

## Case Report

# A Rare Case of Takayasu's Arteritis with Aortic Arch Branching Pattern Variation in a Young Female

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Takayasu's arteritis, formerly known as "pulseless disease", is a chronic inflammatory disease which affects the aorta and its main branches. The prevalence of this disease is higher in Asian countries and among young women. Depending on the progress of the disease, the symptoms and prognosis of the disease is different. Herein, we report the case of a 31-year-old woman with Takayasu's arteritis in the Iranian population. In this case, the size of the aorta and the main branches increased and the large artery walls were thickened. Narrowing of the right and left subclavian artery, carotid artery and left vertebral artery were obvious in these patients. In addition, an anatomic variation was observed in the aortic arch branches. In this patient, the left vertebral artery was branched directly from the aortic arch instead of the left subclavian artery. Hepatomegaly and splenomegaly, indicating a chronic inflammatory disease, were also observed in this case.

**Keywords:** Anatomic variation, Aorta, Computed tomography angiography, Takayasu arteritis

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**Introduction**

Takayasu's arteritis is a rare chronic vasculitis. It mostly affects the aorta and its large branches. It is also called the pulseless disease and is more common in young women. The disease was first reported by Takayasu in a 21-year-old woman in Japan. The prevalence of this disease is more common in Asian countries as well as North America.<sup>2</sup> Some of the symptoms of this disease include weakness, anorexia, hypertension, joint pain, fever, night sweats, weight loss, nausea, vomiting and anemia. The disease is detectable through clinical examinations and imaging techniques. The advancement of imaging techniques has made it possible to diagnose the disease with noninvasive methods such as computed tomography (CT) scan and magnetic resonance angiography (MRA) in early stages.<sup>3</sup> Early diagnosis of this disease will have a great impact on its treatment.

At the International Conference on Takayasu's arteritis, a new classification was made based on angiographic images of the disease (Figure 1).

This type of classification is based on the region of the vessels involved in the disease. The prevalence of each of these classes varies from one community to another.

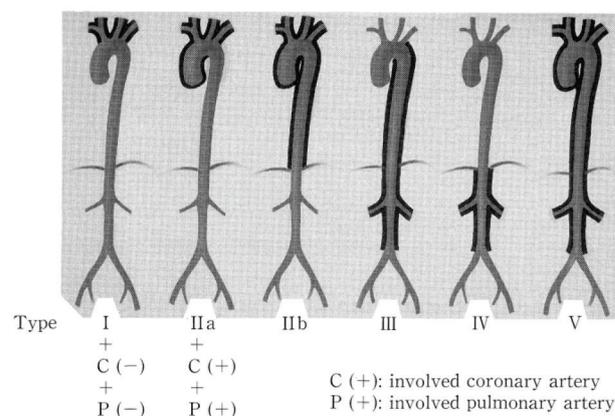
**Case Report**

A 31-year-old woman with chest pain was referred to the emergency department of the hospital. Anemia and very poor pulse were observed in both upper limbs during

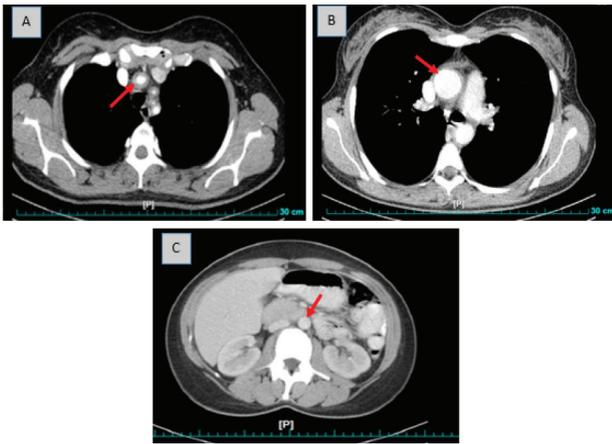
clinical examinations. She was referred to the radiology department to perform a thoraco- abdominal CT scan and CT angiography using 16-slice CT scanners (SOMATOM Sensation 16, Siemens, Germany).

Significant increases in the diameter of the ascending aorta, the aortic arch and its major branches were observed in the thoracic, abdominal and pelvic CT images (Figure 2). In addition, an increase in the thickness of the aortic wall and large arteries was also evident in these regions.

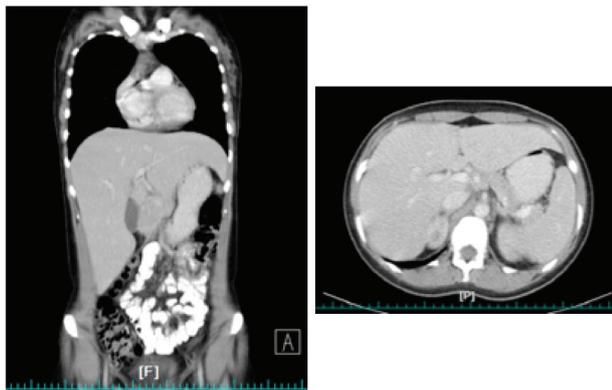
Heart size was normal in this patient. According to Figure 3, hepatomegaly and enlargement of the spleen existed in this patient. Kidneys and other organs had



**Figure 1.** Classification of Takayasu's Arteritis Types Based on Angiographic Results.<sup>4</sup>



**Figure 2.** CT angiography images of aorta and its branches, A) Significant increase in the diameter of the right Brachiocephalic trunk (BCT) along with its thickening wall; B) Ascending aorta had a significant increase in diameter and thickness c) diameter and thickness of the abdominal aorta was normal.

