CASE REPORT

TAKAYASU ARTERITIS IN A YOUNG FEMALE PRESENTING WITH PYREXIA OF UNKNOWN ORIGIN IN THE PRE-PULSELESS PHASE

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Takayasu arteritis is a rare chronic large vessel vasculitis that mainly occurs in young females with a higher prevalence in Asian population. This disease, in its early stages, often goes undiagnosed because of its non-specific and vague symptoms. This poses great difficulty for doctors to accurately make the diagnosis. One of its initial presentations is 'pyrexia of unknown origin (PUO). PUO precedes the pulseless stage of Takayasu arteritis. Here we're going to do a case report on a 26-year-old female patient presenting with PUO as the pre-pulseless stage of Takayasu arteritis. The presentation as PUO in the absence of pulse deficits is very uncommon and has been very underwhelmingly reported.

Keywords: Takayasu arteritis; Pyrexia; Young female

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INTRODUCTION

Different disease processes like infections, inflammation, autoimmune diseases, neoplasia and metabolic disorders can present with fever. Pyrexia of unknown origin (PUO) is defined as a clinically documented persistent temperature of 101°F or higher on several occasions lasting for at least 3 weeks, accompanied by an unremarkable diagnosis despite routine and appropriate investigations, for at least 1 week admitted in a hospital or in outpatient clinic setting. Causes for PUO can be classified as infection, malignancy, noninfectious inflammatory disease, or miscellaneous.[1]

Over 200 different diseases have shown to be presented with PUO, with around 51% of them remaining undiagnosed. Among the different causes infections are responsible for 17–35% of cases, inflammatory causes 24–36%, neoplastic contributing 10–20% and miscellaneous causing 3–15% of the PUO cases.[2]

One of the diseases presenting with PUO is Takayasu arteritis. It is a disease, causing inflammation of large sized blood vessels, mainly the aorta and its branches like the carotid and subclavian arteries. Young females (age between 10–40) are among the most vulnerable patient population. It is more commonly found in Asian populations. Due to its rarity, it has been difficult to report the incidence and prevalence of the disease, but according to a study the estimated incidences were 1–2 per million in Japan, 2.2 per million in Kuwait, 0.5 to 1.5 per million in Europe. The pathology of TA is poorly understood. It is believed to be an autoimmune disease with multifactorial triggers like environmental, genetic and immunological factors. Presentation of TA initially include fever, headaches, malaise, weight loss, arthralgias, dizziness and generalized weakness. During the course of the disease the patient may present with reduced or absent pulses, vascular bruits, hypertension and retinopathies, eventually leading to complications like aneurysms and stroke.

According to American college of rheumatology (ACR) 1990's classification, a patient is diagnosed as TA if he fulfills at least three out of six criteria, and these criteria have a sensitivity of 90.5% and a specificity of 97.8%. ACR criteria include the age of onset <40 years, claudication of extremities, decreased brachial artery pulse, blood pressure difference/greater than 10 mm Hg, vascular bruits over a subclavian artery or aorta, and arteriogram abnormality.[3]

We will now present a case of a female with Takayasu arteritis presenting as PUO, highlighting the diagnostic difficulties and the clinical progression of the disease.

CASE REPORT

A 26-year-old female with no significant past medical history presented in march 2024 with intermittent fever (ranging from 102–104 F) and night sweats since past 5 months in a tertiary care hospital in Islamabad, Pakistan. Her fever progressed from low grade to high grade. The fever was associated with chills and rigor and was relieved by over-the-counter anti pyretic medicines. She had undergone treatment with multiple antibiotics before, but the fever did not resolve. Suspecting enteric fever, she was admitted and prescribed parenteral meropenem for 6 days and discharged. The following month, she presented again with the same recurring symptoms. Her lab work was done and her QuantiFERON TB gold test (QTB) came back positive. She was started on anti-tuberculous

