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Report

Pakistani Origin Takayasu Arteritis: A Case Report with a Brief Review from Asymptomatic Presentation Till Diagnosis and Clinical Management

Shahzad Ahmad[1], Muhammad Sajjad Ali Khan[1], Pardis Bahadori[2], Aimal Khan[1], Wiqar Ahmad[1] Muhammad Ilyas[1], Komal Qayyum[1], Muhammad Saeed[3], Musa Bin Bashir[4, *]

[1]General Medicine and HDU/ICU Department, Northwest General Hospital and Research Center, Peshawar, Pakistan

[2]School of Medicine, Xi'an Jiaotong University, Xi'an, China

[3]Department of Internal Medicine, Al Hussain Medical Service, Sargodha, Pakistan

[4]Department of Cardiology, Second Affiliated Hospital, Xi'an Jiaotong University, Xi'an, China

Email address:

Dr.usmle2021@gmail.com (Musa Bin Bashir)

[*]Corresponding author

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Abstract: Takayasu's arteritis is a rare chronic granulomatous vasculitis that primarily affects the aorta and its branches. It is estimated that this disease impacts approximately 2.6 million individuals every year, with a higher prevalence among women in their second or third decade of life. In this case report, we describe the presentation and management of a 22-year-old female patient who initially sought medical attention from a neurologist due to upper and lower back pain. Initially, the patient's symptoms were managed conservatively. However, a few months later, she presented to the medical department with a high-grade fever, syncopal episodes, generalized body aches, and swelling in her right arm. The findings from the computed tomography angiography (CTA) revealed diffuse thickening of the entire thoracic and abdominal aorta's walls, long segment luminal narrowing of the proximal left common carotid artery and left subclavian artery, as well as diffuse thickening of the abdominal aorta's wall. This case highlights the importance of recognizing that Takayasu's arteritis can present in an unusual manner. Early diagnosis and management are crucial steps towards providing appropriate care for patients. Due to the potential involvement of various arterial segments, Takayasu's arteritis can manifest with diverse symptoms and complications. Therefore, clinicians should maintain a high index of suspicion when encountering patients with unexplained symptoms, particularly those involving the aorta and its branches. In conclusion, the presented case emphasizes the need for prompt recognition and intervention in Takayasu's arteritis. By increasing awareness of this rare condition and its atypical presentations, healthcare professionals can ensure timely diagnosis and appropriate management, ultimately improving patient outcomes and quality of life. Further research is warranted to enhance our understanding of this complex disease and optimize its management strategies.

Keywords: Takayasu Arteritis, Autoimmune, Autoimmune in Females, Vascular Disease, Large Vessel, Vasculitis

1. Introduction

Takayasu arteritis (TA), also known as "pulseless disease," is a primary systemic vasculitis affecting the pulmonary, coronary, medium- and large-sized arteries, as well as the aorta and its branches.

It is a chronic inflammatory condition of uncertain etiology characterized by granulomatous vasculitis, which causes the arteries affected to thicken, enlarge, stenosis, and/or produce aneurysms [1, 2]. Additionally, the signs and symptoms include angiodynia, claudication, peripheral pulselessness, murmurs, ischemic stroke, myocardial

infarction, severe systemic arterial hypertension, etc. and are brought on by systemic inflammation or ischemia of an organ or limb1. 80% of TA sufferers are female, with females typically being more affected than males. The ratio of women to men varies, nevertheless, from 9:1 in Japan and 6.9:1 in Mexico to 1.2:1 in Israel[1]. TA can be fatal and is linked to substantial morbidity. A monophasic and self-limited condition affects about 20% of patients, while other patients may have a progressive or relapsing/remitting sickness. Additionally, the overall 10-year survival rate for this illness is around 90%, however this number can drop in the case of serious complications[2]. A case of TA in a female patient, age 21, is presented here.

