Takayasu Arteritis: Presentation with Renal Artery Stenosis and Left Subclavian Artery Stenosis a Rare Clinical Entity in Young Women

Pankaj Shah[1], Abhinav Rastogi[2], Vibhuti Bhambha[1],Tejinder Talwar[3], Sayeed Hasan[4], Nitin Gupta[4]

[1]Post Graduate Resident, Department of Medicine, [2]Post Graduate DM Resident, Department of Cardiology, [3]Professor, Department of Medicine, [4]Assistant Professor, Department of Medicine, Maharishi Markandeshwar Institute of Medical Science and Research (Deemed to be University), Mullana, Ambala, Haryana

Abstract

Takayasu arteritis is rare form of chronic granulomatous vasculitis, it predominantly affects young adults, female in particular. We report an unusual case of Takayasu arteritis with renal artery stenosis and left subclavian artery stenosis, she presented in medicine OPD with persistent abdominal pain since 2 months, with discrepancy of blood pressure in upper limbs. She was also having renal bruit on examination. The diagnosis of takayasu arteritis was made using physical examination, Doppler upper limb and CT angiography.

Keywords: *Takayasu's arteritis, granulomatous vasculitis, Renal artery stenosis, CT Angiography.*

Introduction

Takayasu's arteritis is a rare, an autoimmune chronic vasculitis that primarily affects the large vessel i.e. aorta and its major branches. Takayasu's arteritis is referred as pulseless disease or aorta arteritis. It is a part of granulomatous arteritis, affecting large and medium sized arteries, primarily the aorta and its major branches and proximal portions of pulmonary, coronary and renal arteries. It affects predominantly young females in about 70-80% of cases (young female arteritis).Renal artery involvement is a potential presentation of TA occuring in 60% of the patients in India.

Takayasu's arteritis was firstly clinically described by Japanese ophthalmologist[1] Takaysu in 1908. Most of the cases are diagnosed in females and most common age of occurence is between 2nd decade- 3rd decade. Patients present with clinical features related to vascular insufficiency of the upper limb, weakness, claudication and fatigue in arms. On physical examination faint or absent pulse and difference of more than 10 mm Hg of systolic blood pressure between two arms[2] .

Case Profile: We present here a young female who presented with persistent abdominal pain from 2 months. On examination she was noticed to have renal artery bruit and discrepancy of Blood pressure in upper limbs. **USG Abdomen showed** Intimal thickening of abdominal aorta. No renal parenchymal changes were noted. Doppler revealed Right renal artery stenosis. **Doppler upper limbs showed** Decreased flow in left subclavian artery.

Corresponding Author:
Dr. Abhinav Rastogi
Department of Cardiology, Maharishi Markandeshwar Institute of Medical Science and Research (Deemed to be University), Mullana, Ambala, Haryana

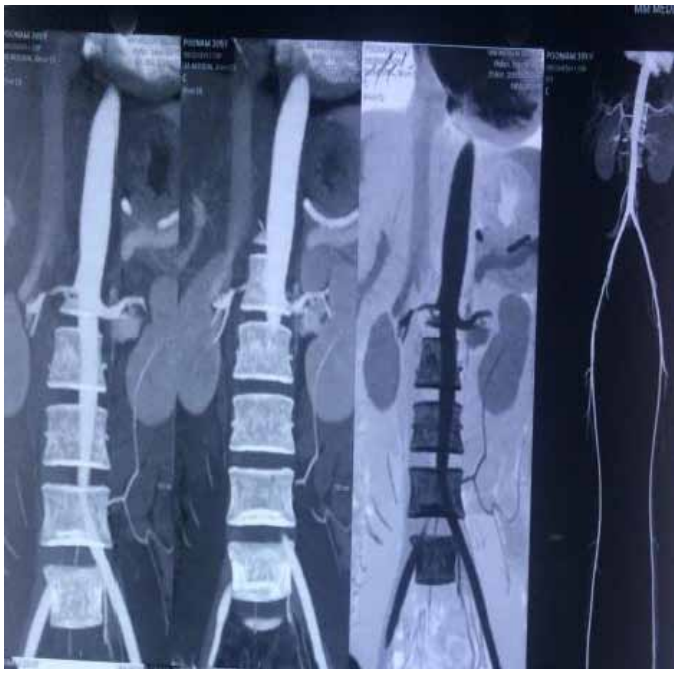

Figure 1: CT Angiography Showing– A. Thickening of upper abdominal aorta for a length of 6.5 cm involving the origin of B/L renal arteries., B. Right renal artery 70 percent stenosis, length of narrowed segment 1.4 cm

