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TAKAYASU ARTERITIS: PRESENTED AS HEADACHE, A RARE CASE REPORT

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ABSTRACT Takayasu's arteritis is a chronic inflammatory disease that primarily involves the aorta and its main branches. Varying degrees of narrowing, occlusion, or dilatation develop in the involved vessel segments. We report a case of 21 year old female who had Headache (holocranial) which was 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.

KEYWORDS : takayasu arteritis, headache, abdominal aorta, young female, renal artery, TNF

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[1]. Annual incidence rate of Takayasu Arteritis is 1.2-2.6 cases/million, mostly Prevalent in adolescent and young women[2]. 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 but their pathological significance is unclear. Constitutional symptoms of Takayasu Arteritis involves Headache (50%-70%), Malaise (35%-65%), Arthralgias (28%-75%), Fever (9%-35%), Weight loss (10%-18%). Dermatological manifestations involve Erythema nodosum (6%-19%), 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 wreathlike 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, DOE grade 1 to 2 since 3 months. No H/o rash, hair loss, photosensitivity. N/o H/o of oral ulcer. No H/o arthralgia, N/o H/o of oliguria, dysuria, N/o breathlessness, edema feet, cough with expectoration ,No H/o chest pain, No H/o of TB/TB contact. General Examination:General Condition-Moderate, Afebrile, Pulse : 90/min, Regular,BP- Right upper limb 150/90 mmHg, Left upper limb 130/80 mmHg, Right lower limb 180/70 mmHg, Left lower limb 150/78 mmHg, RR : 16/min, Carotid Bruit present, Pallor present, No icterus, cyanosis, clubbing, edema feet, lymphadenopathy. Systemic examination RS: clear, Air entry equal on both sides, CVS : S1, S2 normal, No murmur, Per Abdomen : soft, non-tender, CNS: conscious oriented.

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,

ECG S/o LVH, Chest X-Ray –normal, 2D Echo S/o hypertensive heart disease, LVEF 60%, No RWMA, Fundus: mild temporal pallor present, Refraction error present. Serum Iron : 11(50-170), TIBC : 412 (250-450), 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 Coeliac 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 Aortagram -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

controversial. It is now assumed that the underlying pathogenesis is inflammatory with unknown etiology. Several etiologic factors have been proposed, including spirochetes, *Mycobacterium tuberculosis*, streptococci, circulating antibodies due to an autoimmune process, and genetic aspects. One hypothesis states that an antigen deposited in vascular walls activates CD4+ T cells, followed by the release of cytokines chemotactic for monocytes[3]. These monocytes are transformed into macrophages that mediate endothelial damage and granuloma formation in the vessel wall. Human studies, suggesting endothelial cell activation, have demonstrated increased expression of intercellular adhesion molecule-1 and vascular cell adhesion molecule-1 in patients with TA. Humoral immunity may also play a role in the pathogenesis. Antimonocyte antibodies and anti-endothelial cell antibodies are present in patients with TA and correlate with disease activity.

Clinical manifestations of TA are nonspecific. The clinical course of the disease is divided into an early active inflammatory phase and late chronic phase. The active phase lasts for weeks to months and may have a remitting and relapsing course. It is characterized by systemic disease with symptoms of Headache, fever, general malaise, night sweats, loss of appetite, weight loss, dizziness, arthralgia, skin rashes, etc. The acute phase does not occur in all patients, but constitutional symptoms are often seen in children with TA. It should be highlighted that the correct diagnosis of TA is seldom made in the early phase. Evidence of vessel inflammation such as tenderness along arteries, bruits, and aneurysm may point to the diagnosis of TA. The late chronic phase is the result of arterial stenosis and/or occlusion and ischemia of organs. The finding of hypertension and arterial bruits in young adults necessitates the examination of pulses and blood pressures in different limbs in order to detect asymmetry[4]. Elevated erythrocyte sedimentation rate is a common finding; however, caution is advised, because up to 50% of patients may have active TA disease and a normal sedimentation rate. The correct diagnosis usually depends upon the presence of other characteristic features (skin lesions, hilar adenopathy, or Bell's palsy). The diagnosis of TA is reached on the basis of clinical presentation and imaging results; histopathologic confirmation can be obtained in patients who undergo vascular surgery. Pathogenesis of arterial hypertension due to TA is complex, multifactorial, and not fully understood. At present, it is thought to be the result of three mechanisms: (a) mechanical, in which hypertension proximal to narrowed aorta(atypical coarctation) is due to high resistance to cardiac output imposed by narrowing; (b) neural, in which hypertension proximal to narrowed aorta results from aortic arch baroreceptors readjustment and this allows to ensure adequate blood supply to organs distal to narrowed aorta ; and (c) hormonal, in which hypertension is caused by renal hypoperfusion due to stenotic lesions of one or both renal arteries or aorta alone. A decrease in elasticity of arterial walls observed in TA may also contribute to the elevation of the blood pressure[3].

