A CASE REPORT ON TAKAYASU ARTERITIS IN AN ADULT MALE

Divya Jyoti, Puja Dey, Rashmi Sinha, Sanjay Kumar Singh

M.D MEDICINE

RAJENDRA INSTITUTE OF MEDICAL SCIENCES

ABSTRACT

Takayasu arteritis (TA) is a rare large-vessel vasculitis that affects large arteries, mainly the aorta and its branches. It is also called pulseless disease because of diminished or absent pulses in the upper extremities of the patient. The coronary, pulmonary and renal arteries are also affected in the progression of the disease. The prevalence of the disease is more in Asian countries and it has unknown etiopathogenesis. Here we discuss the case of a 45-year-old male who presented to our OPD with complaint of monoparesis of left upper limb. Diagnosis was carried out by CT Aortogram which showed stenosis of left subclavian, distal arch of aorta, entire descending thoracic aorta. Patient was started on steroids and antihypertensive and responded well to the treatment. Early identification and treatment can help in symptom free survival.

INTRODUCTION

Takayasu arteritis (TA) is a rare, autoimmune, idiopathic, large- vessel vasculitis that affects large arteries, mainly the aorta and its branches [1,2]. In 1908 Takayasu, a Japanese ophthalmologist reported the first case of TA [2]. TA has been reported from different parts of the world, but the prevalence is more in oriental countries like Japan, India, Korea and Thailand [3]. The etiopathogenesis of the arthritis is not known, but studies are being conducted regarding the immunological and genetic aspects of the disease [3,4]. In most of the cases, irregular thickening of the walls of the aorta and its branch vessels with intimal stenosis is seen. When the aortic arch is affected, the orifices of aortic branch vessels to the upper part of the body could be narrowed markedly. The coronary, pulmonary and renal arteries are also affected in the disease process [5]. The clinical presentation of TA varies depending on the blood vessels involved. Diminished or absent pulses of upper extremities are found in most of the patients, hence also called pulse less disease[6]. Early reviews suggest that the disease is confined to the female population, but now it has been reported that males are also affected, but disease manifestations may vary among populations [2] .Here we report the case of a 45 year old male who came to our OPD for evaluation of monoparesis of left upper limb .

CASE REPORT

A 45 year old male presented to our OPD on 7th July 2022 with complaints of dizziness, difficulty in holding weights in left hand. He complained of experiencing dizziness , light headedness for the past 6 months, simultaneously he also started experiencing weakness in his left arm which was gradually progressive, he had difficulty in carrying heavy weights in his hands. Over months his weakness progressed such that now he was not able to lift any object or carry any work from his left hand. All these symptoms started about 6 months back. However he did not complain of fever, weight loss, malaise. He did give history of excruciating pain in his left hand occasionally. He also complained of pain and tingling sensation in his left arm about 5 years back for which he consulted local doctors and symptoms improved with injectable Vitamin B_{12}. One month back he complained of slurring of speech which gradually improved with local treatment. However, he did not go for any investigations at that time. His main concern for visiting our OPD was his progressive weakness of his left limb.

On examination, patient had normal built with slightly reduced muscles mass in his left hand as shown in Figure 1. BP as recorded by manual sphygmomanometer was 170/80mmHg, 120/80mmHg in his right and left hand respectively. Pulse rate being 100/min and 60/min in right and left hand respectively. Peripheral pulses of upper limb was palpable on both side but brachial and radial pulse on left side was feeble. However lower limb pulses like popliteal, dorsalis pedis, femoral pulses were absent bilaterally. His left upper arm circumference (10 cm above olecranon process) was 24cm and 27 cm in left and right hand respectively. Forearm circumference (measured from olecranon) was 24cm and 22 cm in right and left hand respectively. Carotid bruit was heard bilaterally.

