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## Case Report

# Takayasu Arteritis and Lower Limbs Claudication: A Rare Presentation

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### ABSTRACT

Takayasu's arteritis is a rare inflammatory disease of large- and medium-sized arteries which predominantly affects women of middle-aged. We report an unusual case of Takayasu arteritis in a 54-year-old woman presenting with progressive lower limbs intermittent claudication. Aneurysmal and stenotic vascular lesions involving both common iliac arteries were identified which also represents a very uncommon anatomical distribution in this pathology. In addition, we share our experience with endovascular intervention for stenotic lesions.

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### Introduction

Takayasu's arteritis (TA) is a rare progressive inflammatory disease of large- and medium-sized arteries. As it commonly involves brachiocephalic vessels particularly the aorta and its main branches, it is also known as "pulseless disease" or "aortic arch syndrome" [1]. In this paper, we report a case of a TA middle-aged patient with progressive bilateral lower limb intermittent claudication which is an atypical clinical presentation of this disease.

### Case Report

A 54-year-old woman with Takayasu arteritis diagnosed at age 34 by clinical and histopathological examination presented to our sanatorium with two days of fever, arthralgias and lower limbs pain associated with coldness, numbness, and paresthesia's. She referred to have 1-year history of bilateral lower limb intermittent claudication (Fontaine's stage IIa) which was progressively decreasing to 100 mts (Fontaine stage IIb) in the last three months.

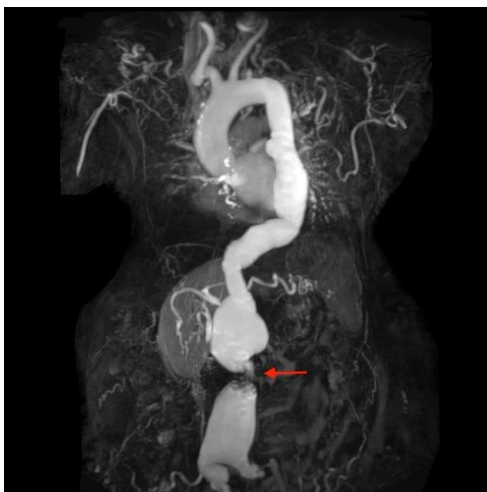
A medical record of tobacco abuse, hypertension, chronic renal insufficiency, and right renal stenting was documented. Current medical treatment included prednisone 60 mg/day, cilostazol 200 mg/day and carvedilol 25 mg/day. Upon physical examination, lower limbs examination showed coldness and paleness bilateral feet with capillaries

refill time delayed and absence of distal pulses. Diminished left radial pulse with a discrepancies in blood pressure (in mmHg) in the right arm (125/55) and left arm (140/65) was also appreciated. Hematology and biochemistry revealed no abnormality except for an elevated erythrocyte sedimentation rate (ESR) of 135 mm/h and C reactive protein (CRP) of 42 mg/L. Electrocardiographic (ECG) and echocardiographic finding were normal.