Hypertension occurs in one third of patients and is usually caused by renal artery stenosis. Coronary artery vasculitis is relatively uncommon, and when it is detected (in fewer than 5% of TA patients), it chiefly affects vessel origins. Left ventricular dysfunction caused by myocarditis has been reported in up to 18% of cases. Suspected TA mandates vascular imaging. While the intra-arterial angiography still remains the standard for diagnosis and evaluation of Takayasu arteritis, it has been largely replaced by computed tomography angiography or magnetic resonance angiography (MRA). The other investigative modalities, such as gallium-67 radionuclide scanning and positron emission tomography utilizing 18F-FDG, are currently in vogue when evaluating such patients, although they are not widely available yet.

Assessing disease activity in patients with Takayasu arteritis is frequently challenging, since clinical, biologic, and radiologic information do not always correlate. Prospective study criteria established by Kerr et al (NIH) are used to assess disease activity in patients with Takayasu arteritis. New onset or worsening of two or more of the following features indicates active disease: systemic features, such as fever and arthralgias (no identified cause), erythrocyte sedimentation rate, Features of vascular ischemia or inflammation, such as claudication, diminished or absent pulse, bruit, carotodynia, or asymmetric blood pressure in either upper or lower limbs (or both).

The mainstay of therapy in TA is immunosuppression, primarily with steroids or methotrexate. Remission occurs in 40% to 60% of patients. Approximately 20% of patients are resistant to any therapy. Nearly 30% of 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 stent placement -- may be needed 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 taper 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 low-dose corticosteroid 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)[5] 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 reduction 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 comorbid 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[6].

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 and the course of her TA, the care 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 clinical 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

Journal of Ophthalmology 58.2 (2010): 148–150. PMC. Web. 27 June 2017.
2. Nalini, Sharma, and Singh Ahanthem Santa. "Takayasu Arteritis with Bilateral Renal Artery Stenosis and Left Subclavian Artery Stenosis in Pregnancy." Journal of Clinical and Diagnostic Research : JCDR 9.9 (2015): QD07–QD08. PMC. Web. 27 June 2017.
3. Sadurska, E., Jawniak, R., Majewski, M., & Czekajska-Chehab, E. (2012). Takayasu arteritis as a cause of arterial hypertension. Case report and literature review. European Journal of Pediatrics, 171(5), 863–869. http://doi.org/10.1007/s00431-012-1674-z
4. Sotoudeh Anvari, M., Masoudkabir, F., Abbasi, K., Boroumand, M. A., Zarghampour, M., & Goodarzynejad, H. (2016). Takayasu's Arteritis Presenting with Headache and Peripheral Facial Palsy: A Case Report . The Journal of Tehran University Heart Center, 11(4), 195–197.
5. Tombetti, E., Di Chio, M. C., Sartorelli, S., Bozzolo, E., Sabbadini, M. G., Manfredi, A. A., & Baldissera, E. (2014). Anti-cytokine treatment for Takayasu arteritis: State of the art. Intractable & Rare Diseases Research, 3(1), 29–33. http://doi.org/10.5582/irdr.3.29
6. Saab, F., Giugliano, R. P., & Giugliano, G. R. (2009). Takayasu Arteritis in a Young Woman: A 4-Year Case History. Texas Heart Institute Journal, 36(5), 470–474.