three year old female presented with occasional attacks of Malaise, Fatigue, Palpitation, Headache for about six months duration. There was no history of fever, joint pain, seizures, blurring of vision or history suggestive of limb claudication, or weight loss / chest pain. Her menstrual cycles were normal and she had no addiction and there was no significant past history. Her father is diabetic.

Examination — She was found to be hypertensive with Blood Pressure (BP) - Right / Upper Limb (UL)-160/92, Left/UL-158/94, Rt/Lower Limb (LL)-120/90 and Lt/LL -134/90, pulse 98 per minute regular. Carotid, brachial, radial pulses were well palpable but femoral, popliteal, posterior tibial and arteria dorsalis pedis were less palpable. No carotid /subclavian/abdominal bruit. CVS- S_1, S_2(normally audible), no murmur, no added sound; chest- Nothing Abnormal Detected (NAD), abdomen – NAD, CNS - NAD.

Investigations — Hb-11.7gm%, ESR-09mm, TWBC-6500/cumm, CRP-0.59mg/l (<6), K^+- 4.4mmol/l, USG of whole abdomen – NAD, Cr-0.77mg%, Glucose (fasting)-84mg/dl, LFT-WNL, TSH-1.59, HB_SAg-negative, ECG-sinus tachycardia, ECHO-NAD, LVEF-68%.

She was put on anti-hypertensives (diuretics, amlodipine, beta-blockers). But her BP was not controlled. So Renal Artery Doppler was done which showed increase AT in Bilateral Renal Arteries and Interlobar Arteries with concentric wall thickening of aorta with focal stenosis involving supracoeliac part. CT Angiography of aorta (Fig 1) showed – Long segment (about 8.5cm in length

Editor's Comment :

- Diagnosis of TA is often delayed and so there must be high index of suspicion for young female patients of Indian origin suffering from hypertension — so, as to treat them early with medical and surgical intervention.

Fig 1 — CT Angiography of aorta

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with inferior extent 4cm above the coeliac trunk) moderate stenosis of thoraco- abdominal aorta, -features likely to suggest TA-there was also mild stenosis at the origin of coeliac trunk. Aortic root, ascending aorta (max. diam. of 27mm), aortic arch and its branches (no plaque/ stenosis), Bilateral Renal Arteries, Superior and Inferior mesenteric arteries and infrarenal and juxtarenal abdominal aorta appear normal, with normal aortic bifurcation.

Visualised illiac arteries appear normal. On conventional Angiography all are normal except long segment stenosis of maximum 70% severity of Descending Thoracic Aorta (DTA) and coeliac ostial discrete lesion of 90%.

Final diagnosis — Takayasu's Arteritis type-III.

Patient was put on immunosuppressants first, followed by, Angiography and Stenting. After Stenting of DTA – BP was better controlled with lesser dose of anti-hypertensives.

Case 2

A twenty five year old female was incidentally detected to have hypertension during pre-anaesthatic check up for a sialoadenoscopic evaluation for her right submandibular duct stone. There was no history of headache, palpitation, abdominal pain, vomiting, fever, joint pain, muscle pain, convulsion, blurring of vision, weakness of limbs, weight loss, limb claudication and chest pain. She had no addiction. Her menstrual cycles were regular. There was no significant Family history or past history.

Examination — Mild pallor was there but there was no cyanosis, no clubbing, no pedal oedema and JVP was not raised. BP was – (Rt/UL-164/90), (Lt/UL-160/94), (Rt/LL-154/90), (Lt/LL-150/92); PULSE – 96/min, equally palpable in all four limbs. Both carotids are equally palpable. There was no carotid/subclavian/abdominal bruit. Chest-NAD, CVS-S_1, S_2 (normally audible), no murmur, no added sound; CNS-NAD, abdomen-no ascites or organomegaly.

Investigations — Hb-9.2gm%, ESR-102mm, TWBC-7720/cumm. DC- normal, CRP-7.2mg/l (<6) , Na^+-141, K^+-4.2 , Cr-0.82mg%, SGPT-10units/l, ECG- Within Normal Limits (WNL), ECHO-NAD.

