REFRACTORY HYPERTENSION IN A CASE OF TYPE III TAKAYASU ARTERITIS TREATED WITH PERCUTANEOUS AORTOPLASTY- A CASE REPORT

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ABSTRACT

Introduction: Takayasu arteritis is a chronic, medium and large vessel vasculitis of unknown aetiology, affecting aorta and its primary branches and is usually seen in young females. Patients present with non-specific symptoms, syncope, claudication, absent pulses, hypertension or discrepancy in blood pressures. Conventional or MR angiography is used routinely for diagnosis, but biopsy of vessel involved is confirmatory. The mainstay of therapy is glucocorticoids or other immunosuppressive agents. Some patients may need angioplasty or bypass grafting. The disease tends to be progressive with intermittent remissions and exacerbations. Case report: We here with present a 20 yr old hypertensive female with head ache, syncopal attack, feeble peripheral pulses, carotid bruits and grade III hypertensive retinopathy, showing involvement of arch and descending aorta and its branches in conventional and MR angiography. Findings were suggestive of TYPE III TAKAYASU ARTERITIS with refractory hypertension, without renal arterial involvement. After initial treatment with steroids followed by aortoplasty, her hypertension improved significantly. An uncommon mechanism of hypertension and prompt response to treatment brings the case for presentation.

KEYWORD

Aortic arch syndrome, Aortoplasty, Refractory hypertension, Takayasu arteritis.

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INTRODUCTION:

Takayasu arteritis (TA) is an inflammatory and stenotic disease of medium and large sized arteries characterized by a strong predilection for the aortic arch and its branches, and hence called "Aortic arch syndrome". The disease was first reported by R Yamamoto in 1830, while the first presentation was in 1905 by Mikito Takayasu, describing a patient with a peculiar optic fundus abnormality [1]. TA is commonly seen in Asian and certain central and South American countries with 10 fold predominance in women, especially in less than 40 years of age [2]. Takayasu arteritis has been associated with different human leucocyte antigen (HLA) alleles in different populations [3]. Hypertension develops in more than one-half of cases due renal artery involvement [4]. Early initiation of treatment with steroids and management of complications decreases mortality and morbidity. We present an interesting case of "TYPE III TAKAYASU ARTERITIS" with refractory hypertension without renal artery involvement and which responded to treatment with complete normalization of blood pressure in a very short period.

CASE REPORT:

A 20 Year old female, hypertensive presented with history of diffuse dull aching type of head ache for 18 months and one episode of syncope. Past history was not significant except for refractory hypertension and anti-hypertensive medication. She was married and has one live child. Family history and obstetric history were not contributory. On examination patient was short in stature and ill nourished with a Body Mass Index of 16.09 Kg/m^2. Left brachial and radial pulses and dorsalis pedis and posterior tibial pulses on both sides were feeble. There was large discrepancy in blood pressures (BP) in limbs with pressures of 180/100 mmHg, 240/110 mmHg, 180/100 mmHg and 200/100 mmHg in left and right upper limbs and left and right lower limbs respectively. Cardiovascular examination revealed bilateral carotid thrill and bruit, heaving apex in normal position and grade IV ejection systolic murmur in aortic area. Grade III hypertensive retinopathy changes were found on fundus examination. Other systems were normal.

Investigations revealed hemoglobin 10.6 g/dl, total leucocyte count 19,200/ml, erythrocyte sedimentation rate (ESR) 85/1st hour and C - reactive protein (CRP) 8mg/dl. Renal parameters, serum electrolytes and fasting lipid profile were normal. Ultrasound abdomen revealed normal kidney size and echo-texture. Chest x ray was normal. ECG was showing left ventricular hypertrophy (LVH). 2D echocardiography showed concentric LVH with grade I diastolic dysfunction, dilated ascending aorta, arch and descending aorta. Aortogram showed 80-90% stenosis of descending thoracic aorta at the level of T6 – T11vertebrae and left subclavian artery stenosis at its origin (fig 1&2). Coronary and renal angiography were normal. CT and MRI scan of thorax and abdomen was showing diffuse circumferential wall thickening of arch of aorta and descending thoracic aorta up to the level of celiac trunk with maximal narrowing in sub carinal level extending up to the diaphragmatic outlet, the length of stenotic segment was 7 cm. As per diagnostic criteria and classification this is a case of "TYPE III TAKAYASU ARTERITIS".

