



Takayasu's Arteritis: A Case Report

Kristin Agustina¹ , Marsha Darmawan¹ , Putu Patriawan² , Pande Anandasari² ,
Ni Putu Ekawati³ 

¹ Faculty of Medicine, University of Udayana-Sanglah Hospital, Denpasar, Bali, Indonesia

² Department of Radiology, Faculty of Medicine, University of Udayana-Sanglah Hospital, Denpasar, Bali, Indonesia

³ Department of Pathology Anatomy, Faculty of Medicine, University of Udayana-Sanglah Hospital, Denpasar, Bali, Indonesia

ABSTRACT

Takayasu's arteritis is an uncommon idiopathic large-vessels inflammation damaging large arteries such as the aorta and its main branches. Clinical manifestations vary from asymptomatic, bruits, unpalpable pulse, and catastrophic neurological disorders that may develop into life-threatening conditions. We report a 23-year-old male with pain and swelling from his right forearm to his fourth finger with significant Doppler ultrasonography and computerized tomography angiography findings. Imaging modalities are essential especially in patients with unspecific clinical manifestations. Appropriate and rapid diagnosis is important to prevent permanent organ damage.

Key Words: Subclavian artery; Takayasu's arteritis; vasculitis.

Takayasu Arteriti: Bir Olgu Sunumu

ÖZ

Takayasu arteriti, aort ve ana dalları gibi büyük arterlere zarar veren nadir bir idiyopatik büyük damar iltihabıdır. Klinik belirtiler asemptomatik, morluklar, ele gelmeyen nabız ve yaşamı tehdit eden durumlara dönüşebilen katastrofik nörolojik bozukluklardan farklıdır. Bu olgu sunumunda sağ ön kolundan dördüncü parmağına kadar ağrı ve şişlik şikayeti olan 23 yaşında bir erkek hasta, önemli Doppler ultrasonografi ve bilgisayarlı tomografi anjiyografi bulgularıyla sunulmuştur. Görüntüleme modaliteleri, özellikle spesifik olmayan klinik belirtileri olan hastalarda çok önemlidir. Kalıcı organ hasarını önlemek için uygun ve hızlı teşhis önemlidir.

Anahtar Kelimeler: Subklavyen arter; Takayasu arteritis; vaskülit.

INTRODUCTION

Takayasu's arteritis (TA), or aortoarteritis and pulseless disease, is an uncommon idiopathic large-vessels inflammation damaging large arteries such as the aorta, main branches, also the proximal pulmonary artery, coronary arteries, and renal arteries⁽¹⁻³⁾. The primary process of this disease is inflammation and several other etiology include infection and autoimmune^(1,2,4). In East Asia, TA is more common in young women, but similar in both gender worldwide⁽³⁾. The clinical manifestations of TA can be asymptomatic and varies depends on the vessels involved. The asymptomatic disease is usually found during the examination, where there is an unpalpable pulse or bruits in the upper extremities (84-96%), thus it is called the pulseless disease⁽¹⁻³⁾. This condition is associated with claudication of the extremities and blood pressure differences between the two upper extremities. Other symptoms can be found after the initial complaint and may develop into life-threatening situations⁽¹⁾. Appropriate and rapid diagnosis is important to prevent permanent organ damage.

CASE REPORT

A 23-year-old Indonesian male came to the hospital complaining of pain and swelling in his right forearm until his fourth finger in the last 7 months. His symptoms are getting worse during the last three months, his hand turned blue, numb, and there was a wound on his fourth finger of the right hand. The patient denied any history of trauma, no history of fever, chest pain, syncope, or blurred vision. Local upper right extremity examination revealed

Cite this article as: Agustina K, Darmawan M, Patriawan P, Anandasari P, Ekawati NP. Takayasu's arteritis: a case report. *Koşuyolu Heart J* 2021;24(2):148-152.