Renal Artery Doppler —There was intimal thickening of aorta, extending into the origin of Left Renal Artery, leading to luminal stenosis. Right renal artery appears normal. Impression-left proximal renal artery stenosis. CT Angiogram of Abdominal Aorta (Fig 2) -small left kidney (72mm), large right kidney (108mm), there was tight stenosis (99%) of proximal left renal artery including ostium with post stenotic dialatation and poor opacification of left kidney, mild narrowing of right renal artery sparing ostium. There was also concentric wall thickening of aorta causing mild narrowing with involment of superior mesenteric artery.

MR Angiogram — Vertex to toe-all vessels were normal except in the perirenal aorta there was uniform

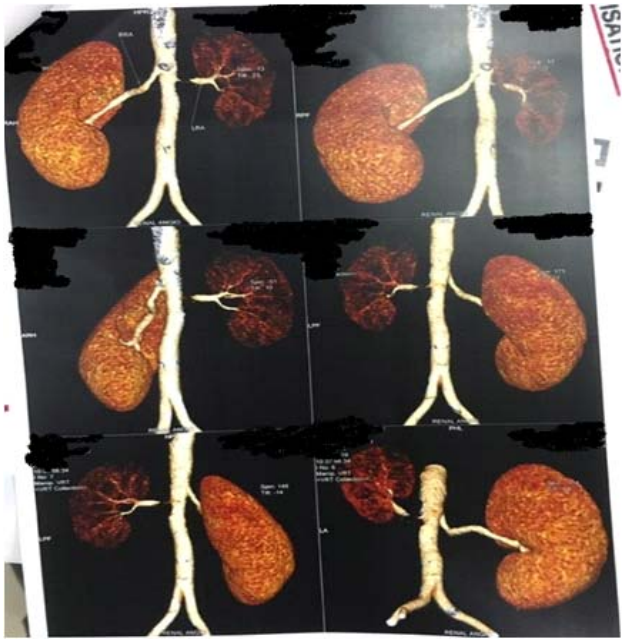

Fig 2 — CT Angiogram of abdominal aorta

circumferential thickening with mid segment narrowing (30%) of Right Renal Artery and Left Renal Artery showed tight stenosis at its origin. Left kidney was reduced in size. Visualised Pulmonary arteries appeared normal. Impression-Takayasu's arteritis-type-IV. Diethylenetriaminepentacetate (DTPA) Renal scan-relative function-left kidney (22%), right kidney (78%). DTPA clearance –left kidney (18 ml), right kidney (61ml). Left kidney was contracted-severely impaired perfusion and parenchymal function; right kidney-good parenchymal function and drainage. Patient was put on anti-hypertensives (amlodipine+metoprolol), immune suppressants (deflazacort + mycophenolate mofetil+ tocilizumab), followed by Angiography and stenting of Left Renal Artery.

Final diagnosis — Takayasu's Arteritis type-IV with Left Renal Artery Stenosis. After Renal Artery Stenting her blood pressure is now normal without any anti-hypertensive medication.

Discussion

TA is more commonly seen in women[5] than men in India and Asia. In clinical practice the diagnosis of TA is almost always secured by an imaging procedure[6] that demonstrates the characteristic abnormalities of the aorta and its major branches. Unfortunately the diagnosis of TA is often delayed. Many of these delays can be prevented by remembering that TA should be included in the differential diagnosis of any person younger than 40 years, who present with FUO, aortic regurgitation, hypertension or unequal or absent pulses. Once an imaging test demonstrates disease of the aorta or its major branches, the differential diagnosis narrows to a set of disorders that are usually differentiated by their

clinical features and other investigations. Most rheumatic diseases that can affect the aorta are distinguished by their associated features. For example, Cogan's syndrome typicaly produces ocular inflammation (especially Keratitis) and vestibuloauditory dysfunction . Giant Cell Arteritis (GCA) can be distinguished from TA by its age range (50years or more – average age of onset – 72 years), visual loss (10%-30%) and less involvement of aorta (25% in GCA whereas 100% in TA)[7]. Infection of the aorta is rare. Tertiary Syphilis can be excluded by a negative fluroscent treponemal antibody test. Other diseases of the aorta are usually readily seperated from TA by the history and physical examination. There has been much speculation about the role of Mycobacterium Tuberculosis in the aetiologies of TA, especially in India[8]. It remains possible that this is simply coincidence of a common infection with a rare vasculitis. In India Numano Types IV and V are most frequent.

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