All routine investigations including CBC, ESR, CRP, RFT, LFT, TSH, RBS, Lipid profile, ANA , RA was sent. We also did an USG whole abdomen ,echocardiography, non-contrast CT of brain and finally went for CT aortogram. CBC, RFT, LFT, TSH, RBS were in normal range with mild anemia noted. ESR, CRP were raised. Dyslipidemia was noted with Triglyceride- 320mg/dl, Total cholesterol 144.14, ANA profile and RA factor was negative. Echocardiography revealed concentric LVH with EF 65%. NCCT Brain suggestive of few basal ganglia infarcts. Ultrasound was normal. A CT Aortogram was done which showed significant stenosis of distal arch of aorta, descending thoracic abdominal aorta and left subclavian artery. Mild narrowing of proximal left carotid artery and right brachiocephalic artery was also noted as shown in Figure 2.

He was started on oral prednisolone at a dose of 1mg/kg body weight for 4 weeks, once daily morning dose. We attempted to taper the dose steadily to achieve the dose of 20mg/dl by 3 months as he showed clinical improvement. Antihypertensives were also started simultaneously. We are following the patient on monthy basis.

FIGURE 1: Mid-arm circumference

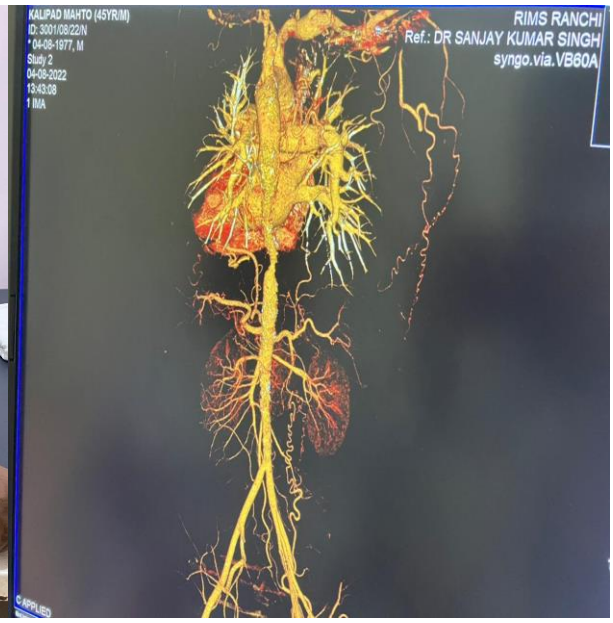

FIGURE 2: CT Aortogram of the patient

DISCUSSION

TA is a chronic, progressive, granulomatous vasculitis which commonly occurs in the second or third decade of life[2]. The most common clinical features are significantly low blood pressure (BP) and weaker pulses in the upper extremities associated with coldness or numbness of the fingers[4,5]. The clinical course of the arteritis is usually divided into an active inflammatory phase and chronic phase. The early active phase is characterised by the systemic disease with signs of fever, malaise, loss of appetite, weight loss, headaches, dizziness, arthralgia, skin rashes, etc which lasts for weeks to months. The most common presenting vascular symptoms are claudication (35%), reduced or absent pulse (25%), carotid bruit (20%), hypertension (20%), carotidynia (20%) light-headedness (20%), and asymmetrical arm blood pressures (15%). Stroke, aortic regurgitation, and visual abnormalities are present at onset in less than 10% of patients[9]. The clinical manifestations in the late, chronic phase may vary depending on the location of the arterial stenosis. The arteries involved are coronary arteries, pulmonary arteries, renal arteries, aortic arch and aortic branches and clinical manifestations include ischemic heart disease, dyspnea, ischemia of stomach and intestines, arterial hypertension, congestive heart failure, etc[6,7] . According to American College of Rheumatology, at least three out of the following six criteria should be satisfied for a definite diagnosis of TA. These criteria are :

1. age under 40 at disease onset
2. claudication of extremities
3. decreased brachial artery pulse
4. blood pressure difference more than 10 mmHg between arms
5. a bruit over subclavian arteries or aorta and angiogram abnormalities
6. occlusion or narrowing in the aorta or its main branches[5] .

Considering this patient, 4 out of 6 criteria were met. The patient also had raised ESR, CRP and anemia which

reflects the underlying inflammatory process. But these laboratory investigations are usually nonspecific for diagnosis of TA. In a study conducted by Park MC *et al.* in South Korea, 23 % of the patients showed normal laboratory results even with active disease[5,8]. There is no diagnostic serological test for TA and the symptoms of the disease are constitutional like fever, malaise, loss of appetite, weight loss, headaches, dizziness, arthralgia in the initial phase. This makes difficulty in early diagnosis of the disease[1].Classification of the disease based on the vessel involved has been shown in Table 1.

