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Emerging Hypertension in Young Adults: A Detailed Case Report on Takayasu Arteritis in a 29-Year-Old Female from South India

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Abstract

Introduction: *Takayasu arteritis is a rare vasculitic disorder predominantly affecting young women, characterized by inflammation of the aorta and its major branches usually seen in the East Asian population. Due to its varied clinical presentation, early diagnosis remains challenging, necessitating a high index of suspicion.*

Case Report: *A 29-year-old South Indian female presented with exertional dyspnea to the emergency department and was initially diagnosed and treated as middle lobe pneumonia. Further physical and laboratory evaluations done at that time were unremarkable. Following successful treatment, she remained largely asymptomatic and sought evaluation at the local clinic due to elevated blood pressure detected during home monitoring. On work up of hypertension Echocardiography was done which showed unusual wall thickening of the ascending aorta and mild aortic regurgitation .CT aortogram was then done which revealed aortic thickening and left renal artery stenosis. Confirmation of takayasu arteritis was achieved through PET-CT imaging at a higher tertiary care centre, leading to initiation of immunosuppressive therapy and subsequent percutaneous transluminal angioplasty stenting of the left renal artery.*

Conclusions: *A high index of suspicion is crucial in young females with unusual presentations especially in the South Indian population where the disease is not prevalent. Due to financial constraints it is very likely that appropriate imaging is not done in cases where takayasu arteritis is not suspected by the treating physician. Lack of appropriate work up of hypertension can result in missed diagnoses which can lead to potentially disastrous side effects to the disease*

Keywords: *Takayasu arteritis, young female, vasculitis, immunosuppressive therapy, angioplasty.*

Introduction

Takayasu arteritis (TAK) is a rare vasculitis primarily affecting young women, characterized by inflammation of the aorta and its major branches. This condition often manifests as nonspecific symptoms in its early stages.[1] Early recognition of TAK is crucial, as delayed diagnosis can lead to complications and suboptimal outcomes.[2] The diagnosis of takayasu arteritis is usually done based on a high degree of clinical suspicion . Specific blood tests for the same are unavailable. Imaging remains the diagnostic modality and is also used to assess the extent and severity of the disease. Despite its rarity, understanding the range of presentations of TAK is essential for timely intervention and effective management.[3,4] Delaying the diagnosis of a chronic inflammatory disease exacerbates the extent of inflammatory damage inflicted. Our case presents a young Indian female with atypical symptoms whose initial diagnosis of takayasu arteritis was missed and was found out on timely work up of her hypertension.

Case Report

A 29-year-old Indian female, presented to the emergency department with exertional dyspnea (NYHA Class 2) and a vague symptom of bilateral lower limb claudication which had worsened over the past month. There were no complaints of effort-related angina, abdominal angina, jaw claudication, carotidynia, giddiness or syncope. A detailed physical exam was done and was found to be unremarkable .Past medical history was unremarkable for cardiovascular events. On her background of dyspnea, Computed tomography pulmonary angiography (CTPA) was performed at the emergency department rule out a pulmonary embolism, revealing ground glass opacities in the right middle lobe, leading to a diagnosis of middle lobe pneumonia for which she was treated accordingly.

The patient was then predominantly asymptomatic and presented for routine care to her primary care physician following repeated high blood pressure values noted at home measurement. Routine physical examinations and lab values including an ESR were unremarkable. Echocardiography was done to follow up her hypertension and showed aortic valve trileaflet and mild to moderate aortic regurgitation with normal diastolic function and right ventricular function, with no evidence of clot, vegetations, or pericardial effusion. A detailed assessment of aortic dimensions showed normal values, except for unusual wall thickening along the ascending aorta and proximal arch of the aorta, suggestive of large vessel vasculitis. She was then referred for a CT aortogram which showed aortic wall thickening and 90 percent osteoproximal left renal artery stenosis.

The patient was referred to a higher center for further evaluation where further characterization through a whole-body PET CT scan unveiled metabolically active lesions consistent with vasculitis along the ascending aorta and proximal arch, as well as near occlusion of the left renal artery origin. Bilateral subcentimetric level II cervical and subcarinal nodes were observed, likely inflammatory in nature. Additionally, metabolically inactive subcentimetric axillary nodes and a left adnexal cyst were noted on CT imaging. It confirmed large vessel vasculitis, consistent with Takayasu arteritis type 4. She was commenced on immunosuppressive drugs for disease management and scheduled for left renal artery stenting.

Following three months of immunosuppressive therapy, a procedural intervention was performed, involving bilateral transfemoral angiography, pulmonary angiography and stenting of the left upper and lower renal artery under under general anesthesia. This addressed a significant 90% stenosis in the left renal artery. The procedure involved meticulous stenting and simultaneous kissing balloon inflation to ensure adequate arterial patency and flow restoration. This procedure was done within 7 months of the initial diagnosis.