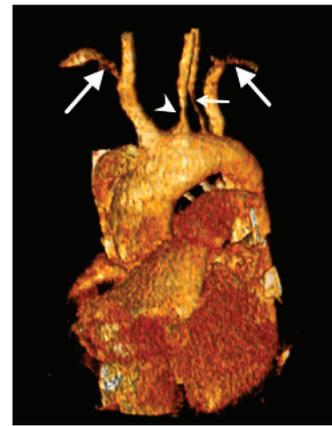


**Figure 3.** CT Scan Images Of Lung, Abdomen and Pelvis of the Patient in the Coronal (left) and Axial (right) Sections with Contrast Media.

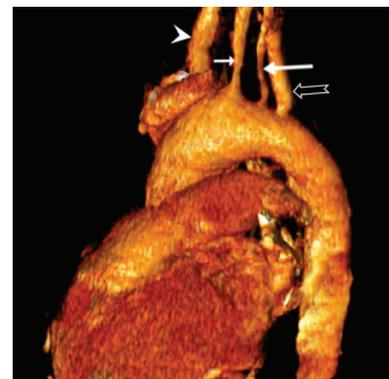
normal shape and dimensions.

In the study of angiographic images, occlusion of right subclavian artery was observed. The ascending aorta and aortic arch as well as carotid arteries, brachiocephalic trunk and subclavian arteries showed wall thickening. Proximal portion of descending aorta was also involved. Left subclavian artery stenosis (when exiting the thorax), right subclavian artery origin stenosis, left carotid and vertebral arteries proximal portion stenosis was noted (Figure 4). No obvious abdominal aorta involvement was revealed in this case.

According to the angiographic images, Takayasu’s arteritis was type IIb in this case. Additionally, variation in aortic arch branches was observed in this case. As shown in Figure 5, the left vertebral artery was separated directly from the aortic arch. In normal mode, this artery is separated from the left subclavian. Therefore, in this case, the aortic arch branches were brachiocephalic trunk, left



**Figure 4.** Three-dimensional CT Angiography of the Aortic Arch and its Branches; large arrows represent narrowing of the right and left subclavian arteries; small arrows indicate narrowing of the origin of left vertebral artery and the flash indicates narrowing of the origin of the left common carotid artery.



**Figure 5.** Three-dimensional CT angiography of this patient showed that aortic arch branches were brachiocephalic trunk (arrowhead), left common carotid artery (small arrow), left vertebral artery (large arrow) and left subclavian artery (black arrow) from right to left.

common carotid, left vertebral and left subclavian arteries from right to left.

**Discussion**

Takayasu’s arteritis is a chronic inflammatory disease with an unknown cause. The most common way to detect this disease is to use CT angiography and MRA. The largest lesion observed in this disease is in the form of narrowing of the aorta or its major branches.<sup>5</sup> Other manifestations such as irregular narrowing or occlusion are less common.<sup>6</sup> Various locations of vascular involvement have been reported in different communities. Previous studies have shown that in Northern Europe, the aortic arch branches are mostly involved.<sup>7</sup> A study of people with Takayasu’s arteritis in an Indian community showed that the abdominal aorta is involved in most cases and the patients mostly suffer from hypertension. Another study showed that the abdominal aorta is involved in the Indian and

Japanese populations.<sup>8,9</sup> Investigating the reported cases in the Iranian population indicated a higher prevalence of type I and II in this population.<sup>10</sup> The case introduced in the present study was also consistent with these findings.

So far, the definitive cause for the disease has not been determined. Many researchers believe that Takayasu's arteritis is a multi-agent illness. Factors such as environment and genetics are effective in its occurrence.<sup>11</sup> Various mechanisms are described to explain the onset of the disease. Some researchers believe that the expression of a 65 kDa heat-shock protein in aortic tissue may trigger a cascade of events that cause inflammation.<sup>12</sup>

Prevalence of this disease in families has been studied in some populations. For example, in the Japanese population, the prevalence was observed in 21 families. In addition, observation of Takayasu's arteritis disease in identical twins indicates the effective role of genetic factors in the pathogenesis of this disease. Research on this subject demonstrates the involvement of human lymphocyte antigen (HLA) in this disease. The results of a study on 38 members of 21 families with the disease show that 77% of these people carry the HLA B52 antigen. In another study, the expression of different HLA regions was studied in patients with Takayasu's arteritis and compared with a control group. In this study, there was significant increase in expression of B52, D12 and A24 in patients. The results showed that Haplotype A24-B52-D12 has the highest prevalence in patients with Takayasu's arteritis in the Japanese population. Therefore, it can be said that genetic factors associated with this haplotype play an important role in the development of Takayasu's arteritis.<sup>13,14</sup>

Diagnosis and early treatment of Takayasu's arteritis can prevent the onset of a severe condition in the patient. The progression of the disease can lead to cardiac arrhythmia, stroke and congestive heart failure. These are the most common causes of death from Takayasu's arteritis.

#### Authors' Contribution

Conceptualization: BJK; Methodology: FA; Writing-review & editing: All authors; Funding Acquisition: MHA; and Supervision: BJK.

#### Conflict of Interest Disclosures

The authors have no conflicts of interest.

#### Ethical Statement

There were no ethical considerations to be considered in this research.

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