Case Report

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A rare case of Takayasu arteritis with aortic dissection in a young male patient presented with hypertensive urgency

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Introduction and importance: Takayasu arteritis (TA) is a granulomatous, large vessel inflammation characterized by occlusion, stenosis or aneurysm of the aorta and its branches. Aortic dissection (AD) is one of the rare manifestations of TA, which can present as uncontrolled hypertension. This case illustrates the severe rare cardiovascular complication that arises in a 21-year male patient with TA, including hypertensive urgency, AD and significant renal artery involvement.

Case presentation: A 21-year-old male presented with complaints of fever, malaise and dizziness. On clinical examinations, a discrepancy in blood pressure was noted in bilateral arms. All the laboratory parameters were within normal limits. Echocardiography confirmed moderate concentric hypertrophy with Grade II diastolic dysfunction and a preserved ejection fraction. To rule out secondary causes of hypertension, an arterial Doppler was done, which showed features of renal artery stenosis and stenosis in the descending aorta. CTA revealed an intimal tear/dissection in the thoracoabdominal aorta consistent with Stanford Type B AD.

Clinical discussion: Based on clinical presentation and imaging findings, large vessel vasculitis such as TA or giant cell arteritis (GCA) was suspected. Later, TA was diagnosed using the 2022 American College of Rheumatology/EULAR classification criteria for TA. The patient had rapid symptomatic improvement using antihypertensive drugs, corticosteroids and immunosuppressants.

Conclusion: This explains rare manifestation of TA in a young male patient with normal inflammatory markers and underlying AD with hypertensive urgency. In those suspected with TA, screening for life threatening conditions such as AD should be done.

Keywords: aortic dissection, case report, hypertensive urgency, Takayasu arteritis

Introduction

Takayasu arteritis (TA) is a chronic, granulomatous, large vessel inflammation characterized by occlusion, stenosis or sometimes aneurysm of the aorta and its branches. Other large vessels, including pulmonary and medium-sized coronary arteries, might also be involved[1]. TA is an overwhelming illness of young adults, predominantly females, in the second and third decades of life. Despite being worldwide, the disease is more prevalent in regions such as India and East Asia[2]. The development of vascular inflammation is thought to be a significant factor in the dysregulation of T cells, particularly CD4+ and CD8+ T cells. Renal involvement is related to an imbalance of pro-inflammatory and anti-inflammatory cytokines, such as interferon-gamma and IL-6[3], which could be the reason for a secondary cause of hypertension/hypertensive urgency in

HIGHLIGHTS

- Takayasu arteritis (TA) is an idiopathic granulomatous inflammatory and stenotic disease of medium and large sized arteries characterized by strong predilection for the aorta and its branches. Aortic dissection (AD) is a rare manifestation of TA.
- This case highlights the importance of keeping TA as one of the differential diagnoses even in young male patient with unexplained hypertension.
- TA patients with poorly controlled or long-standing hypertension are likely to develop AD where initial clinical manifestations could be hypertensive urgency.
- Elevated levels of inflammatory markers such as Erythrocyte sedimentation rate (ESR), C-reactive protein (CRP) and cytokines are associated with active phase of TA while their normal levels indicate inactive phase and does not rule out TA.
- Treatment of Takayasu arteritis includes endovascular methods, surgery and drugs such as corticosteroids, immunosuppressives, antihypertensives, vasodilators and anti-inflammatory drugs which can assist with decreasing vascular inflammation and prevention of complications.

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young patients. A systolic blood pressure of 180 mm Hg or a diastolic blood pressure of 110 mm Hg without accompanying end-organ damage is considered hypertensive urgency. In addition to being entirely asymptomatic, hypertensive urgency can also be accompanied by symptoms including headache, dyspnea, and anxiety and are associated with a higher incidence of cardiovascular events[4]. Uncontrolled hypertension is one of the risk factors of aortic dissection (AD), a life-threatening condition caused by a tear in the intimal layer of the aorta or bleeding within the aortic wall resulting in separation of the layers[5]. AD is a rare complication of large vessel disease such as TA[6].

This is an atypical case of a young male patient who presented with fever and hypertensive urgency. Later, it was proved to be a complex diagnosis, such as TA. This case report has been reported in line with SCARE Criteria[7].