2. Case Report

A 22-year-old Asian woman presented to the outpatient department with acute febrile illness, generalized body aches, syncope and right arm swelling. She was then admitted to medical high dependency unit (HDU). She had a history of stillbirth (2^{nd} pregnancy loss) few months back. After few months of pregnancy loss, she developed weakness with whole back pain for which visited neurologist. Patient visited gynecologist for a regular checkup and got diagnosed as having Toxoplasmosis for which was treated with azithromycin and she was recommended an MMR vaccination. After the MMR vaccination she developed fever which had spike early morning with rigors and chills. she complained that her condition worsened after the MMR vaccination. She was having syncopal episodes while doing her house chores as well as bending over to pick something up. She did her ESR and CBC on a primary physician's advice; the ESR was raised (42mm/hr.), MCV was low (77mg/dl) and platelets were high (573mg/dl). She was complaining of fatigue, weakness and upper limbs cramps and came to see Gynecologist in Northwest General Hospital. She was referred to medical specialist who admitted her in the hospital for further investigation and management. On physical examination, she was afebrile with blood pressure 160/87 mmHg and normal oxygen saturation. ECG and ultrasound abdomen had no significant findings while CT CAP showed "Diffuse thickening of the wall of entire thoracic and abdominal aorta, diffuse long segment luminal narrowing of the scanned proximal left common carotid artery as well as the left subclavian artery and Diffuse thickening of the wall of the abdominal aorta is noted up to its bifurcation".

On the basis of the CT-CAP diagnosis was made. The patient was initially managed with antibiotics later after the diagnosis was made patient was given oral steroids on tapered basis. Patient was also put on PPIs, CA^{+2} supplement and vitamin-D supplements.

[1]Koide, 'Takayasu Arteritis in Japan.'

[2]Manfrini and Bugiardini, 'Takayasu's Arteritis: A Case Report and a Brief Review of the Literature.' 2006.

3. Laboratory Test and Imaging

Laboratory tests frequently lack specificity. In the early stages of the disease, the erythrocyte sedimentation rate may be high—generally greater than 50 mm/h—but surprisingly, it is frequently normal afterwards. A normal or slightly higher leukocyte count is possible. Patients with advanced illness may have a moderate, normochromic or hypochromic anemia, thrombocytosis with neutrophilia showing left s. Antinuclear antibodies, rheumatoid factor, and antineutrophil cytoplasmic antibodies are just as common in the general population as autoantibodies seen in other connective tissue diseases. Numerous results include hypoalbuminemia and elevated levels of fibrinogen, C reactive protein (CRP), and gamma globulin. There is no clear relationship that has been verified by HLA typing. The gold standard for criteria is angiograms. The aorta, its principal branches, or big arteries in the proximal upper or lower extremities must be narrowed according to the angiographic criteria. Typically, focal or segmental changes occur. The prognostic value of angiographical categorization is limited since it does not allow comparison of patient characteristics based on the vessels involved in the procedure. Aorta thickness can be measured using ultrasound or computed tomography (CT) scanning. Despite less accurate, magnetic resonance (MR) can be utilized to noninvasively evaluate the vasculature. Gallium and whole-body positron emission tomography (PET) scanning, as well as ultrasonography, may be able to help determine how much the vessels have been affected by inflammation.

4. Discussion

Takayasu's illness is a long-term inflammatory condition that affects the large and medium-sized arteries, affecting the coronary branch, the arteries of the lung, and the principal branches of the aorta. Since Takayasu's disease first came to light in 1908, there have been somewhere in the range of 2.6 cases per million people worldwide each year, with women reportedly more susceptible than men. Individuals in their 30s experience the peak onset. Although Western research have also been published, the majority of the disease's studies has been done in Japan . Cardiac characteristics can be found in up to 40% of cases. Patients with atheromatous aortas typically have no atherosclerosis risk factors, highlighting the significance of inflammation in atherosclerosis.

Vascular abnormalities result in a number of significant complications: pulmonary hypertension, aortic or arterial aneurysm, and hypertension, which is most frequently brought on by renal artery stenosis or, less frequently, narrowing of the suprarenal aorta [13, 14]. Additionally described conditions include cardiomyopathy, myocarditis, and pericarditis. Pneumonia, interstitial pulmonary fibrosis, and alveolar destruction can all occur in patients with pulmonary arteritis. Vertebrobasilar ischemia, carotid stenosis, and hypertensive encephalopathy are further

clinical signs [5, 6]. Inflammatory bowel disease, glomerulonephritis, systemic lupus, rheumatoid arthritis, and ankylosing spondylitis have also been linked to Takayasu's disease. Sensorineural hearing loss has been linked to fewer common connections. Only around 25% of patients get the Takayasu's retinopathy, which is typically accompanied by carotid artery involvement [15, 16].