Correspondence

Kristin Agustina

E-mail: katharinaa.yohana@gmail.com

Submitted: 31.03.2021

Accepted: 20.04.2021

Available Online Date: 26.04.2021

© Copyright 2021 by Koşuyolu Heart Journal.
Available on-line at
www.kosuyoluheartjournal.com

a dry wound on the fourth finger, exposed bone (+), edema, and redness to his 1st until 5th finger. There were tenderness, unpalpable brachial and radial artery pulses of the right upper extremity, and a limited range of movement (ROM) was noticed. The left upper extremity pulse and blood pressure were normal 130/90 mmHg, while the blood pressure in his right arm was undetermined. Oxygen saturation (SpO₂) was not detected in the fingers of his right hand, while it was 97% in the fingers of his left hand. Laboratory tests showed unremarkable results, except for a slight increase in leukocytes (11.100/ μ L) (Figure 1).

The patient was referred to the radiology department to evaluate the cause of his different blood pressure and arterial pulses between his right and left upper extremities. Doppler USG showed partial obstruction to the right subclavian artery until the right proximal axillary artery (Figure 2). There were no visible abnormalities in the arterial system of the left upper limb. There was no deep vein thrombosis in the right and left upper extremities.

Further examination with CTA and arteriography was recommended to the patient. The CTA examination revealed an intraluminal thrombus causing a partial obstruction of the right

subclavian artery (Figure 3). The aorta, right and left common carotid artery, subclavian artery, and left vertebral artery were normal. There was no hypertrophy of the right thoracic outlet muscles nor organ compression around the right thoracic outlet against the right subclavian artery and vein. Arteriography revealed partial obstruction of the right subclavian artery (Figure 4). The right common carotid artery and vertebral artery were normal. Based on these clinical and radiological findings, the patient was diagnosed with TA.

Therapy for this patient is immunosuppressants, analgesics, and a thrombectomy. But unfortunately, the thrombectomy failed but clinicians managed to take a sample of the intraluminal thrombus and sent it to pathology. Later on, an innominate brachial artery bypass surgery with sternotomy was planned for this patient but the family refused to go forward with the procedure and the patient went home with antibiotics, immunosuppressants, and analgesics. The result from pathology suggests arteritis with thickened tunica intima layer, recanalization, and distribution of lymphocytes and one or two plasma cells (Figure 5).



Figure 1. The clinical picture of the patient's right hand and fingers.

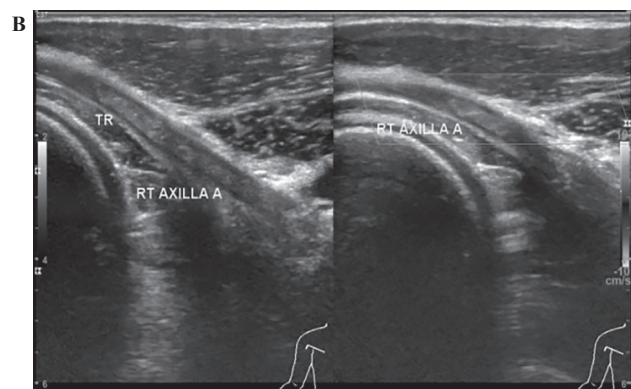
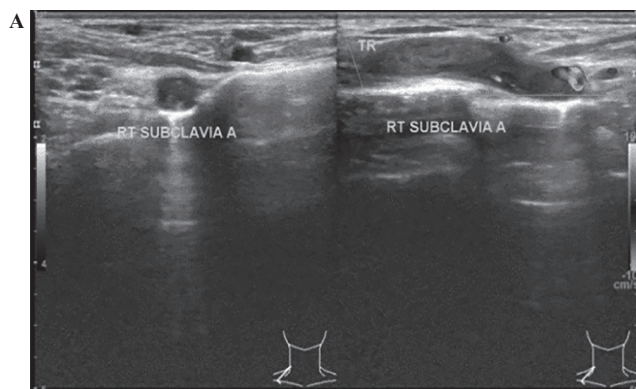


Figure 2. The Doppler USG examination result: (A) A partial intraluminal thrombus, (B) Little to no flow detected with CDUS in the supraclavicular region of the right subclavian artery until the right proximal axillary artery that causes a partial obstruction.