The patient was initially treated with oral prednisolone at a dose of 40 mg per day and continued on ACE inhibitors, beta

blockers, calcium channel blockers and diuretics without effective control of hypertension. With administration of steroid for one week, ESR normalized and the patient symptomatically improved. Then the patient was subjected to percutaneous aortoplasty and 14×100 mm self expandable stent was placed after dilating the stenotic segment of aorta (fig 3&4). Left subclavian angioplasty was also done. A fall of 50 mm Hg of intra aortic pressure in descending thoracic aorta was noted during the procedure. Post operatively the patient was given intravenous heparin and anti-platelet therapy, atorvastatin 80mg daily and then gradually shifted to oral anticoagulants.

Immediately following the procedure, she was hemodynamically stable and interestingly the blood pressures in all 4 limbs reduced to near normal values. She was discharged with dual anti-platelet drugs and atorvastatin and no antihypertensive medication was advised at discharge as she could maintain normal blood pressure without drugs. She is on regular follow up and is maintaining normal BP without drugs. Complete and immediate normalization of blood pressures was not expected after procedure and this makes the case unique for presentation.

DISCUSSION:

Takayasu Arteritis is a chronic inflammatory disease affecting the aorta and its primary branches. The inflammation is either localized or may involve the entire vessel. The initial vascular lesions frequently occur in the left middle or proximal subclavian artery, but there may be variation in disease expression. As the disease progresses, aorta or its branches may also be affected. The abdominal aorta and pulmonary arteries are involved in approximately 50% of patients.

TA is classified as follows

Type I: Involvement localised to aortic arch and its branches.

Type II: Involves the thoraco abdominal aorta and its branches, sparing the arch.

Type III: Involves aortic arch as well as descending aorta.

Type IV: Includes pulmonary arterial involvement.

Type V: Aneurismal type

More than half the patients develop an initial systemic illness characterised by fever, anorexia, malaise, weight loss, night sweats, arthralgias, pleuritic pain and fatigue. After a latent period of variable duration, symptoms and signs referable to the obliterative and inflammatory changes in the vessels develop. Patients with Types I and III disease manifest with typical findings of absent or diminished upper limb pulses (and hence called pulseless disease) and barely detectable blood pressure in the arms, higher pressures in the lower extremities, bruits overlying the diseased arteries and syncope. Patients with Type II arteritis may have abdominal angina, claudication of the limbs and hypertension.

Hypertension in this disease can be due to several causes:

- Renal artery stenosis, present in 35-85% of cases
- Involvement of baroreceptors by aortitis
- Loss of elasticity of aorta
- Coarctation-like lesion
- Aortic regurgitation

In our case there was refractory hypertension without involvement of renal artery.

Aortic regurgitation (AR) may occur due to severe hypertension or to inflammation with scarring of the aortic valve. Hypertension and AR may result in heart failure. The ostia and proximal segments of the coronary arteries may also be affected, resulting in angina or myocardial infarction. Neurologic symptoms include syncope, headache, hemiplegia, paraplegia, visual disturbances and hypertensive encephalopathy. The retinopathy described by Takayasu is seen in only 25% of cases and is usually associated with carotid arterial involvement. Other eye signs include A-V anastomosis, iris atrophy, retinal haemorrhage, micro aneurysms, mydriasis, hyperaemia of conjunctiva, retinal detachment and visual loss. Though Pulmonary artery involvement is found in about 50%, clinical pulmonary manifestations are rare or absent.

During acute phase, low-grade leucocytosis and mild anaemia of chronic disease are frequent. Acute phase reactants such as an elevated ESR, serum CRP, alpha-2 globulin concentrations, anti-streptolysin titres and hypoalbuminemia are a reflection of the underlying inflammatory process. These tests are not always precise or invariably reliable indicators, but reflect disease activity. Chest X-ray is usually unrevealing, although a rim of calcification is sometimes seen in the walls of the affected arteries. Chest X-ray and ECG may show signs of pulmonary hypertension and right ventricular enlargement in type IV.

Arterial biopsy may be positive in 20-35% of cases. Because of the central location of the arteries involved, arterial biopsy is not an option. So confirmation is best done by conventional or MR angiography. In angiography changes are most pronounced in the region of the aortic arch and its primary branches or limited to the more distal aorta and its branches. Most common lesion is a smooth, concentric, arterial or aortic narrowing (85%). Irregular narrowing, complete occlusion and fusiform or saccular aneurysms are less commonly seen. The affected thoracic aorta is described as having a 'rat-tail' appearance. Collateral circulation is often prominent because of the chronic nature of the disease.

Contrast-enhanced MR perfusion imaging, ultrasonography and positron emission tomography (PET) are new, non-invasive methods of assessment that are likely to replace conventional angiography. PET scan utilizing radioactively labelled fluoro deoxy glucose (FDG) can be used, but has limited availability [5,6]. Areas of increased uptake of the tracer correlate well with abnormal arterial segments noted by MRI. It may be more sensitive than MRI in detecting segmental arterial inflammation[7] and can distinguish active inflammation from scar.