Figure 2. CT Angiogrpahy Showing– A. Diffuse circumferential inflammatory thickening and enhancement of the aortic arch., B. Diffuse thickening of the proximal part of left subclavian artery causing its mild narrowing.

American College of Rheumatology (ACR) criteria led to the diagnosis of Takayasu arteritis. She was started on oral steroids with good clinical response. She is planned for Right renal artery angioplasty on follow up.

Discussion

Takayasu Arteritis is a chronic inflammatory disease of unidentified etiology affecting large and medium vessel arteries. Although Takayasu Arteritis has been described all over the world, it occurs most commonly in Japan, India, China and Southeast Asia. The first case of Takayasu Arteritis was described in 1908 by Dr. Mikito Takayasu (Japanese ophthalmologist) as a wreathlike appearance of blood vessels in retina. Takayasu Arteritis affects 2nd-3rd decade of life. Female sex is affected 7-8 times more frequent than males. Takayasu Arteritis is manifested by granulomatous inflammation of the aorta and its major branches, leading to stenosis, thrombosis and aneurysm formation.

The vast majority of patients present with symptoms and signs of vascular insufficiency (from stenosis, occlusion, or aneurysm), systemic inflammation, or both. The most common presenting vascular symptoms are claudication (35%), diminished or absent pulse (25%), carotid bruit (20%), hypertension (20%), headache (20%) and asymmetrical arm blood pressures (15%). Aortic regurgitation, Stroke and vision abnormalities are present at onset in less than10% of patients[3]. The 1990 ACR criterion for the classification of TA [Table 1] remains the gold standard for diagnosis[4]. Ishikawa categorised clinical groups based on the natural history and complications of the disease [Table 2]. The significant complications included Secondary hypertension, retinopathy, aortic regurgitation and aneurysm development, which are graded as mild/ moderate or severe [5]. New angiographic classification of Takayasu arteritis [Table 3], categorizes based on vessel involved, aids in surgical planning but doesn't offer much in prognosis.[6]

Table 1:1990 American College of Rheumatology criteria for the classification of TA [2]

1	Age at onset ≤ 40 years
2	Limb claudication
3	Diminished brachial pulse
4	Difference of > 10 mmHg systolic pressure between arms
5	Bruit over the subclavian artery or aorta
6	Abnormal angiogram
For diagnosis ≥ 3 criteria should be present (Sensitivity: 90.5%, Specificity: 97.8 %)	

Table 2: Ishikawa clinical classification of Takayasu arteritis [3]

Groups	*Clinical features*
Group I	Uncomplicated disease, with or without pulmonary artery involvement
Group IIA	Mild/moderate single complication together with uncomplicated disease
Group IIB	Severe single complication together with uncomplicated disease
Group III	Two or more complications together with uncomplicated disease

Table 3: New angiographic classification of Takayasu Arteritis, Takayasu conference 1994 [4]

Type	*Vessel involvement*
Type I	Branches from the aortic arch
Type IIA	Ascending aorta, aortic arch and its branches
Type IIB	Ascending aorta, aortic arch and its branches, thoracic descending aorta
Type III	Thoracic descending aorta, abdominal aorta, and/or renal arteries
Type IV	Abdominal aorta and/or renal arteries
Type V	Combined features of types IIB and IV
Involvement of the coronary or pulmonary arteries should be designated as C (+) or P (+), respectively.	