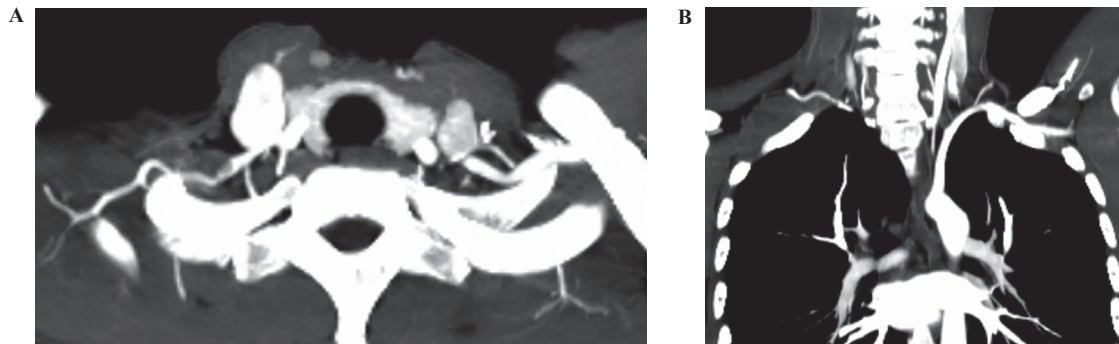


Figure 3. CT scan Angiography, (A) Axial and (B) Coronal view of post-contrast CTA showed an intraluminal thrombus in the right subclavian artery (Blue arrow) causing partial obstruction to the artery.

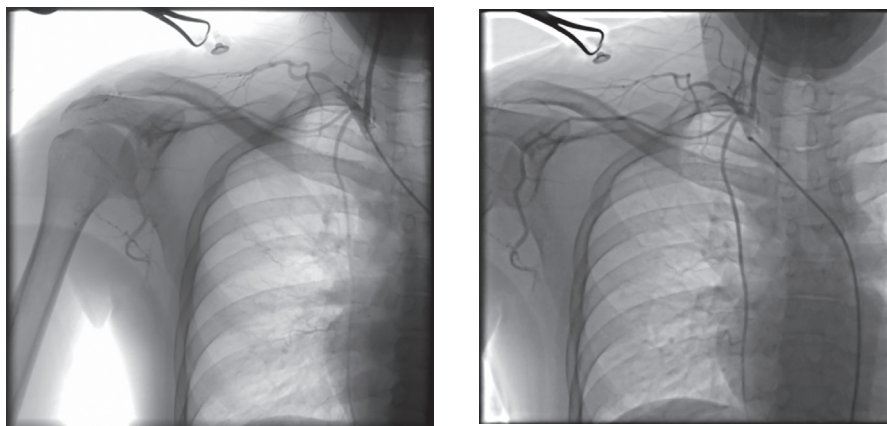


Figure 4. Arteriography showing a partial obstruction in the right subclavian artery. The right common carotid artery and right vertebral artery were normal.