Case presentation

A 21-year-old male with no known co-morbidity presented primarily in OPD of a tertiary care center in Kathmandu, Nepal, with a history of fever associated with headache and malaise for 10 days. He also complained of increased dizziness 5 days prior to the presentation. There was no blurry vision, chest pain or dyspnea. He had no history of drug abuse, but he smoked 2 pack years. Upon physical examination, no deformities were noted. On systemic evaluation, he had normal vesicular breath sounds. In the cardiovascular examination, no murmurs were heard. His abdomen was soft and non-tender, and no organomegaly was present. There were no signs of cyanosis and peripheral edema. The vital signs were as follows: His systolic and diastolic readings taken at multiple intervals of time at a supine position had an average blood pressure of 200/100 mm Hg in the right arm, 180/90 mm Hg in the left arm, femoral blood pressures were 210/100 mm Hg bilaterally. There was no radio-radial and radio-femoral delay, respiratory rate: 22 breaths/min, pulse rate: 104 beats/min, and Temperature: 39.4°C. Also, bruits were heard over the left subclavian artery, aorta and bilateral renal arteries.

Investigation and management

Based on the above findings, baseline lab investigations showed hemoglobin of 9.52 gm/dL, total lymphocyte count of 8590/μL, platelets of 405 300/μL, and creatinine of 0.7 mg/dL. Liver function tests, urinalysis and thyroid function tests were within normal range. His hsCRP was <0.34 mg/L and ESR was 10 mm/hr. Antinuclear antibody, rheumatoid factor, anticardiolipin antibodies and venereal disease research laboratory test (VDRL) were negative. To rule out causes of fever as a tropical illness, Salmonella, Dengue, Scrub Typhus, and Leptospira Antibody tests were done, and the results were negative. Serology for Rickettsia, agents of viral hepatitis, Epstein-Barr virus, Cytomegalovirus, and HIV were negative. The malaria smear was negative. Ultrasound of the Abdomen and Pelvis was done, which showed a normal scan.

The electrocardiogram suggested left ventricular hypertrophy (Fig. 1). The echocardiography report suggested moderate concentric left ventricular hypertrophy, Grade II left ventricular diastolic dysfunction with a left ventricular ejection fraction of 60%. Other heart chambers were normal. To rule out the secondary cause of hypertension in young patients, arterial Doppler was performed, which showed tardus parvus waveform in bilateral renal arteries and atheromatous wall calcification in the aorta. Features were favorable for stenosis in the descending aorta; hence, a CT aortogram was suggested.

CT aortogram showed calcified and non-calcified atherosclerotic plaque noted in the distal arch of the aorta with a narrowed caliber of 1.1 cm in diameter at the D10 vertebral level. There was a hypodense intimal flap in the descending thoracoabdominal aorta distal to narrow segment of descending thoracoabdominal aorta extending from D11-D12 to L2 vertebral body (Stanford B AD) without differential luminal attenuation, mural hematoma and contrast extravasation. Occlusion of the proximal renal artery followed by collateral reformation was noted distally, and the visualized part of the proximal left superficial femoral artery was narrow in caliber (Fig. 2). AD Stanford B was diagnosed based on clinical and CT aortogram findings. Although AD is a rare presentation of large vessel vasculitis, the clinical presentation of our patient includes age, fever of unknown origin, discrepancies in blood pressure in upper and lower limbs, multiple occlusions and narrowing of large and medium-sized blood vessels, suggesting suspicion towards large vessel vasculitis such as TA or giant cell arteritis (GCA). According to the criteria for TA defined by the American College of Rheumatology/EULAR, our patient had met the absolute requirement of age ≤60 years, evidence of vasculitis on imaging, including other additional criteria such as vascular bruit (2 points), systolic blood pressure difference in arms ≥20 mm Hg (1 point), three or more affected arterial territories (3 points), abdominal aorta involvement with renal artery involvement (3 points), making it a total of 9 points out of 22 points in which a score of ≥5 is needed for the diagnosis of TA.

