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*Case Report*

# A case of Takayasu's arteritis with pulsatile neck mass

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## Abstract

Takayasu's arteritis (TA), also known as pulseless disease or occlusive thromboaortopathy, is a form of vasculitis of unknown cause that chiefly affects the aorta and its major branches, most frequently in young women. We describe an 18-year-old female with a soft and pulsatile mass in the left side of her neck.

**KEYWORDS:** Takayasu's Arteritis, Vasculitis, Pulselessness, Aneurysm.

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Takayasu's arteritis (TA) is a chronic inflammation of main arteries which manifests with variable clinical presentations.[1,2] This granulomatous disease imposes a considerable morbidity and mortality to the affected cases.[3] A mass of knowledge about vasculitis, derived from different studies, opened new horizons to the general field of vasculitis that was first reported by Kussmaul and Maier in 1866.[4] Among the vasculitis involving medium and large arteries is Takayasu's arteritis.[5] This chronic and relapsing arteritis, classified as a primary vasculitis, manifests mainly by constitutional symptoms and fatigue.[6-8] The other disease presentations are complications of perfusion insufficiency including muscle pain, claudication, dizziness, decreased arterial pulses, carotidynia and hypertension.[2,9-14]

Criteria for active Takayasu's arteritis include new onset or worsening of two or more of the following: 1) Fever or other systemic features (in the absence of other cause); 2) Elevated erythrocyte sedimentation rate (ESR); 3) Symptoms or signs of vascular ischemia or inflammation (claudication, absent pulse, or carotidynia); and 4) Typical angiographic lesions.[15] Although about 85% of TA patients present active disease, about 15% do not.[15-17]

TA is a form of vasculitis most frequently requiring revascularization procedures.[15] Medical therapy rarely reduces or reverses stenotic lesions. Treating stenotic or aneurysmal lesions may require bypass surgery (especially of stenotic cervicobrachial arteries, coronary arteries, or renal arteries), aortic valve replacement (for aortic regurgitation), or percutaneous transluminal angioplasty (especially for stenotic renal arteries causing hypertension).

Twenty percent of TA patient have a self-limited disease. The rest have a relapsing-remitting or progressive course requiring chronic corticosteroid therapy. Nearly two thirds of patients experience new angiographic lesions.[17] No parameters at disease onset have been shown to predict mortality.[17] The survival is 92.9% at 5 years, 87.2% at 10 years, [16] and 73.5% at 20 years.[17] Congestive heart failure and renal failure are the commonest causes of death.[18] Pregnancy appears to be relatively well tolerated in the presence of good medical care and in the absence of abdominal aortic involvement.[15]

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## Case Report

The patient was an 18-year-old female who referred to Rheumatology Clinic in Alzahra Hospital. She was complaining from low back pain and paravertebral muscle spasm since two weeks prior to admission. The pain was mainly localized in the right side of her waist and this sudden onset of the pain made her troubles walking. Three days later, she found the pain radiating to her left ear and her lower left molar teeth. At this time, she got aware of a painless huge mass in the left side of her neck. She complained from weight loss about 3 kg over 6 months but she denied any fever or other constitutional symptoms. Increasing pain and the mass in the left side of her neck caused her seek for medication. On admission, she was stable and afebrile. Her left radial pulse was undetectable, but other pulses were completely normal. No sign of ischemia was present on her limbs. A soft and pulsatile mass was observed in the left side of her neck. On auscultation, vascular bruit was audible on the carotid arteries. No positive finding was detected in neurological examination, and fundoscopic evaluation of the eyes was also normal. Left and right arm blood pressures were 80/60 and 120/80 mmHg, respectively. Right and left foot had equal pulse pressure (120/80 mmHg). The laboratory data is summarized in table 1.

Color Doppler of carotid arteries showed a left common carotid artery aneurysm with dimensions of about 15×17×21 mm with increased intima and media wall thickness in site of dilation. Computed tomography (CT) angiography revealed aneurysmal dilation at mid common carotid in a length of about 4 cm. Diameter of aneurysm at the most dilated portion was about 15 mm. In addition, diameters proximal and distal to aneurysm were 5 mm and 4 mm, respectively. Left carotid distal to aneurysm was well visualized. Rest of carotid and vertebral arterial system showed normal caliber with no significant stenosis. No definite plaque formation was identified. In the region of aortic area, left vertebral artery was arising from the aortic arch. Left subclavian artery was not visualized. There was also narrowing at the origin of right subclavian artery (Figure 1).

The chest radiography, abdominal sonography, and thyroid gland sonography were normal. Echocardiographic findings were compatible with trivial mitral and tricuspid regurgitation. Based on these vascular data, unequal arm blood pressure, leukocytosis and elevated ESR, the diagnosis of the Takayasu's arteritis was made.

