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## Case report

### TAKAYASU'S ARTERITIS: AN UNUSUAL PRESENTATION OF A RARE DISEASE

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#### ABSTRACT

Takayasu's arteritis, also called tak, aortic arch syndrome, pulse less disease or occlusive thromboaropathy is a rare chronic, progressive, autoimmune, idiopathic disease involving inflammation in the walls of the largest arteries in the body: the aorta and its main branches that affect primarily adolescent girls and young women. It most often occurs in people ages 15–40 years, but sometimes affects younger children or middle-aged adults. Here is a case of a young girl diagnosed with takayasu's arteritis whose initial complaints were predominantly high grade fever and malaise with minimal signs of vascular insufficiency.

**Keywords:** Takayasu's arteritis, American rheumatological society, Fever of unknown origin, CT angiography

#### INTRODUCTION

Takayasu's arteritis (TA), also known as aortoarteritis and pulse less disease, is a rare condition. The cause of TA is unknown. It is a form of granulomatous arteritis, which affects large- and medium sized arteries, primarily the aorta and its large branches as well as proximal portions of pulmonary, coronary, and renal arteries. It was first described in 1908, in a Japanese patient with retinal abnormalities<sup>1,2</sup>. TA affects women eight times more frequently than men. Here is discussing a case of a girl who was diagnosed with TA at 23 years of age, the course of the disease during the previous 3 years prior to diagnosis, and different treatments for the rare disorder.

#### CASE REPORT

In September 2014, a young 23 year old girl presented with only complaint of marked generalized fatigue. On enquiry, she revealed that she had a high grade fever 3 years back along with joint pains (which were not marked) for which she was given a prolonged course of antibiotics but with minimal relief. She was then evaluated for infectious pathology but most of her tests were unremarkable except for her erythrocyte sedimentation rate (ESR) which was 189 mm/hr and her complete blood count which showed severe

anaemia (7gm %) of dimorphic origin. She was labeled as fever of unknown origin (FUO) and started on anti-tubercular therapy (ATT) without any evidence of the same. Meanwhile, she visited a higher centre where her ATT was stopped after 1 month, and again started on intravenous antibiotics but remained febrile. She was intensively investigated there, but apart from persistently high ESR and anaemia, most of the tests, including rheumatoid factor, antinuclear antibodies (ANA), thyroid profile, x-ray chest and echocardiography came out to be negative. Finally, she was discharged on tab doxycycline 100mg twice a day for 40 days. Her fever and joint pains subsided over a period of time. However, she continued to have persistently raised ESR and moderate to severe anaemia with relapsing and remitting complaints of generalized fatigue and weakness until she presented to me in September 2014 with the above mentioned complaints. She denied any current history of fever, joint pains, claudication, dizziness, headache, visual blurring and breathlessness.

On physical examination, the patient was comfortable and was not in any distress. She was a febrile with a regular pulse rate of 86 beats per minute with unequal radial pulses, left radial pulse being markedly

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diminished. Blood pressure was 110/90 mm Hg in the right arm while it was difficult to auscultate on left arm with a blood pressure of approximately 90/70 mm Hg, and respiratory rate of 16 breaths per minute. Her cardiovascular examination revealed bilateral carotid and subclavian artery bruit, more prominent on the left side. Cardiac auscultation revealed normal sounds. There was no abdominal aortic bruit. The femoral and pedal pulses were normal, equal and palpable bilaterally.

The initial workup included complete blood cell count (CBC), basic metabolic panel, coagulation profile, erythrocyte sedimentation rate, C-reactive protein, hepatitis panel and tests for human immunodeficiency virus. She had a white blood cell (WBC) count of 6000, haemoglobin (Hb) of 10.4 gm% and platelets (PLTs) of 1.94 lakhs/cumm. Her basic metabolic, renal and liver function tests as well as coagulation panel were unremarkable. Serology for antiHIV, antiHCV and HBsAg was negative. Her Doppler abdomen was unremarkable. Her inflammatory biomarkers such as C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) were both elevated, with >5ug/ml and 35mm/h (0 to 20mm/h) respectively. Her electrocardiogram (EKG) and echocardiogram were both normal.

Carotid Doppler suggested diffuse intima-media complex thickening involving both common carotid arteries (L>R). Flow through left brachial artery showed parvus and tardus kind of spectral waveform s/o proximal occlusion. CT angiography showed diffuse intima-media thickening of the wall of arch and descending aorta. Similar diffuse thickening was also noted of the walls of carotid artery bilaterally with complete occlusion noted at the origin of left subclavian artery; features suggestive of takayasu's arteritis.