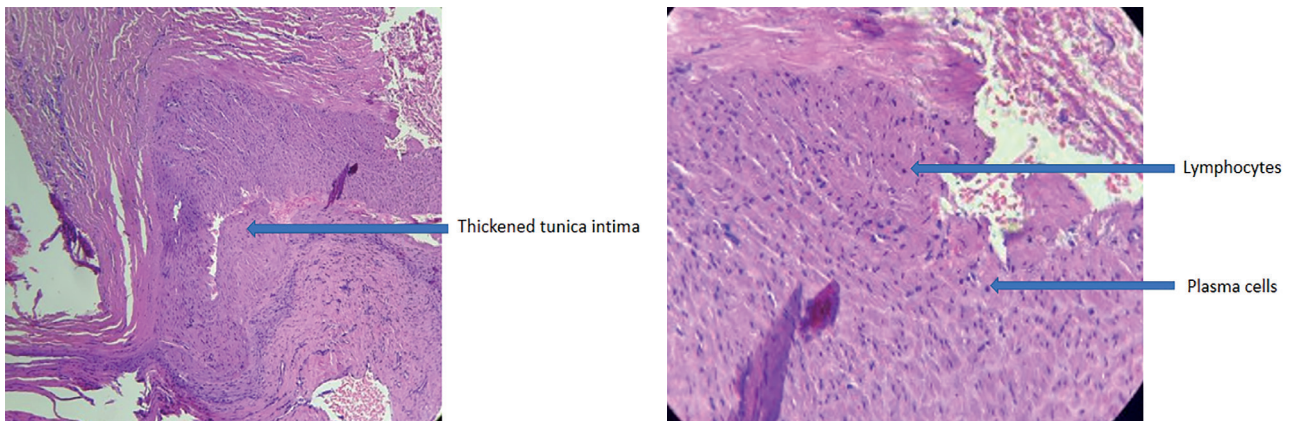


Figure 5. The vascular tissue sample was composed of the tunica intima, media, and adventitia layers. It was showed thickened tunica intima layer, recanalization, and distribution of lymphocytes and one or two plasma cells.

DISCUSSION

Takayasu's arteritis is an uncommon, long-lasting, idiopathic, large-vessels inflammation that is commonly found in young women, mostly in East Asia^(5,6). However, data show similar incidence in both genders worldwide⁽⁶⁾. The ratio of TA in women to men varies from 9:1 in reports from Japan, 6.9:1 in Mexico, and 1.2:1 in Israel^(7,8). Inflammation to the walls of

blood vessels causes thickening, fibrosis, stenosis, occlusion, and dilatation of the blood vessels. Further acute inflammation can damage the medial artery and causing aneurysms formation in the affected vessels^(1,3,6). This disease usually occurs in the 2nd-3rd decade of life and the diagnosis is often late from months to years since the first onset of the symptom⁽³⁾. About 25% of cases can occur before the age of 20 years old and 10-20% occur after the age of 40⁽⁸⁾.

The clinical course of TA is not specific and is classified as an early active inflammatory phase and an advanced long-lasting phase. The active phase is characterized by systemic symptoms, such as high temperature, discontent, loss of appetite, weight loss, headache, dizziness, pain to joints, and skin rash, that lasts for weeks to months, and episodes of remission and relapses can occur. The acute phase may not happen to all patients therefore the diagnosis of TA is rarely made in this phase. The advanced phase of TA results from stenosis or occluded arteries and organ ischemia. The clinical manifestations vary depending on the vessels involved. For example, arterial hypertension and renal failure can happen due to renal artery stenosis^(1,2). Our patient did not have an early acute phase. Advanced phase manifestations in this patient were unpalpable pulses of the brachial and radial arteries of the patient's right upper extremity, edema, and erythema of his 1st until 5th right-hand fingers and ulcers on his 4th right-hand finger.

If TA is suspected, vascular imaging, either invasive or non-invasive, should be performed. Although arterial angiography is the standard diagnosis and evaluation of TA, it has been generally replaced by CTA or MRA. According to the American College of Rheumatology (ACR), at least three of the six criteria must be met for a definite diagnosis of TA (Table 1) and will result in a sensitivity of 90.5% and specificity of 97.8%^(1,2,6,9).

Our patient was a 23-year-old male that was consistent with TA predilection of onset, which is in the 2nd and 3rd decade of life. There are five of the six ACR criteria found in our patient and the patient also had an elevated leukocyte count which reflects the underlying inflammatory process. Unfortunately, laboratory test results are usually not specific for the diagnosis of TA. A study conducted in South Korea by Park MC et al. found that 23% of patients showed normal laboratory results even in the active phase of TA⁽¹⁰⁾. There was no diagnostic serological test for the diagnosis of TA⁽²⁾.