Table 1: New angiographic classification of Takayasu arteritis, Takayasu conference 1994

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

According to this classification system, the involvement of the coronary or pulmonary arteries should be designated as C (+) or P (+), respectively[2,10].

Our patient was diagnosed as TA Type V (P+) as there was concentric wall thickening of aorta, renal artery stenosis and complete stenosis of bilateral descending pulmonary artery in CT aortogram.

Diseases which can mimic TA are rheumatic (giant cell arteritis, Cogan's syndrome, relapsing polychrondritis, ankylosing spondylitis, rheumatoid arthritis, systemic lupus erythematosus, Buerger's disease, Behçet's disease), infectious (syphilis, tuberculosis) and others (atherosclerosis, ergotism, radiation-induced damage, retroperitoneal fibrosis, inflammatory bowel disease, sarcoidosis, neurofibromatosis, congenital coarctation or Marfan's syndrome). Diagnosis is mainly based on physician awareness, along with a high index of suspicion. If TA is suspected, it is essential to palpate peripheral pulses, listen for bruits, and measure blood pressure in all four limbs. Patient should be evaluated for evidence of an acute phase response (elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) and normocytic-normochromic anaemia, and the diagnosis is confirmed on imaging.

Management of systemic and vascular inflammation is carried out using corticosteroids and immunosuppressive agents. Treatment may be initiated with high dose corticosteroids, such as prednisolone. Methotrexate, Cyclophosphamide, azathioprine are used for immunosuppression. Cyclophosphamide is usually used for the treatment of systemic vasculitis when the condition is severe or life- threatening. In chronic stages of TA, endovascular revascularization procedures or interventions like balloon angioplasty or stent placement should be considered. Procedures should be undertaken only after the suppression of inflammation in the affected arteries. Surgical procedures carry risk and success rate depends upon the location and stage of stenosis of blood vessel[6,11].

CONCLUSION

TA is a rare clinical entity, can present in wide variety of ways, many with a typical history of other conditions. The use of steroids along with immunosuppressant like methotrexate is a cost-effective regimen and has a good initial response. However, chances of relapse should be kept in mind while treating the patient. Regular follow up is necessary for the assessment of the disease progression.

REFERENCES

1.Saab F, Giugliano RP, Giugliano GR. Takayasu arteritis in a young woman A 4-year case history. Tex Heart 2009;36:470–4.

2.Johnston SL, Lock RJ, Gompel MM. Takayasu arteritis: a review. J Clin Pathol 2002;55:481–6.

3.Jain S, Pondaiah SK. Takayasu's arteritis: a review of epidemiology and etiopathogenesis. Indian J Rheumatol 2015;10:S22–9.

4.Das D, Mondal KK, Ray B, Chakrabarti A. A case of unusual presentation of Takayasu's arteritis. Indian J Ophthalmol 2010;58:148–50.

5.Al-Bishri J. Takayasu's arteritis: a review article. Br J Med Med Res 2013;3:811-20.

6.Khan MAM, Banoo H. A case report of takayasu's arteritis. Med Today 2012;24:79-81.

7.Kim HJ, Suh DC, Kim JK, Kim SJ, Lee JH, Choi CG, *et al.* Correlation of neurological manifestations of Takayasu arteritis with cerebral angiographic findings. Clin Imaging 2005;29:79-85.

8.Park MC, Park YB, Jung SY, Lee KH, Lee SK. Anti-endothelial cell antibodies and antiphospholipid antibodies in Takayasu's arteritis: correlations of their titers and isotype distributions with disease activity. Clin 2006;24(2, Suppl 41):S10-6

9.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.

10.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;48:369–79.

11.Keser G, Direskeneli H, Aksu K. Management of Takayasu arteritis: a systematic review. Rheumatology (Oxford) 2014;53:793-801.

12.Sharmila Mohan, R Lakshmi. A case report on takayasu's arteritis. Int J Pharm Pharm Sci 2017;9(3):296-297.