Post-procedure, the patient was stabilized and transferred to the ward for further management. Plans were made to initiate dual antiplatelet therapy and schedule mandatory follow-up angiography at 6-9 months to monitor the response to treatment and disease progression. The patient had an uneventful post-procedure course and was discharged with medication recommendations and follow-up plans. The patient is currently stable and on antihypertensive and immunosuppressive drugs and is under continual follow-up.

Discussion

Takayasu arteritis (TAK) is notorious for its heterogeneity in clinical presentation, often mimicking other conditions, thereby posing diagnostic challenges.(5) In this case, the patient initially presented with exertional dyspnea and vague \lower limb claudication, symptoms not pathognomonic for TAK. At her initial presentation a diagnosis of takayasu arteritis was missed. Furthermore, the absence of classical features such as abdominal angina or carotidynia further delayed the diagnosis. Hence, a high index of suspicion needs to be maintained for TAK, particularly in young individuals with nonspecific symptoms suggestive of systemic inflammation.

Khadka et al. described a case of Takayasu's arteritis in a 26-year-old woman presenting with syncope and dizziness. Magnetic Resonance Imaging angiography showed thickening of the aortic arch and its branches. Both cases involve young female patients with symptoms suggestive of Takayasu arteritis, showing commonalities such as elevated inflammatory markers and arterial wall abnormalities. However, our case presented with exertional dyspnea, claudication, and hypertension, whereas Khadka et al. 's patient experienced syncopal attacks, unequal blood pressure, and myalgia. These differences may arise from varying disease progression, individual patient characteristics, and arterial involvement.[6]

A case reported in a Japanese patient mirrors our own, both involving young females with symptoms suggesting Takayasu arteritis (TA). Despite demographic and clinical differences, both cases share common features such as elevated blood pressure, diminished pulses, and arterial stenosis, notably in the renal arteries. However, management approaches diverged; our patient underwent PTA stenting and immunosuppressive therapy, while the Japanese patient received oral medications. These differences may arise from varying disease severity, treatment response, or institutional practices, alongside healthcare setting disparities and individual patient factors.[7]

Manfrini et al has reported a case of TAK in a Caucasian woman. Similar to ours, it depicts large vessel vasculitis but exhibits differing clinical presentations. Our shows Takayasu arteritis in a younger patient with renal artery involvement and pulmonary manifestations, while the Caucasian lady was an older patient which had giant cell arteritis with severe aortic involvement and coronary artery disease. Plausible reasons for differences include varied underlying pathophysiology, genetic factors, and timing of symptom onset.[1] Cases of TAK associated with Crohn's disease have also been reported.[8–10]

In a comparative study conducted Jain et al ,106 patients were documented over a period of 16 years. These patients presented with hypertension (51.3%) as the most common presenting symptom found in 82 percent of the patients (3). The diagnostic journey for TAK often involves a combination of imaging modalities and laboratory tests. In this case, initial investigations such as CTPA came back inconclusive to the disease. Echocardiography provided valuable insights, albeit not definitive for TAK. However, it was the PET-CT imaging that confirmed the diagnosis, demonstrating metabolically active findings consistent with large vessel vasculitis. This highlights the role of PET-CT as a powerful tool

in diagnosing TAK, particularly when other modalities yield inconclusive results.

The cornerstone of TAK management revolves around immunosuppressive therapy to control disease activity and prevent vascular damage. In this case, the patient was promptly initiated on immunosuppressive drugs following confirmation of TAK diagnosis. Additionally, the patient underwent percutaneous transluminal angioplasty (PTA) stenting of the left renal artery, emphasizing the importance of a multidisciplinary approach involving rheumatologists, cardiologists, and surgeons in managing TAK effectively.

While TAK can be a challenging condition to manage, early diagnosis and appropriate treatment initiation significantly impact prognosis. Long-term management typically involves close monitoring for disease activity, optimization of immunosuppressive therapy, and surveillance for complications such as arterial stenosis or aneurysm formation. Moreover, patient education and adherence to treatment regimens are paramount in achieving favourable outcomes.

Conclusions

Recognition of atypical presentations of Takayasu arteritis is crucial for timely management and optimal outcomes. Multidisciplinary collaboration and targeted investigations are essential in cases with subtle symptoms. This case shows the importance of considering TAK in the differential diagnosis of young adults presenting with nonspecific symptoms suggestive of systemic inflammation. In developing countries there is a high chance that such diagnoses can be missed due to lack of proper timely imaging due to financial constraints of the patient Early recognition, comprehensive evaluation, and a multidisciplinary approach are essential for accurate diagnosis and effective management of TAK, thereby mitigating the risk of complications and optimizing patient outcomes. Further clinical awareness and work up in all Indian patients presenting with are warranted to enhance our understanding and management of this rare vasculitic disorder.

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