**Table 1.** Laboratory findings of the patient

| | |
|---|---|
| RBC = 4,300,000 | P-ANCA = Negative |
| WBC= 13500 | RF = 56 IU/ml (NL = 10) |
| Hb = 10.7 mg/dl | HBSAg = Neg |
| PLT = 595 × 103/$mm^3$ | HCVAb = Neg |
| ESR = 130 mm/h | Acl-IgG = Neg |
| CRP = 56 mg/l (NL = 6) | Acl-IgM = 11.3 IU/ml (> 7 IU/ml positive) |
| BUN = 17 mg/dl | Serum protein electrophoresis<br>Albumin = 38.4% (52-65) |
| Cr = 0.9 mg/dl | Alpha1 Globulin = 3.7 g/l (2.5-5) |
| C3 = NL | Alpha2 Globulin = 16.9 g/l (7-13) |
| C4 = NL | Beta Globulin = 15.2 g/l (8-14) |
| C-ANCA = Negative | Gamma Globulin = 25.8 g/l (12-22) |

RBC: Red blood cells; WBC: White blood cells; Hb: Hemoglobin; PLT: Platelets; ESR: Erythrocyte sedimentation rate; CRP: Capsid related peptide; BUN: Blood urea nitrogen; Cr: Creatinine; C3: Complement-3; C4: Complement-4; C-ANCA: Cytoplasmic antineutrophil cytoplasmic antibodies; P-ANCA: Perinuclear antineutrophil cytoplasmic antibodies; RF: Rheumatoid factor; HBSAg: Hepatitis B surface antigen; HCVAb: Hepatitis C virus antibody; Acl-IgG: Anti cardiolipin-IgG; Anti cardiolipin–IgM; NL: Normal.

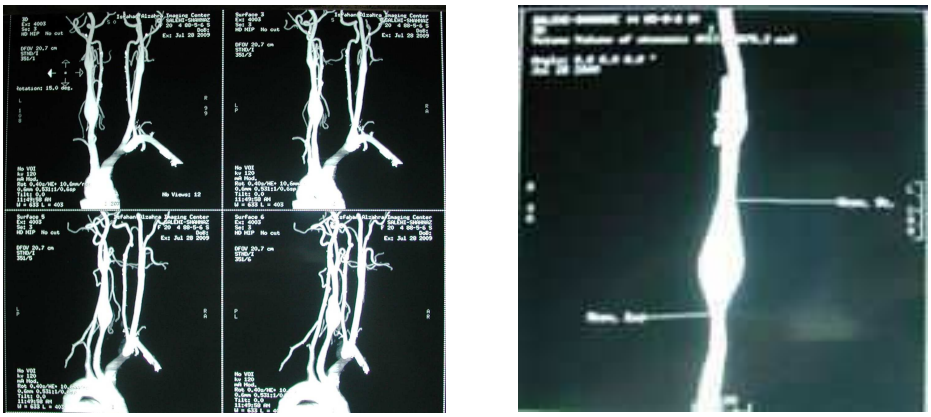

**Figure 1.** Fusiform aneurysmal dilation of mid common carotid in a length of about 4 cm

Heparin infusion was started for the patient (2500 units every 3 hours) followed by oral warfarin therapy. She received prednisolone tab (50 mg) daily (in divided doses). She was also treated with infusions of monthly 700 mg cyclophosphamide pulse. After 6 cyclophosphamide pulses, a significant improvement was seen, constitutional symptoms vanished, neck mass was reduced 50% in size, and ESR got the normal range.

The left radial artery found to have a weak pulse. Totally, she received 6 pulses of cyclophosphamide within six months. Then, she was treated with 15 mg intramuscular methotrexate per week, 1 mg oral folic acid daily, and 80 mg aspirin daily. In addition, prednisolone was tapered. Like any other patient on chronic corticosteroids, to prevent osteoporosis, she was prescribed with calcium, vitamin D, and a bisphosphonate and perform weight-bearing exercises. Modifiable risk factors for atherosclerosis especially hypertension, smoking, inactivity, diabetes, and hyperlipidemia should be treated maximally.

## Discussion

Our case was an interesting one manifesting a neck mass, unilateral radial pulselessness, and significant weight loss. She felt it herself, but no attention was paid. One interesting fact about this patient was the switching phenotype of such a subtle disease to a fulminant course. Absence of ischemic signs in the hand with occlusion of subclavian artery may be a clue toward disease chronicity due to the collateral formation. It is noteworthy that the symptoms and signs of the Takayasu's arteritis fluctuate in severity, but there is a consensus on the systemic manifestations as the presentations of the early disease. Moreover, the presence of signs and symptoms related to occlusion, such as pulselessness and vascular aneurysm, are considered to appear in the later phases of the disease. Chronic inflammation of the vessel wall leads to aneurysm formation, stenosis, or thrombosis.[19] The other interesting observation about our patient was diminished size of the carotid aneurysm after 6 months treatment just by prednisolone and cyclophosphamide which was documented by physical examination and ultrasound of the carotid vessels. This may be partly due to subsidence of the inflammation. Since medical therapy rarely reduces or reverses stenotic lesions, reduced size of the carotid aneurysm by cyclophosphamide pulse seems like an appropriate preparation of the patient for surgical operation if necessary.

Although the pulse was not tangible at first, weak radial pulse was found after treatment. These non-distinguished primary symptoms need high index of suspicion which is achievable by reports from rare and new cases reported by different researchers. New therapeutic strategies derived from these reports may help the clinicians to overcome the present

controversies in the treatment of such poor responsive vasculitis.

## Conclusion

Our patient was an 18-year-old female with a soft and pulsatile mass in the left side of her neck. Radial pulselessness without any other symptom may be a subtle manifestation of the Takayasu's arteritis which needs further follow-up. Typical angiographic lesions, elevated ESR, absence of radial pulse, and carotidynia are always considered as symptoms of active disease and medical therapy must be started as soon as possible.

## Conflict of Interests

Authors have no conflict of interests.

## Authors' Contributions

All authors planned and conducted the study procedure and wrote. All authors read and approved the final draft of the manuscript.

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