treatment (ATT). On subsequent visits, after 3 months, she started reporting of pain in back of the neck and upper back. The pain was not associated with rest or activity. She also complained of frequent vomiting, loss of appetite and weight loss. There was no cough or sore throat. Patient had abdominal pain but no constipation, diarrhea, vomiting or blood in stool. Her imaging was done and found unremarkable. Her abdomen ultrasound and chest x ray were clear. CT abdomen and pelvis were unremarkable. No vegetations were seen in transthoracic echocardiogram (TTE). Autoimmune workup was unremarkable. Her lab work showed elevated erythrocyte sedimentation rate (ESR) and C reactive protein (CRP) along with neutrophilic leukocytosis and anemia as shown in Table-1. She was started on oral steroids. Bone marrow biopsy was also carried out and found unremarkable. Throughout her visits she was mildly hypotensive, with her blood pressure recorded in the range of 100–110/ 70–60. On her 6th month follow up visit, her posterior cervical lymph nodes were palpable but sub centimetric. On her 9th month follow up she complained of persistent back and neck pain along with fever and lethargy. On examination her left radial and brachial pulse were weak and right sided carotid bruit was auscultated and thrill was felt. Her blood pressure was recorded as 170/100 in the right arm and 100/70 in the left arm. Her CT aortogram showed narrowing of aorta, extending to the branches, with thickened aortic walls and multiple saccular aneurysms. The patient's CT angiography in Figure-1 showed findings consistent with Takayasu arteritis such as irregular luminal narrowing of aortic arch, descending thoracic aorta and abdominal aorta along with segmental stenosis of the left subclavian artery, brachiocephalic artery and right common carotid artery. Based on the CT findings along with the elevated inflammatory markers, a diagnosis of Takayasu arteritis was made. She was advised to continue her steroid treatment and was referred to rheumatology for review.

Table-1: Relevant investigations

Investigations	Results
White blood cell count/µl	11.2
Hemoglobin (gm/dl)	7.7
Platelet count/ µl	762000
LDH u/l	132
Creatinine mg/dl	0.47
CRP	126
ESR	150
MTB DNA	Negative
Anti-HIV	Non-reactive
Brucella IgG/IgM	Negative
ANA	Negative
P-ANCA	Negative
C-ANCA	Negative
Blood c/s	Negative
Urine c/s	Negative
Bone marrow biopsy	Negative
Echo	No vegetations

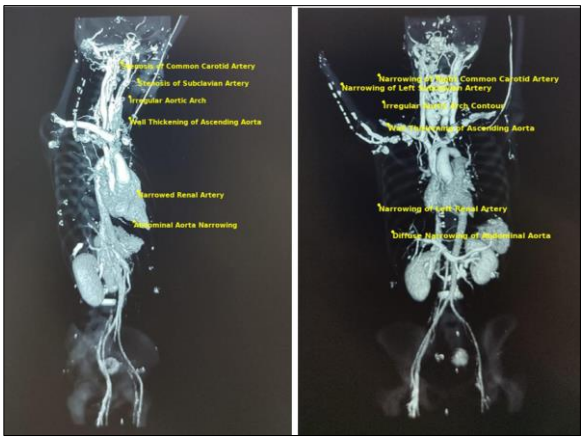

Figure-1: CT Angiography of the head, neck, chest, abdomen and pelvis in coronal reconstruction front and lateral view

DISCUSSION

Takayasu arteritis is chronic large vessel vasculitis which mostly affects aorta and its major branches. This disease often manifests in two phases; The initial phase involves non-specific inflammatory symptoms and a later phase characterized by vascular insufficiency leading to diminished or absent pulses, hence the name 'pulseless disease.

In our case, the patient presented with fever, night sweats, later back of the neck and upper back pain, epigastric and mid-abdominal pain, accompanied by weight loss. While going through other literature, we found a case report of a male in his 20s in India presenting with fever, weight loss, abdominal pain and blood in his stool.[4] Another case of a 32-year-old female in India presented with recurrent fever chest pain and pleural effusion. Angiography revealed right subclavian artery and carotid artery occlusion.[5]

The absence of specific clinical symptoms in the early phase of this disease poses a significant difficulty in diagnosis. Routine laboratory tests may reveal elevated inflammatory markers but these are not specific to TA. In this case ESR and CRP were elevated. MRI showed disc bulges at T1-T2 and T2-3, streak of fluid in the left pleural cavity with trace perinephric fluid as well. The patient was treated with anti-tubercular therapy because of positive family history but there was no improvement. Although there was no association of TB with Takayasu arteritis in our patient, but a study showed that microbial agents, including TB bacteria, could initiate or sustain Takayasu arteritis through mechanisms such as molecular mimicry, particularly between mHSP65 and hHSP60.[6] Then after a few months, she presented with right neck pain and a right carotid bruit. On further examination, there was weak radial and brachial

pulses on the left side. On CT aortogram, there was narrowing of aorta with extension to major branches (suggestive if TA).

She was treated with steroids leading to improvement of her condition. The 2021 American College of Rheumatology guidelines suggested a combination of non-glucocorticoid and glucocorticoid use in patients with active TA rather than glucocorticoids alone.[7] Methotrexate and azathioprine are used as initial non-glucocorticoid agents.

According to our knowledge, this is the first case report of Takayasu arteritis presenting with PUO in Pakistan, and signifies that Takayasu arteritis may be considered as a differential in patient presenting with similar symptoms and findings.

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