TAKAYASU ARTERITIS: PRESENTED AS HEADACHE, A RARE CASE REPORT

INTRODUCTION
TAKAYASU'S ARTERITIS is an inflammatory and stenotic disease of medium and large sized arteries characterized by a strong predilection for the aorta and its branches. It is also known as AORTIC ARCH SYNDROME, PULSELESS DISEASE. Dr. Takayasu described the retinal changes of the disease in 1905 at the Annual Ophthalmology Society meeting in Japan, and his abstract was subsequently published in 1908. The condition is now called Takayasu arteritis in his honor. Annual incidence rate of Takayasu Arteritis is 1.2-2.6 cases/million, mostly Prevalent in adolescent and young women. In India, the female-to-male ratio is 1.6:1. Most patients are aged 4-63 years, mean age of onset - 30 years. <15% of cases present in individuals older than 40 years. Takayasu arteritis is observed more frequently in patients of Asian or Indian descent. Japanese have a higher incidence of aortic arch involvement. Subclavian artery (93%) is involved most commonly followed by Common Carotid (58%), Abdominal Aorta (47%), Renal Arteries (38%) and Aortic Arch (root) (35%). India report higher incidences of abdominal involvement. Pathology of Takayasu Arteritis involves Panarteritis, inflammatory mononuclear infiltrates and occasionally giant cells, with marked intimal proliferation, fibrosis, scarring and vascularisation of media, disruption, degeneration of elastic lamina. Also there is narrowing of lumen due to thrombus. Circulating immune complexes—demonstrated in 65%), Arthralgias (28%-75%), Fever (9%-35%), Weight loss (10%-65%), Ulcerated sub-acute nodular lesions (<2.5%), Pyoderma gangrenosum (<1%). The most discriminatory finding in TA is a systolic blood pressure difference (>10 mm Hg) between arms. Hypertension due to renal artery involvement is also found in approximately 50% of patients. Absent or diminished pulses are the clinical hallmark of Takayasu arteritis, but pulses are normal in many patients and upper limbs are affected more often than lower limbs. Carotidynia may be present. Bruits are often noted. Aortic regurgitation is a common finding. Ophthalmologic examination may show retinal hemorrhages, cotton-wool exudates, venous dilatation and beading, microaneurysms of peripheral retina, optic atrophy, vitreous hemorrhage, and classic wrinkle-like peripapillary arteriovenous anastomoses (extremely rare). Investigations done to diagnose Takayasu Arteritis are Complete Blood Count, C-reactive protein (CRP), Erythrocyte sedimentation rate (ESR), Electrocardiogram (ECG), X-ray of the chest, Ultrasound, Arteriogram, Angiogram, Magnetic resonance angiography (MRA), Magnetic resonance imaging (MRI), USG Doppler, MRA or CT angiography.

CASE REPORT
21 year female, admitted with complaints of Headache, holocranial, on and off, throbbing type since 3 months. Intermittent low grade fever since 3 months. Palpitation on and off since 3 months. We investigated the patient, USG Abdomen & Pelvis, Aorta and Renal Doppler study & CT Aortogram was done. Involvement of Abdominal aorta, Right renal artery and Right subclavian artery was seen. Hence the diagnosis of Takayasu Arteritis Type 4 was made.

Her Investigations were Hb-10.4, WBC-5600, Platlets-21700, MCV-70, MCH-23, MCHC-33, BSL-103, BUN-18, Sr.Bil-0.5, SGOT-18, SGPT-9, Na-144, K-4.4, Creat-0.6. Urine routine and microscopy: 5-6 pus cell. Albumin 1+ sugar absent, ESR : 30, CRP : 12, P-ANCA : negative, C-ANCA : negative, ANA : negative, Transferrin Saturation : 2.8 (10-35), Montoux Test : negative, CRP : 12, P-ANCA : negative, C-ANCA : negative, ANA : negative.

USG Abdomen S/o thick intima media in abdominal aorta measures 0.19 cm. abdominal aorta measures 1.58 cm above bifurcation which small segment of narrowing in mid 1/3rd of abdominal aorta measures 0.83 cm. RK 6.4+2.4 cm,LK 9.8+4.9 cm. Aorta and Renal Doppler study-50% stenosis of the abdominal aorta is noted in the region of the SMA and Coelica axis. There are multiple stenotic segments at the origin of and mid portion of left renal artery S/o of aorta-arteritis with associated bilateral renal artery stenosis. CT Aortogram - Short segment wall thickening and narrowing of abdominal aorta with narrowing at the origin of iliac trunk and SMA. Stenosis at the origin and proximal part of right renal artery and shrunken right kidney. Stenosis at origin and accessory left renal artery. Small right subclavian artery with multiple collateral in right paravertebral and anterior chest wall.