Based on the vessel involvement, the latest angiographic classification divides TA into five types⁽¹¹⁾ (Figure 6):

Type I: involves only branches of the aortic arch

Type II:

Type IIa: involves ascending aorta, aortic arch, and its branches

Type IIb: involves ascending aorta, aortic arch, and its branches, thoracic descending aorta

Type III: involves thoracic descending aorta, abdominal aorta, and/or renal arteries. The ascending aorta, aortic arch, and its branches are not affected.

Type IV: only involves the abdominal aorta and/or renal arteries

Type V: combined features of type IIb and IV

The involvement of coronary and pulmonary arteries should be indicated as C (+) or P (+). Type V has been documented as the most common type of TA. In our patient, the partial obstruction occurs in the right subclavian artery, which is consistent with the TA type I classification features⁽¹¹⁾.

CONCLUSION

Takayasu's arteritis is an uncommon idiopathic large vessel inflammation that occurs frequently in young women, in the 2nd-3rd decades of life. The diagnosis of TA is rarely done in the early phase due to its unspecific manifestations. This case report aims to emphasize the importance of imaging modalities in the diagnosis of TA. Typical clinical findings and appropriate imaging techniques are required to make an accurate diagnosis and assist in deciding and prioritizing its management, especially in a patient with non-specific clinical manifestations.

The pointed out the renewed TA information for the researcher is accurate diagnosis and early diagnosis is paramount in managing TA. This also can help in deciding

Table 1. The diagnostic criteria of Takayasu's arteritis according to ACR (1990)⁽⁹⁾

Criteria	Definition
Age at disease onset ≤ 40 years	Development of symptoms or findings related to Takayasu's arteritis at age ≤ 40 years
Claudication of the extremities	Development and worsening of fatigue and discomfort in muscles of one or more extremities while in use, especially the upper extremities
Decreased brachial artery pulse	Decreased pulsation of one or both brachial arteries
Blood pressure difference > 10 mmHg	Difference of > 10 mmHg in systolic blood pressure between right and left arms
Bruit over subclavian artery or aorta	Bruits audible on auscultation over one or both subclavian arteries/abdominal aorta
Arteriogram abnormality	Arteriographic narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities, not due to arteriosclerosis, fibromuscular dysplasia, or similar causes; changes are usually focal or segmental