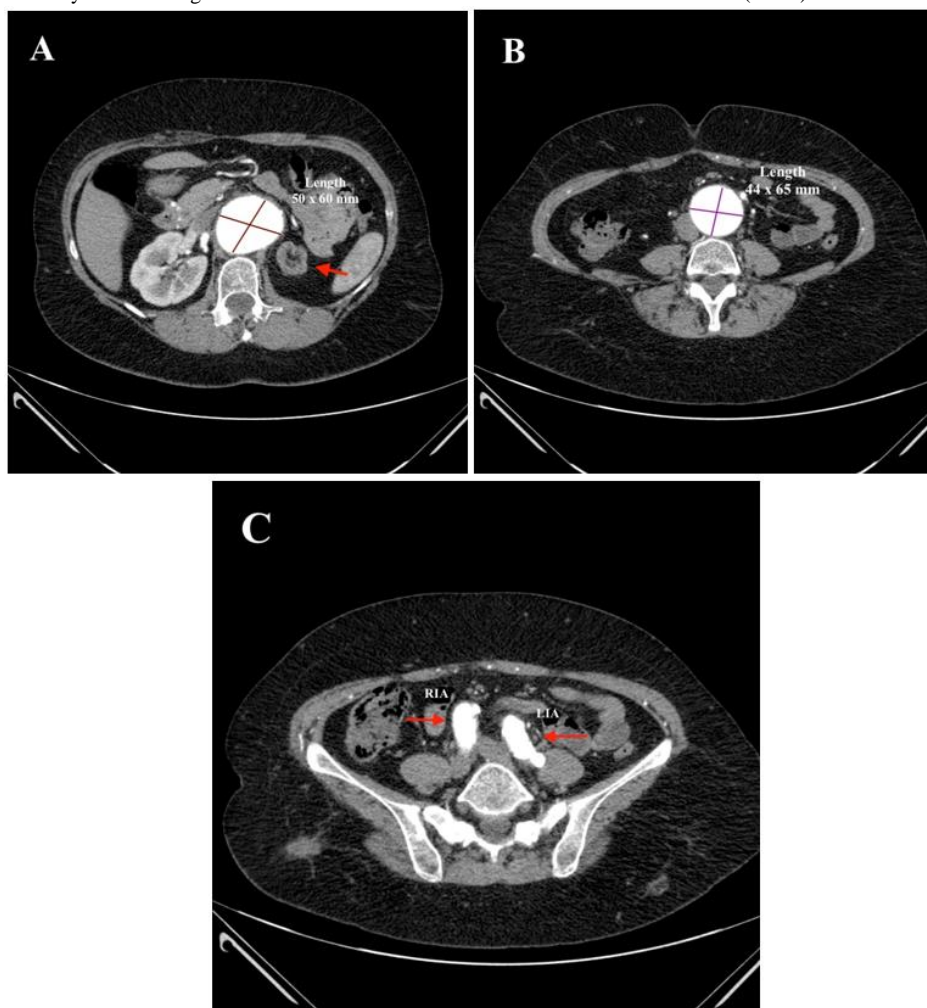
Doppler exploration of the lower limbs showed no significant stenosis but monophasic waveform at the bilateral common femoral, superficial femoral, popliteal, posterior tibial and dorsal pedis arteries with an ankle-brachial index (ABI) of 0.5 in each limb. CTA scan of thorax, abdomen and pelvis demonstrated occlusion in the left subclavian artery as well as chronic obstructions in celiac trunk and superior mesenteric artery. Although little development of collateral circulation was observed, patient did not re-fer any symptom of abdominal angina. Left renal artery occlusion with ipsilateral kidney atrophy was appreciated but good stent expansion and patency was observed at right renal artery.

A fusiform dilatation extended from descending thoracic aorta to aortic bifurcation (Crawford type II classification) was detected (Figure 1). The maximum anteroposterior diameter of the aneurysm was 50 x 60 mm below the diaphragm. A severe and focal stricture distal to the renal arteries followed by another dilatation measuring 44 x 65 mm with extension to the iliac arteries was observed. (Figures 2A-2C)

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**Figure 1:** CTA scan on coronal plane showing a fusiform dilatation extended from descending aorta to the iliac arteries, consistent with a Crawford type II thoracoabdominal aortic aneurysm measuring 60 mm in maximum diameter with an infrarenal focal stenosis (arrow).



**Figure 2:** CTA scan on axial plane. **A)** Shows abdominal aortic dilatation measuring 50 x 60 mm before stenotic lesion. Left Kidney atrophy (arrow). **B)** shows abdominal aortic dilatation measuring 44 x 65 mm after stenotic lesion. **C)** shows right and left iliac common artery aneurysm (arrow). RIA: Right Iliac Artery; LIA: Left Iliac Artery.

