Case Report

# Aortoarteritis: A rare pulmonary-renal disease

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## ABSTRACT

Takayasu arteritis (TAK) is an autoimmune disease majorly affecting young females. It alters the vascular wall, resulting in stenosis, occlusion, or dilatation. It has no distinct clinical manifestation. Here, we present the case of an 18-year-old girl who presented with generalized tonic-clonic seizure and hypoxia. Blood investigations showed deranged urea and creatinine values. Computed tomography angiography revealed bilateral artery occlusion, decreased kidney size, and pulmonary artery dilatation, confirming generalized vascular disease that caused hypertension and ischemic nephropathy in the patient. Our case represents a rare autoimmune disease, leading to pulmonary hypertension and renal artery stenosis. TAK should be considered as a differential diagnosis in young female patients presenting with pulmonary as well as renal signs and symptoms, especially if there is discrepancy in blood pressure levels in all limbs.

**Key words:** Bilateral renal arteries, Cortical stenosis, Granulomatous diseases, Ischemic nephropathy, Pulmonary hypertension, Vasculitis

Takayasu arteritis (TAK) is a granulomatous inflammatory disease of medium to large vessels, resulting in stenosis, occlusion, and dilatation of involved arteries [1]. The clinical manifestations of the disease can range from non-specific constitutional symptoms such as fever, malaise, anorexia, and dyspnea to more distinctive features caused by stenosis/occlusion of the vascular territories involved [2]. The disease is frequently misdiagnosed due to its non-specific symptoms and may have a relapsing course [3]. TAK, also known as aortoarteritis, affects 2.6 people per million each year, with Japan having the highest prevalence [4]. According to studies, the involvement of the descending aorta is less common in patients with pulmonary artery involvement (PAI) than in patients without PAI [5].

We present a case of aortoarteritis with involvement of the pulmonary artery, descending aorta, and bilateral renal arteries.

## CASE REPORT

An 18-year-old girl presented to our emergency room with an episode of generalized tonic-clonic seizure 8 h back and hypoxia. On inquiry, this was preceded by dyspnea, headache, and decreased urine output for the past 3 days. She had no previous history of hypertension, chest pain, and dyspnea and was very active in her everyday life.

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The physical examination revealed accelerated hypertension significant inter-arm and inter-limb differences in blood pressure (BP) and bruit over the right subclavian artery. BP in the right upper limb was 180/120, 150/70 in the left upper limb and 200/140 in both lower limbs. Her pulse rate was 102 beats per minute and respiratory rate was 32 per min.

On admission, she had deranged creatinine concentration of 4.4 mg/dl and urea of 96 mg/dl. Erythrocyte sedimentation rate (ESR) was 52 mm/h, hemoglobin was 11 g/dl, and other blood investigations were normal. Urinalysis revealed trace proteinuria, suggestive of tubulointerstitial, or vascular disease. Computed tomography (CT) aortogram revealed 4 cm long thickening of the aorta (Fig. 1a) at the level of the origin of renal arteries, causing occlusion of bilateral renal arteries 3 mm from the origin with cortical stenosis (Fig. 1b). The right kidney measured 6.4 × 3.9 cm, and the left measured 5.7 × 3.2, both small in size. The pulmonary arteries were dilated, suggestive of pulmonary hypertension. These findings confirmed our initial suspicion of generalized vascular disease causing hypertension and ischemic nephropathy.

The patient was administered calcium channel blockers, alpha-adrenergic blocking agents, and beta-adrenergic blocking agents. BP was partially controlled. Angiotensin receptor blockers and angiotensin-converting enzyme inhibitors were avoided in view of bilateral renal arteries stenosis. Blood investigations revealed improvement in creatinine level from 4.4 mg/dl to 2.8 mg/dl. ESR improved from 52 to 35. Interventional radiologist opined that balloon angioplasty or stenting was not feasible as there was a severe narrowing of the ostia.