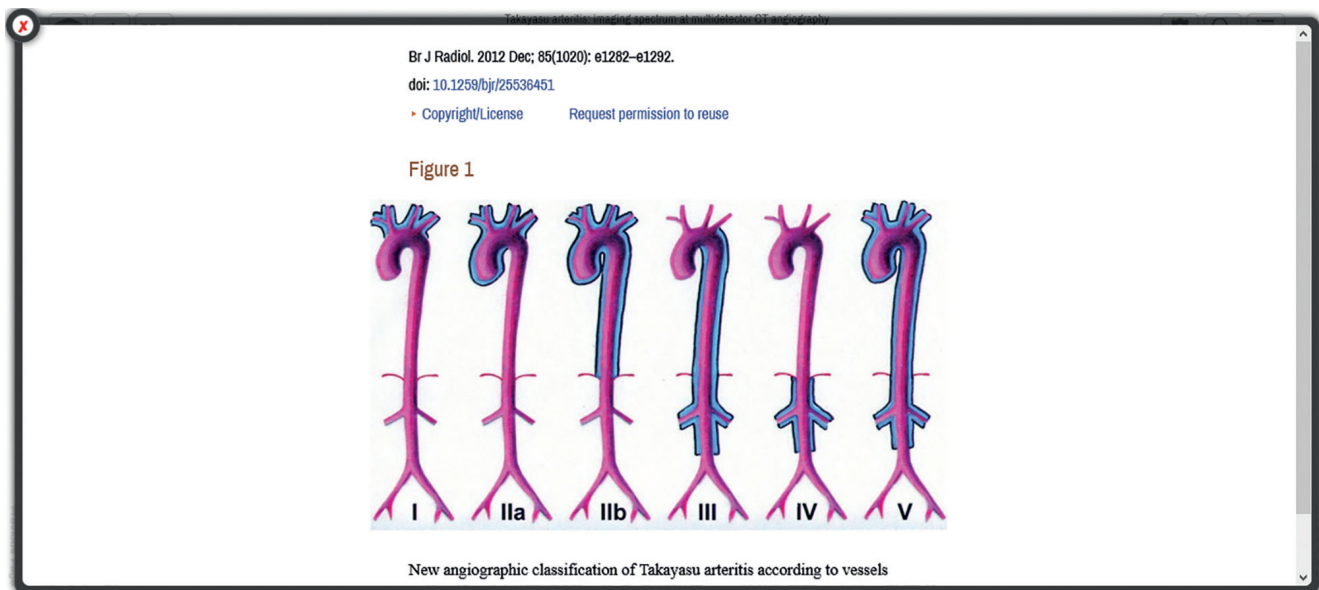


Figure 6. The latest angiographic classification of Takayasu arteritis is based on vessel involvement (Hata et al, 1996).

and prioritizing its management in a patient with non-specific clinical manifestations.

Informed Consent: Informed consent was obtained.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept/Design - KA; Analysis/Interpretation - MD; Data Collection - PP; Writing - KA; Critical Revision - PA; Statistical Analysis - PA; Overall Responsibility - MD; Final Approval - All of authors.

Conflict of Interest: The authors have no conflicts of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

REFERENCES

1. Khan M, Banoo H. A case report of Takayasu's arteritis. *Med Today* 2013;24:79-81. [\[Crossref\]](#)
2. Shelma R, Paul W, Sharma CP. Chitin nanofibre reinforced thin chitosan films for wound healing application. *Trends Biomater Artif Organs* Published online 2008. [\[Crossref\]](#)
3. Johnston SL, Lock RJ, Gompels MM. Takayasu arteritis: a review. *J Clin Pathol* 2002;55:481-6. [\[Crossref\]](#)
4. Espinoza JL, Ai S, Matsumura I. New insights on the pathogenesis of Takayasu arteritis: revisiting the microbial theory. *Pathog (Basel, Switzerland)* 2018;7:73. [\[Crossref\]](#)
5. Mason JC. Takayasu arteritis-advances in diagnosis and management. *Nat Rev Rheumatol* 2010;6:406-15. [\[Crossref\]](#)
6. Dhar Dwivedi AN. Takayasu's arteritis-a case report and a brief review of diagnostic Imaging. *Clin Radiol Imaging J* 2019;3. [\[Crossref\]](#)
7. Landhani S, Tulloh R, Anderson D. Takayasu disease masquerading as interruption of the aortic arch in a 2 year old child. *Cardiol Young* 2001;11:244-6. [\[Crossref\]](#)
8. Brunner J, Feldman BM, Tyrrell PN, Kuemmerle-Deschner JB, Zimmerhackl LB, Gassner I, et al. Takayasu arteritis in children and adolescents. *Rheumatology* 2010;49:1806-14. [\[Crossref\]](#)
9. Arend WP, Michel BA, Bloch DA, Hunder GG, Calabrese LH, Edworthy SM, et al. The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis. *Arthritis Rheum* 2010;33:1129-34. [\[Crossref\]](#)
10. Park MC, Park YB, Jung SY, Lee KH, Lee SK. Anti-endothelial cell antibodies and antiphospholipid antibodies in Takayasu's arteritis: correlations of their titers and isotype distributions with disease activity. *Clin Exp Rheumatol* 2006;24(2 Suppl 41):S10-6. [\[Crossref\]](#)
11. Hata A, Noda M, Moriwaki R, Numano F. Angiographic findings of Takayasu arteritis: new classification. *Int J Cardiol* 1996;54:S155-S163. [\[Crossref\]](#)