5. Conclusion

Takayasu's arteritis is a rare form of large vessels' vasculitis in which there tends to be a delay in diagnosis [3]. The prognosis may be neurological impairment or asymptomatic with an impalpable pulse [4]. The onset of disease ranges from months to years. Asians are prone to have Takayasu's Arteritis, with a higher frequency in females in their early juvenile years, mostly in 2nd or 3rd decades of their life [5, 6]. In another study the onset of disease is stated to be less than 20 years age. The earlier acute phase is inflammatory with systemic and cardiovascular symptoms. compared to other ethnicities. r as reported by Manfirini et al., there is a lower prevalence of 9–10% involvement of the coronary arteries. Based on autopsy reports, angina pectoris is only evident in 6–16% of scenarios. [8, 9]. Zheng deyu et. al in a retrospective study of 530 cases out of which 346 patients were diagnosed through aortography the ratio of female to male was 2.9:1. Diagnostic criteria proposed and applied by Ishikawa K. et. al included an essential standard (age less than or equal to 40 years), two significant requirements (left and right mid subclavian artery lesions) and nine minor criteria (high erythrocyte sedimentation rate, common carotid artery tenderness, hypertension, aortic regurgitation or annul aortic ectasia and lesions of the pulmonary artery, left mid common carotid artery, distal brachiocephalic trunk, thoracic aorta and abdominal aorta). In addition to the mandated state, the presence of two major criteria, one major plus two or more minor criteria, or four or more minor criteria indicates a significant likelihood of Takayasu's illness. The utilization of radiographic and angiographic examinations reinforces this diagnostic criterion. The topology of the incidence suggests that the injury is limited to the supraoptic trunk, an alternation of the intermediate thoraco-abdominal aorta, or a combination of the two [12]. The involvement of aorta is of four types viz. arch type, extensive type, descending thoracic and abdominal type according to Kiku et. al. findings of clinical report of 84 patients reported that arch type, extensive type and descending thoracic and abdominal type differs by ischemic signs in terms of type of lesions and minor differences between males and females. [11]. Furthermore, findings from laboratory examination in terms of enhanced ESR, Increased WBC number, Anemia, increased gamma globulin and ASLO With positive CRP, RA, Wassermann and tuberculin reaction. Increased cardiothoracic ration as well as aortic calcification and abnormal EKG are some of the related diagnostic tools . In order to detect early warning manifestations like impalpable pulses and disparity in blood pressure, patients should have appropriate examinations and have even nonspecific symptoms like malaise, headache, and exhaustion thoroughly evaluated. Early illness detection can reduce morbidity and mortality since patients who arrive with symptoms including acute MI, pulmonary edema, and visual impairment later on are less likely to survive. Cerebral hemorrhage is the primary cause of death in Takayasu arteritis. Patients should be treated surgically if possible [17]. In recent studies parenchyma of various organs also seems to be involved with various dermatological manifestations like erythema nodosum, facial lupus rash and erythema induratum [10]. The involvement of the lungs in Takayasu arteritis is uncommon. One case report describes fulminant lung failure due by acute interstitial pneumonia [7]. Furthermore, in another case report Takayasu arteritis is also reported with ulcerative colitis.

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Ethical Approval

Written informed consent for publication was obtained from the patient.

Author Contributions

SA contributed to the initial management of patient, conception of case report, data collection and assisted in echocardiography, literature review and writing of first draft. MSAK contributed to literature review and critical revision of manuscript. AK contributed to literature review and review of manuscript. SH contributed to literature review and revision of manuscript. HHK contributed to management of the patient, did the echocardiography and contributed to data collection, critical revision of the manuscript. All authors read and approved the final manuscript.

Conflicts of Interest

None declared.

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