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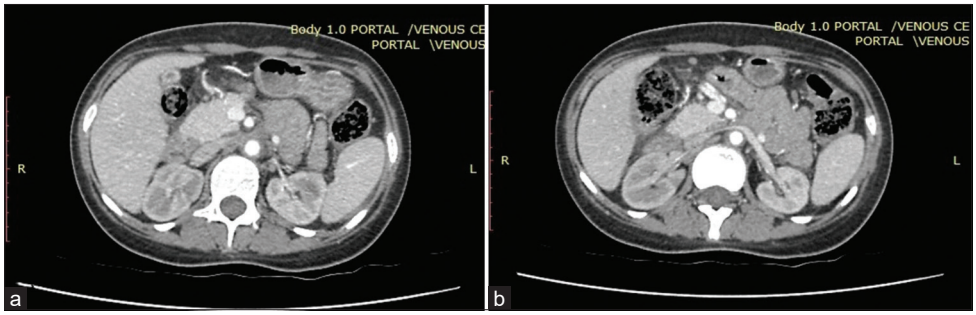


**Figure 1: (a) Computed tomography (CT) angiogram showing inflammation at the descending aorta; (b) CT angiogram showing stenosis of the right renal artery along with inflammation at the descending aorta**

She was discharged and has been on a regular follow-up to date. Her creatinine level is still 1.8 mg/dl. There were no further episodes of hypertensive emergencies thereafter.

## DISCUSSION

Aortoarteritis, also known as Takayasu's disease, has a wide spectrum of symptoms, with the majority of patients remaining asymptomatic or minimally symptomatic for varying periods of time over the course of the disease [6]. As a result, aortoarteritis is frequently misdiagnosed until it has progressed to the point where serious complications are caused by the disease, resulting in a poor prognosis [6]. A study from China suggested that the mean age at onset of the disease was 28.9±12 years, but, in another study, it was 23 years [7]. In Poland, a retrospective study stated a mean age of 26 years, and another affirmed a mean age of 45.4 years [7]. Our patient was 18 years of age when she was first diagnosed with aortoarteritis.

The disease was observed more often in females than males, with males having a greater risk of complications than females [2]. The patient in the case was a female with complications which were actively managed. The condition is more common in Asian countries. The occurrence of aortoarteritis can be related to many different factors, but genetic and personal factors are considered more relevant [7]. Some genetic factors such as HLA-B*52 and FCGR2A/FCGR3A are still under study [8].

Patients with TAK who had PAI had dyspnea as one of the major symptoms, indicating that PAI should be considered when TAK patients complain of dyspnea [5]. In tropical areas, recurring fever is considered one of the major symptoms of TA when infectious cause is eliminated [9]. Blood tests, such as elevated ESR, C-reactive protein, interleukin 6, and pentraxin levels, can aid in the diagnosis [2]. Previously, traditional angiography was the gold standard for diagnosis, but due to its invasive nature, it has been replaced by duplex ultrasonography, CT scan, and magnetic resonance angiography [9]. Positron emission tomography CT is becoming increasingly popular among physicians and is an important diagnostic tool [10]. However, for diagnosis, it is preferable to use a combination of imaging modalities as well as blood analysis. The above case was diagnosed with the non-invasive imaging technique, CT angiography along with the blood investigations.

In the active stage of the disease, conservative care with corticosteroid medication is frequently recommended, as are oral anticoagulant medicines. In TAK with aortic dissection, surgical treatment in inactive disease is considered beneficial [11]. For symptomatic treatment, digitalis, antihypertensive medications, and antibiotics are routinely employed. In individuals with significant symptoms that are not resolved by conservative therapy, balloon angioplasty or stenting may be considered for surgical repair [6]. The patient was administered calcium channel blockers, alpha-adrenergic blocking agents, and beta-adrenergic blocking agents which significantly lowered the BP in the patient.

The case had presented with pulmonary and renal symptoms, for which small vessel vasculitis is generally the most common differential diagnosis; whereas the above case was of large vessel vasculitis. The case had a unique presentation of involvement of descending aorta along with pulmonary arteries which are not the common type of presentation in the patients with aortoarteritis.

## CONCLUSION

Early diagnosis and management are the key in treatment of aortoarteritis. Patients showing renal and pulmonary signs and symptoms in conjunction, large vessel vasculitis should be considered and ruled out. Such cases can be effectively managed conservatively.

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