Treatment and follow-up

Since the patient presented with fever and hypertensive urgency, he was admitted and managed with antipyretics (Paracetamol), a combination dose of calcium channel blocker and angiotensin receptor blocker (Amlodipine 5 mg and Losartan 50 mg) and statin (Atorvastatin 20 mg). Blood pressure was still not controlled; hence, a Beta-blocker (Metoprolol 25 mg) was added. Half-hourly BP monitoring was done, which showed normalization of blood pressure after a few hours. In line to treat TA, Prednisolone 50 mg also started, which was tapered later. The patient's condition remained stable throughout the hospital stay. Blood Pressure was maintained at 130/80 mm Hg in the right arm and 120/90 in the left arm, with no other subjective complaints. After 6 days, the patient was discharged with antihypertensive medications (Metoprolol, Amlodipine and Losartan) and Prednisolone. During follow-up after 2 weeks, Prednisolone was tapered to 40 mg for the next 2 weeks, 30 mg for 2 weeks, 20 mg for 2 weeks, 10 mg for 2 weeks and 5 mg to continue. Tab Methotrexate 15 mg weekly was also started along with Folic acid 5 mg twice weekly to enhance therapeutic efficacy. After three months of follow-up, he exhibited notable clinical improvements, including resolving symptoms such as fever, dizziness, headache, and normal blood pressure. Further investigations showed normal lab parameters in follow-up, including baseline investigations, ESR and CRP.

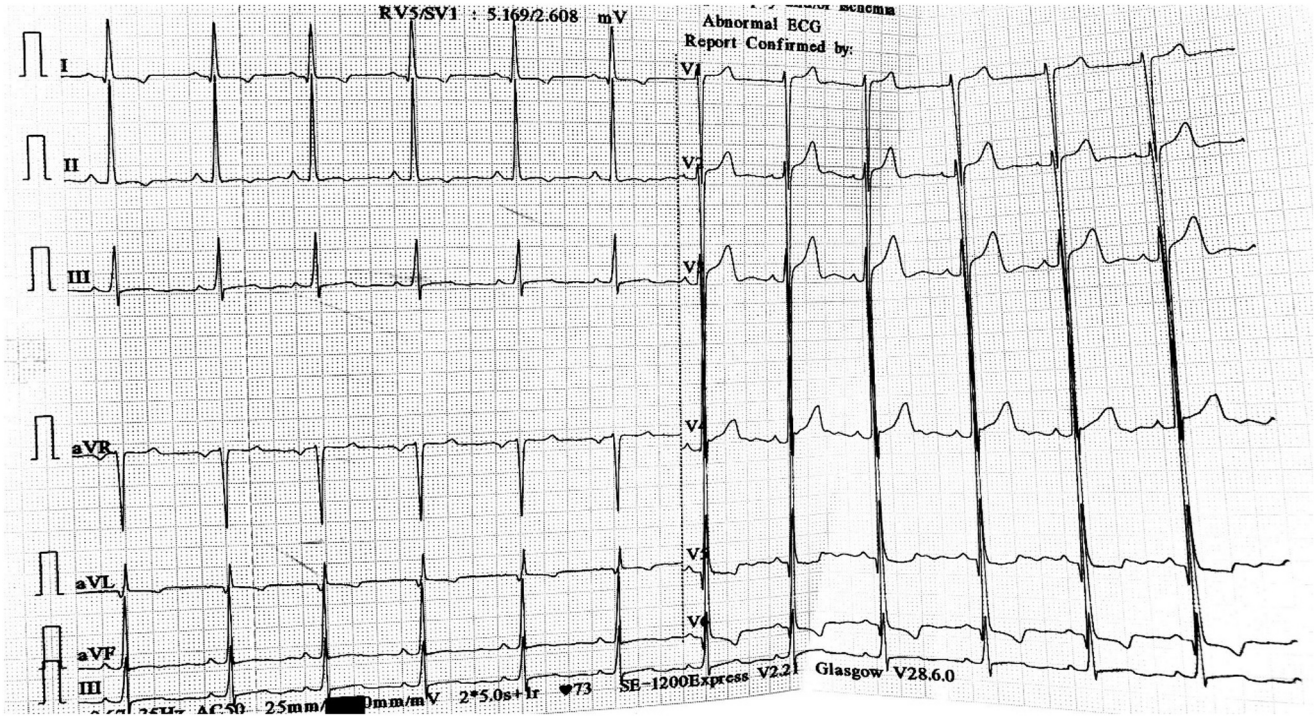

Figure 1. ECG showing features of Left Ventricular Hypertrophy.

