

**A CASE REPORT OF A YOUNG FEMALE WITH TAKAYASU ARTERITIS IN STENOTIC STAGE**Pradeep Sharma<sup>1</sup>, Atul Gupta\*<sup>2</sup> and Rajesh Kumar<sup>3</sup><sup>1</sup>Medical Officer (Specialist), DHS Office, Kasumapatti- Shimla, Himachal Pradesh, India.<sup>2</sup>District Programme Officer, District Kullu, Himachal Pradesh, India.<sup>3</sup>Medical Officer (Specialist), Regional Hospital, Kullu, Himachal Pradesh, India.**\*Corresponding Author: Dr. Atul Gupta**

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**ABSTRACT**

Takayasu arteritis (TA) is a disease of chronic granulomatous inflammation of the aorta or its branches. This inflammation will eventually cause fibrinoid necrosis, and fibrosis, narrowing and aneurysm of the vessel wall. Takayasu arteritis in early phase has non-specific complaints leading to delayed diagnosis in stenotic phase. It can cause debilitating complications without treatment. Here the author presents a case of TA with detailed history and steps in diagnosis of the disease.

**KEYWORDS:** Takayasu arteritis, Chronic granulomatous inflammation, Large vessel vasculitis.**INTRODUCTION**

Takayasu arteritis (TA) is a rare systemic disease more frequently found in Asian countries but is prevalent all over the world.<sup>[1]</sup> TA is a disease that can cause debilitating complications if left untreated. Therefore, early recognition and prompt treatment are key to managing patients with TA. Early diagnosis, however, is challenging because of the disease's unspecific early signs and symptoms.<sup>[2]</sup> Herein, we present a case of a 20-year-old woman from north India, who was diagnosed with TA.

**CASE**

A 20 year old unmarried female with no previous comorbidities presented in the OPD with fever of 7 month duration. The fever was low grade, intermittent documented up to 101°F, not associated with chills and rigor. Fever was associated with significant weight loss and decreased appetite. There was no localizing

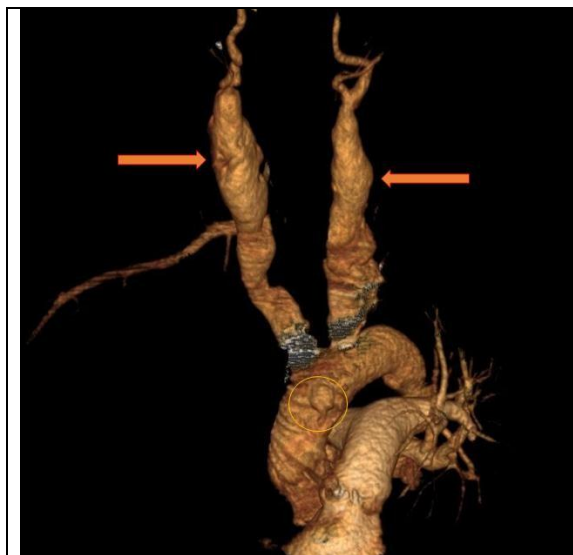
symptom of fever other than dry cough. Fever partially responded to antipyretics and antibiotics. Initially patient was evaluated for tuberculosis. Sputum was sterile and mantoux was negative, however patient was started on empirical ATT by a private practitioner. Fever and cough resolved after one month of ATT start but there was no improvement in appetite and weight gain. Two months into the illness patient noticed pain in left arm which used to increase with activity and relieve on rest. Pain was not associated with restriction of movement, weakness and any sensory deficit. Patient noticed swelling on both sides of neck which was soft in consistency and pulsatile. There was no visual disturbances, headache, vertigo, dizziness, chest pain. On examination there was pallor and arterial pulses were not palpable in left upper limb. On neck examination there were oval shaped, soft, pulsatile swelling on both side and bruit was heard over left supraclavicular region (Figure 1). Systemic examination was unremarkable.



**Picture 1: Swelling on neck, which was pulsatile; a bruit was heard on auscultation.**

Laboratory evaluation revealed anemia, thrombocytosis, raised ESR and high CRP titre. Autoimmune workup showed 1+ ANA cytoplasmic reticular pattern and possibility of vasculitis was kept. On USG Doppler bilateral common carotid artery aneurysm were seen. CT angiography (Head, neck and thorax) showed circumferential mural thickening with dilatation of both common carotid arteries with diffuse circumferential mural thickening involving arch of aorta and significant

narrowing of left subclavian artery (Figure 2). 2D Echocardiography revealed moderate left ventricular dysfunction with mild aortic regurgitation. All these findings were suggestive of large vessel vasculitis and final diagnosis of Takayasu arteritis was kept. Patient was started on oral steroids and methotrexate, improved clinically and was referred to higher institute for further management.



**Figure 2: CT angiography showing circumferential mural thickening with dilatation of both common carotid arteries.**

## DISCUSSION

The presented case of a 20-year-old woman presented with low grade, recurrent fever, claudication of arm, and swelling on both side of the neck which produced bruit on auscultation. These were caused by the narrowing of the subclavian artery and the common carotid arteries. Differential diagnoses, in this case, include other primary vasculitis; giant cell arteritis, polyarteritis nodosa, Henoch-Schonlein purpura, granulomatosis with polyangiitis and eosinophilic granulomatosis with polyangiitis. In our patient, the disease affected both the superior branches of the aorta, as supported by the physical examination and investigations. This clinical presentation and imaging suggested TA. The diagnosis of TA has always been a challenge. This delay of diagnosis can be attributed to a few factors. First, both the early and late manifestation of TA is not specific.<sup>[3]</sup> Laboratory findings include acute-phase reactant markers or ESR, which are not specific for TA. Tools to diagnosis like CT angiography and Echocardiography are expensive and not readily available especially in rural areas.<sup>[4]</sup> Therefore, the most crucial factor of early diagnosis is the physician's awareness of the clinical findings. India is a tropical country and is endemic to many infectious diseases, such as tuberculosis. Therefore, it is more likely that infectious disease is suspected first in a patient with fever. The case presented here also had non-specific symptoms which delayed the diagnosis and was diagnosed in late stenotic phase. The treatment for TA consists of immunosuppressant therapy with high dose corticosteroid. It is recommended to add a

second immunosuppressive agent such as methotrexate, cyclosporine A, tacrolimus, azathioprine, cyclophosphamide and mycophenolate mofetil.<sup>[5,6]</sup> TA is a disease of chronic granulomatous inflammation of the aorta or its branches. This inflammation will eventually cause fibrinoid necrosis, and fibrosis, narrowing and aneurysm of the vessel wall. TA manifestation can be divided into two phases: an initial phase of unspecific findings, for example, fever, malaise and headache, and a late stenotic phase characterised by findings such as hypertension, claudication, pulselessness, bruit and angina.<sup>[7,8]</sup> If left untreated, TA will eventually cause irreversible target organ damage and severe morbidity.

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