DISCUSSION
Although our knowledge of TA has considerably improved over the last decade, the etiology and pathogenesis of this disease still remain
The mainstay of therapy in TA is immunosuppression, primarily with corticosteroids or methotrexate. Remission occurs in 40% to 60% of patients. Approximately 20% of patients are resistant to any therapy. Nearly all TA patients require surgery, usually for large-vessel vasculitis that warrants CABG or replacement of the aortic root or valve. Treatment of Takayasu arteritis is difficult, but patients who do have the right treatment can see positive results. Early detection is important. Most patients are treated with steroids and immunosuppressive drugs. Surgery is reserved for complications caused by narrowed arteries. Surgery to bypass narrowed arteries -- angioplasty or stents may be sufficient to supply blood or open up the constriction. Corticosteroids are the mainstay of therapy for active Takayasu arteritis, and some patients may require additional cytotoxic agents to achieve remission and tapers of chronic corticosteroid treatment. Oral corticosteroids are started at 1 mg/kg daily or divided twice daily and tapered over weeks to months as symptoms subside. Long-term aspirin therapy may be required. Cytotoxic agents are used for patients whose disease is steroid-resistant or relapsing. These agents are usually continued for at least one year after remission and are then tapered to discontinuation. The following agents with their respective doses are as follows: Methotrexate - 7.5-25 mg/week oral, Azathioprine - 1-2 mg/kg/day oral, Cyclophosphamide - 2 mg/kg/day oral (should be reserved for patients with the most severe and refractory disease states). Strict management of traditional cardiovascular risk factors such as dyslipidemia, hypertension, and lifestyle factors is mandatory to minimize secondary cardiovascular complications, which are the major cause of death in this disease. Additionally, low-dose aspirin may have a therapeutic effect in large vessel vasculitis. In a study of Tombetti et al, adjunctive treatment with anti-tumor necrosis factor (TNF) agents was effective in patients with active, relapsing Takayasu arteritis despite treatment with steroids and multiple other immunosuppressive agents. Critical stenotic lesions should be treated by angioplasty or surgical revascularization during periods of remission. Indications for surgical repair or angioplasty are as follows: stenosis causing hypertension, Coronary artery stenosis leading to myocardial ischemia, Extremity claudication induced by routine activity, Cerebral ischemia and/or critical stenosis of 3 or more cerebral vessels, Aortic regurgitation, Thoracic or abdominal aneurysms larger than 5 cm in diameter, Severe coarctation of the aorta.

Takayasu arteritis is a chronic, progressive disease. Its degree of activity varies over time; the intensity of its inflammatory processes typically fluctuates between exacerbation and remission or remission. Vascular involvement tends to be progressive. Vascular complications of the cardiac, renal, and central nervous systems are the chief causes of morbidity and death in TA, which is usually fatal when it remains untreated. Remission remains the goal of therapy. Identifying confounding complications (such as hypertension) and initiating aggressive treatment may afford better chances of symptom-free survival. In several follow-up studies, 5-year survival rates of 80% to 90% have been reported. Poor outcome depends chiefly on the presence of such complications as hypertension, aortic regurgitation, and aneurysm, and on a rapidly progressive course. In 1 study, the 15-year survival rate was 66% in patients who had a major complication versus 96% in patients who did not, and 68% in patients with a progressive course versus 93% in patients without.

After diagnosing Takayasu arteritis in our patient, conservative management with steroids and methotrexate was started and improvement in her symptoms was seen. As is evidenced by our patient’s presentation, death from Takayasu arteritis and death of patients who have the disease can be very complex. A fundamental understanding of the pathophysiology of TA, availability of diagnosing modalities is imperative in the choice of optimal care.

CONCLUSION
Takayasu's arteritis is a rare disease, with a potential for devastating consequences. As Clinical presentation is varied, and associated worsening of the symptoms due to late diagnosis, there is a need for further studies that could establish a strategy for the early diagnosis and treatment of takayasu Arteritis. It should be emphasized that despite the significant advance in noninvasive imaging modalities over past decade, detailed medical history and thorough physical examination still remain important for clinical diagnosis.

REFERENCES:
1. Das, Debabrata et al. "A Case of Unusual Presentation of Takayasu's Arteritis." Indian