Discussion

TA is a rare form of vasculitis, essentially influencing large blood vessels like the aorta and its fundamental branches. Classical presentation includes limb claudication, reduced or absent pulses, and vascular bruits. TA can likewise manifest unusually, adding complexity to its diagnosis and management[8]. Similarly, etiology and pathogenesis remain controversial. One of the hypotheses states the cause to be the activation of CD4+ T cells after deposition in the vascular wall, which releases cytokines attracting monocytes that are later transformed into macrophages, resulting in endothelial damage and granuloma formation. Another hypothesis suggests the involvement of humoral immunity in the pathogenesis of TA. Anti-endothelial cell antibodies and anti-monocyte antibodies are also present in patients with TA. However, further extensive studies are required to elucidate the pathogenesis of TA[2].

In TA patients, gender strongly influences the pattern of vascular involvement and clinical presentation. In one such study, it was found that females below 40 are commonly affected as compared to males. In female TA patients, the artery most frequently involved was the left subclavian artery, and in male TA patients, the artery most frequently affected was the abdominal aorta and renal arteries[9]. While stenosis is the most common pattern of arterial lesion and AD, have been reported in only 25% and is considered rare. Hypertension is considered the most prevalent risk factor for AD. Any part of the aorta can develop dissections in patients with TA, but the descending aorta tends to be involved more often, which can be diagnosed with CTA for early detection of AD in TA patients[6]. Our case also represents a rare case of TA in a young male with AD.

The disease has two distinct phases in its clinical course: an early aggressive inflammatory phase and a late chronic phase/ inactive phase. The active phase may be remitting and relapsing,

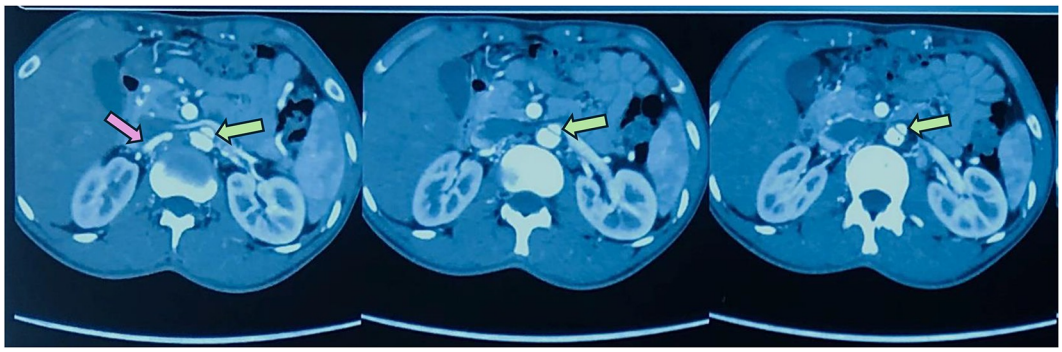

Figure 2. Features of intimal tear/ dissection in thoracoabdominal aorta from D11-D12 to lower level of L2 vertebral body (Stanford B) (Green Arrow), Occlusion of proximal right renal artery (Red Arrow).

lasting for weeks to months. Headaches, dizziness, arthralgia, fever, night sweats, appetite loss, weight loss, and other symptoms indicate a systemic disease. However, some patients do not experience the acute phase but have constitutional symptoms. Thus, normal ESR and CRP levels might indicate inactive disease[10].

Another unusual manifestation of TA is the difference between the BP of the upper and lower limbs. Although the exact pathogenesis is multifactorial, some of the causes of the difference in BP could be due to a decrease in the elasticity of the arterial wall or renal hypoperfusion due to stenotic lesions of one or both renal arteries or aorta alone[2]. Renal artery stenosis is one of the most common secondary causes of hypertension, which activates the renin-angiotensin-aldosterone (RAS) system, leading to renovascular hypertension[11]. Similarly, the presence of systolic blood pressure in the lower limb is generally greater than in the upper limb due to anatomical variations. Some literature suggests that clinicians should expect blood pressure difference in the lower to upper limbs by at least 15 mm Hg in the supine position[12]. On the other hand, discrepancies in blood pressure in bilateral arms are common presentations in TA, likely due to subclavian artery stenosis resulting in the narrowing of blood vessels and decreasing blood flow, causing a reduction in blood pressure. In our case, bruit was heard over the left subclavian artery area, and the blood pressure in the left arm was lower than in the right[13].