The American College of Rheumatology has established classification criteria for Takayasu arteritis to distinguish this disorder from other forms of vasculitis

- Age at disease onset ≤ 40 years
- Claudication of the extremities
- Decreased pulsation of one or both brachial arteries
- Difference of at least 10 mmHg in systolic blood pressure between the arms
- Bruit over one or both subclavian arteries or the abdominal aorta
- Arteriographic narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities, not due to arteriosclerosis, fibro muscular dysplasia or other causes

Patients are said to have Takayasu arteritis if at least 3 of 6 criteria are present; this classification yields a sensitivity and specificity of 90.5 and 97.8 percent, respectively[8].

Since the aetiology of TA is still not fully understood, treatment is not curative but rather primarily medical and symptomatic. The mainstay of therapy is glucocorticoids. An initial daily dose of 45 to 60 mg of prednisone or its equivalent dose should be started as soon as the diagnosis is suspected. A decrease and eventual disappearance of constitutional

symptoms accompanied by a decrease in acute phase reactants is expected. CT or MRI scans can be used to follow the response. If the patient presents after arterial damage has occurred, the disease is less likely to respond to immuno-suppression. Gluco-corticoid dose can be gradually tapered and discontinued if the disease goes into remission, while the dose should be increased if exacerbations occur. Long-term low-dose prednisone therapy may be necessary to prevent progression of arterial stenosis. The adjunctive use of methotrexate, azathioprine and other cytotoxic agents are reserved for patients with steroid resistance or relapse. Anti-tumor necrosis factor (TNF) agents might be another treatment possibility [9]. Treatment of hypertension and heart failure should be instituted if these complications occur.

Percutaneous transluminal angioplasty or bypass grafts may be considered in symptomatic patients with irreversible arterial stenosis and it should be postponed until disease remission if possible [10]. Renovascular hypertension, coarctation of aorta, severe cerebral ischemia and severe aortic regurgitation causing congestive heart failure or progressive aneurismal enlargement or dissection may all require prompt surgical treatment.

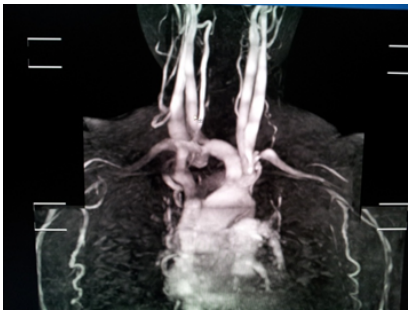

Figure.1 MR angiography showing left subclavian artery stenosis at its origin

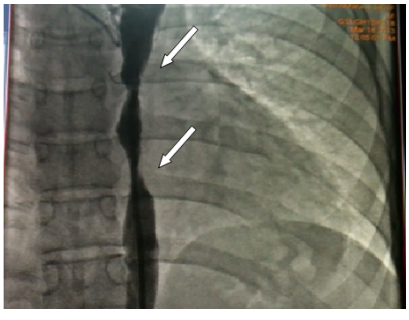

Figure.2 Aortography showing stenosis of descending thoracic aorta.

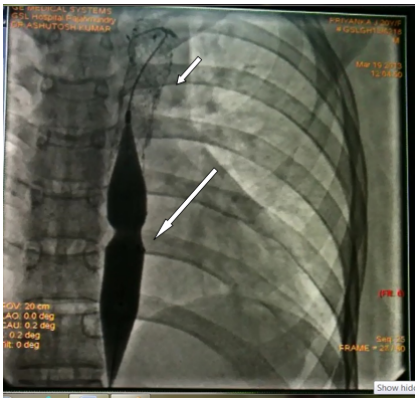

Figure3.

Contrast angiography showing ballon dilatation of aortic stent after it is deployed.

Short arrow is showing stent in place and long arrow is showing contrast filled ballon.

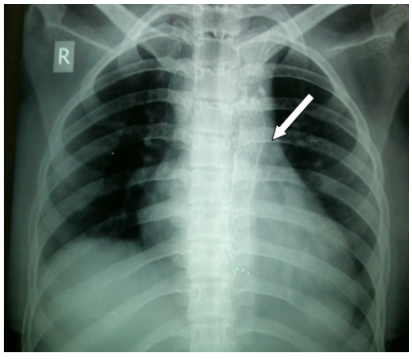

Figure.4 Chest radiography showing metallic stent in descending thoracic aorta.

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