## DISCUSSION

Although TA has been described worldwide, it occurs most commonly in Japan, China, India and Southeast Asia. It is an autoimmune disease involving the arterial walls of large arteries, causing Panarteritis<sup>3</sup>. Takayasu arteritis is a systemic disorder that affects multiple organs. The diagnosis of TA can be a challenge, especially in its initial phases. Clinicians divide TA into two phases: Systemic and Occlusive. Patients may have features of both phases at the same time. In the early or the systemic phase of TA, clinical features

include fatigue, low grade fever, weight loss and lethargy. As the disease progresses into the occlusive phase, vascular involvement and insufficiency become apparent due to dilatation, narrowing and occlusion of the main vessels like the aorta and its branches<sup>4</sup>. These may include pain in limbs, claudication, dizziness upon standing up, headaches, and visual disturbances. Lung involvement is a rare presenting feature, but involvement of the pulmonary arteries has been reported.<sup>5</sup>

The American Rheumatological Society considers three of the following six criteria necessary for a definite diagnosis of Takayasu's disease.<sup>6</sup> The presence of any three or more criteria yields a sensitivity of 90% and a specificity of 97.8%.<sup>7</sup>

1. Onset before 40 years
2. Claudication of the extremities
3. Decrease in the brachial pulse in one or both arms
4. A difference of 10 mm Hg or more in blood pressure measured in both arms
5. Audible bruit on auscultation of the aorta or subclavian artery
6. Narrowing at the aorta or its primary branches on arteriogram

However, in clinical practice, the diagnosis of TA is almost always secured by an imaging procedure that demonstrates the characteristic abnormalities of the aorta and its major branches. The current patient fulfilled 5/6 criteria (including an imaging procedure) at the time of the diagnosis.

Unfortunately, the diagnosis of TA is often delayed. One of the reasons for this is that some patients have striking features of inflammation that camouflage or overshadow the somewhat more familiar vascular abnormalities. Indeed a few patients with TA present chiefly with FUO as in this case. Most of these patients have other, albeit subtle, manifestations of TA such as bruits, diminished pulses, unequal arm blood pressures or aortic regurgitation. Thus the most frequent impediment to a speedy diagnosis is a physician's failure to consider TA in the differential diagnosis. Delays in diagnosis can be reduced by carefully searching for unequal or absent upper extremity pulses and by listening for bruits not only over the carotid arteries, but also above and below the clavicle for subclavian artery bruits and over the abdomen and flanks for renal and other mesenteric artery bruits.

The differential Diagnosis of TA includes infection (tuberculosis, mycoses and syphilis), congenital collagen disorders (Ehlers-Danlos, Marfan's syndrome), Fibrous Dysplasias (FD), and rheumatic diseases (Giant cell arteritis, Rheumatoid arthritis, cogan's syndrome, SLE, Buerger's disease, Sarcoidosis, and Spondyloarthropathies). Imaging is very useful in differentiating most of the possible diagnoses except for giant cell arteritis. Giant cell arteritis like TA involves large arteries showing granulomatous vasculitis on histologic examination. A distinction can be made based on the age of the patient and distribution of lesions.<sup>8,9</sup>

The course of the disease is variable, and although spontaneous remissions may occur, TA has been most often chronic and relapsing. Corticosteroids are the cornerstone of treatment of active TA. Prednisolone, at a dose of 0.5 to 1 mg/kg per day, is indicated for the treatment of active disease. Open trials have suggested that weekly oral methotrexate is a moderately effective corticosteroid sparing agent.<sup>10</sup> TA is the form of vasculitis most frequently requiring revascularization procedures.<sup>11,12,13</sup> In the presence of symptomatic stenotic or occlusive lesions, endovascular revascularization procedures like bypass grafts, patch angioplasty, endarterectomy, percutaneous transluminal angioplasty, or stent placement should be taken into consideration<sup>14</sup>.

## CONCLUSION

Takayasu arteritis is a relatively rare disease with various and sometimes initially only systemic clinical manifestations, such as FUO leading to delay in diagnosis as in our case. As early, appropriate diagnosis and initiation of proper therapy could avoid further progression and reduce complications of the disease, many of these delays can be prevented by remembering that TA should be included in the differential diagnosis of any person younger than 40 years who presents primarily with FUO, musculoskeletal and other symptoms of systemic inflammation.

**Conflict of Interest: Nil**

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