Diseases which can mimic Takayasu arteritis are rheumatic (giant cell arteritis, Cogan's syndrome, relapsing polychrondritis, rheumatoid arthritis, ankylosing spondylitis, systemic lupus erythematosus, Buerger's disease, Behçet's disease), infectious (tuberculosis, syphilis,) and others (atherosclerosis, ergotism, radiation-induced damage, retroperitoneal fibrosis, inflammatory bowel disease, sarcoidosis, neurofibromatosis, Marfan'ssyndrome orcongenital coarctation). Diagnosis is mainly based on physician examination along with a high index of suspicion. If Takayasu arteritis is suspected, it is essential to palpate peripheral pulses, listen for bruits and measure blood pressure in all four limbs. Patient is screened for an acute phase response (elevated erythrocyte sedimentation rate (ESR), C-reactive protein (CRP) and normocytic-normochromic anemia[7] and definitive diagnosis is confirmed on imaging. Conventional arteriography is gold standard but its invasive technique. CT and MR angiography have replaced the conventional arteriography because of noninvasive method, they are useful in early stages,follow up and prognosis. 18F-Fluorodeoxyglucose (18F-FDG) PET imaging is increasingly being used in patients with large vessel vasculitis, allowing more precise anatomic location of metabolic activity with enhanced sensitivity, particularly in the event of moderate FDG accumulation. The principal advantage of FDG PETCT is the diagnosis of early pre-stenotic disease, an event that can be missed by intra-arterial angiography [8]. Majority of the patients have good response to steroids (oral prednisolone 1 mg/ kg/day) with 80-90% remission but about half of the patients relapse when the drug is tapered or stopped, despite good initial response [9]. Immunosuppressants (azathioprine, methotrexate, MMF, etanercept and infliximab) are used in these patients that enable the dose of corticosteroids, reduce their adverse effects and increase the rate of sustained remission. The decision for intervention is based on refractory secondary hypertension, symptomatic cardiovascular disease, severe aortic regurgitation, lesions manifesting as critical limb ischemia and aneurysms prone to rupture[10]. Common surgical interventions are angioplasty and reconstructive surgery.

Conclusion: Clinical signs to alert Takayasu's arteritis :

- Hypertension in young female
- Difference in blood pressure/ pulses between two arms.
- Claudication of extremities
- Bruit over subclavian / renal artery.

The aim of this case report is to increase the awareness of this condition among primary physicians because early diagnosis and the timely intervention or treatment can lead to better outcomes in this poorly understood and less thought clinical enigma.

Ethical Clearance: Taken from Institutional Ethics Committee (IEC).

Source of Funding: Self

Conflict of Interest: Nil

References

1. Numano F. The story of Takayasu's arteritis. Rheumatology 2002; 41: 103 - 6.
2. Fraga A, Medina F. Takayasu's arteritis. CurrRheumatol Rep 2002; 4: 30 - 3 -8.
3. Lupi-Herrera E, Sánchez-Torres G, Marcushamer J, Mispireta J, Horwitz S, Vela JE. Takayasu arteritis. Clinical study of 107 cases. Am Heart J. 1977;93:94-103.
4. Arend WP, Michel BA, Bloch DA, Hunder CG, Calabrese LH, Edworthy SM. The American college of Rheumatology 1990 criteria for the classification of Takayasu Arteritis. Arth Rheum. 1990;33:1129-1134.
5. Ishikawa K. Natural history and classification of occlusive thromboaortopathy (Takayasu's disease). Circulation. 1978;57:27-35.
6. Moriwaki R, Noda M, Yajima M, Sharma BK, Numano F. Clinical manifestations of Takayasu arteritis in India and Japan-new classification of angiographic findings. Angiology. 1997;48:369-379.
7. Nazareth R, Mason JC. Takayasu arteritis: severe consequences of delayed diagnosis. QJM. 2011;104(9):797-800.
8. Warembourg H, Devulder B. Takayasu'sarteritis diagnosed at the early systemic phase:diagnosis with noninvasive investigation despitenormal findings on angiography. J Rheumatol. 1998;25:376-377.

9. MukhtyarC, Guillevin L, Cid MC, Dasgupta B, de Groot K, Gross W. EULAR recommendations for themanagement of large vessel Vasculitis. Ann Rheum Dis. 2009;68(3):318-323.
10. Mason MC. Takayasu Arteritis: Surgical intervention. CurrOpin Rheum. 2015;27:45-52.