In clinical practice, the diagnosis of TA was done using the American College of Rheumatology/EULAR criteria, which include age ≤60 years, evidence of vasculitis on imaging as an absolute criteria, while other additional criteria are systolic blood pressure difference between two arms ≥20 mm Hg, female sex, angina, arm or leg claudication, vascular bruit, reduced pulse in upper extremities and others with maximum score 22 points in which our case satisfied criteria for classification of TA which is ≥5[14].

Other differential diagnoses, such as GCA, can be ruled out as per the 2022 ACR/EULAR classification for GCA absolute criteria, which states that age must be ≥50 years at the time of diagnosis. In our case, other features such as morning stiffness, visual loss, jaw or tongue claudication, and scalp tenderness are also absent[15].

In the 2021 large-vessel vasculitis guidelines, the American College of Rheumatology and Vasculitis Foundation recommend noninvasive imaging to include computed tomography angiography (CTA), positron emission tomography (PET) or magnetic resonance angiography (MRA) for determining potential vessel wall inflammation, without pointing out the preferred imaging modality[14]. In our case, computed tomography angiography was done and showed features of intimal tear/ dissection in thoracoabdominal aorta from D11—D12 to lower level of L2 vertebral body (Stanford B) without differential luminal attenuation, mural hematoma or contrast extravasation. CTA is a comprehensive technique for vascular evaluation in TA patients. It allows evaluation of both vessel lumen and vessel wall changes, of which the later may precede vascular changes[16]. Doppler USG is used to evaluate stenosis in the main renal artery and by analyzing the Doppler waveform obtained from the intrarenal artery[17]. In our patient, a Doppler ultrasound of the renal artery was performed, which showed a tardus parvus waveform in bilateral renal arteries—atheromatous wall calcification in the aorta with tardus parvus waveform. Features were in favor of stenosis in the descending aorta.

The relatively low incidence, atypical symptoms, and absence of risk factors may lead to misdiagnosis in the TA population. To improve the early diagnosis rate of TA, various diagnostic procedures should be evaluated together, including meticulous physical examination, especially the blood pressure of both upper limbs, the vascular murmur of the neck, upper clavicle, or abdomen, laboratory tests such as ESR and CRP, and angiographic findings[18].

The treatment of TA includes drugs, endovascular methods, and surgery. During the acute inflammatory stage, medical treatment normally incorporates corticosteroids, immunosuppressive medications, antihypertensives, anticoagulants, antiplatelets, vasodilators, and anti-inflammatory drugs, which can assist with decreasing vascular inflammation and advance the relapse of vessel injuries[19]. The patient responded well to an extensive treatment routine, including antihypertensives, corticosteroids, and enhancements, prompting a critical improvement in side effects. Beta-blockers also play a crucial role in reducing aortic wall stress in AD and help reduce high blood pressure[20]. Blood pressure was successfully normalized, and the patient was stable throughout follow-ups. Since this was an uncomplicated Stanford B AD case, the patient was managed medically[21]. However, proper counseling was done to address the probable need for surgical intervention if medical management is insufficient. The patient is under close monitoring with follow-ups from cardiology, rheumatology, radiologist, cardiac and vascular surgeons.

Conclusion

Hypertensive urgency, AD and notable involvement of the renal arteries are some of the alarming cardiovascular impacts of TA, which is described in this case. TA patients with unexplained hypertension should be screened for AD using CTA or other imaging modalities. Considering different sites of involvement of AD, tailored therapy should be initiated by a team comprising cardiologists, cardiac and vascular surgeons and interventional radiologists. The case also underlines a high index of suspicion of TA in a young male patient with unexplained hypertension and vascular diseases, even with normal inflammatory markers such as ESR and CRP, as early diagnosis can determine the course of management and decrease the rate of life-threatening complications such as AD. Therefore, vigilant future follow-ups are also crucial to determine AD development in patients with TA.

Ethical approval

Not applicable.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author's contribution

K.R.P.: manuscript preparation, obtaining consent from patient, edit and Review; M.G., P.B.: manuscript preparation, editing; B.S.: manuscript review, patient care, editing; S.T.: patient care during follow up, manuscript review, monitoring; S.S.: manuscript review.

Conflicts of interest disclosures

All the authors declare to have no conflicts of interest relevant to this study.

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Not applicable.

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