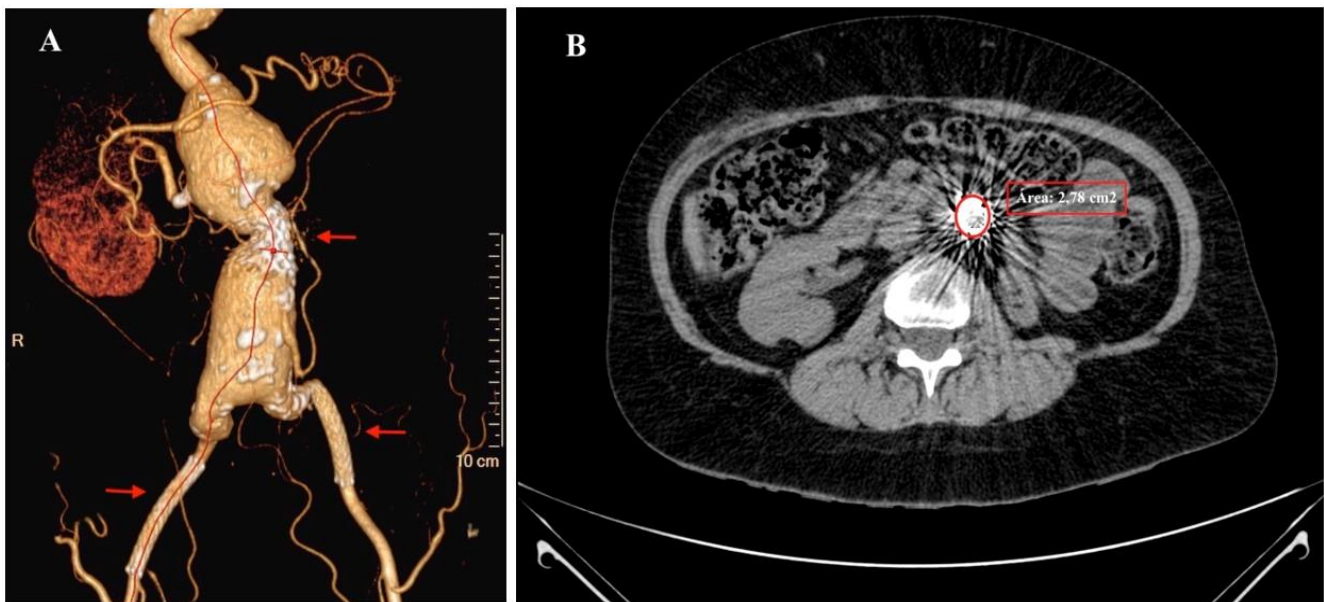
In the first instance, medical treatment with methylprednisolone 500 mg/d for 3 days (15 mg/kg) with subsequent maintenance of prednisone 60 mg/d (1mg/kg) and methotrexate at a dose of 8 mg/kg/week were indicated. Ischaemic symptoms and signs improved at five days after initial therapeutic and inflammatory parameters normalized within 3-weeks. Since the presence of peripheral artery disease symptoms which interfered with the patient's lifestyle develop, surgical intervention was considered for vascular lesions after clinical remission was obtained.

Our initial decision making was open surgery due to anatomy unsuitable for EVAR and inadequate stent graft landing zones. Given that patient did not accept a conventional open procedure, an endovascular intervention was proposed with the aim to improve the run off and achieve symptoms relief. We adopted the strategy of repair the stenotic stricture by using a balloon expandable covered stent (CP Stent 20 x 40 mm). Because of tortuosity and stenotic iliac lesions, a bilateral femoral access was performed by using two different-sized self-expanding covered stents, 8 x 61 mm for the left common iliac artery and 9 x 61 mm for the right common iliac artery (Epic™ Vascular Self Expanding Covered Stent System - Boston Scientific, USA). Systolic blood pressure

(SBP) gradients values showed a decreased from 30 mmHg before stent placement to 5 mmHg after stenting.

Arterial duplex performed two days after procedure informed an ABI of 0.7 on the left and 0.65 on the right. Clinical presentation, biomarkers and imaging findings improved. Methylprednisone was gradually reduced at a maintenance dosage of 15 mg/day and patient was discharged from hospital on the third postoperative day. Medical regiment included methylprednisone at a maintenance dosage of 15 mg/day, aspirin 100 mg and clopidogrel 75 mg daily.

Annual follow-up assessment was performed by duplex and CTA scan. At three-years follow-up visit, patient remained free of symptoms. Limb claudication was referred to > 600 mts. Upon physical examination, lower limbs distal pulses were positive with a normal capillaries refill time and signs of good perfusion. Her blood ESR level was 6 mm/hr and CRP was 0.1 mg/dL. Duplex arterial ABPI determination was of 0.75 and last CTA scan showed good stent patency rate (intra-stent area: 2,78 cm<sup>2</sup>) with no aneurysmal diameter changes (Figure 3A & 3B). Methylprednisone dose after discharge was not varied. The patient gave full consent for data and imaging use.



**Figure 3:** A) CTA scan on coronal plane showing endovascular treatment (arrows) performed by using covered stents. B) CTA scan on axial plane showing intra-stent area (2,78 cm<sup>2</sup>) after endovascular treatment.

## Discussion

Takayasu arteritis (TA) is a rare systemic vasculitis of large- and medium-sized arteries with an overall annual incidence of 0.3-3.4 per million and prevalence of 0.9-4.0 per million. Although most commonly seen in Japan and South East Asia now it has been recognized worldwide. It usually presents in the 2nd or 3rd decade of age with a female/male ratio ranges from 1:9 to 1:5 [1].

The diagnosis is based on a set of symptoms, signs and imaging findings according to criteria proposed by The American College of Rheumatology in 1990 [2]. Following angiographic classification, our case may be labelled as type IV as imaging revealed involvement of

thoracic descending and abdominal aorta. Although not typical, lower limb vascular lesions and symptoms may manifest on these group of patients. To date not many cases describing lower extremity claudication have been published demonstrating that it is certainly quite infrequent [1-10]. In a recent large series lead by Mwipatayi and his colleagues, lower limb symptoms were found in only 15% of patients [1]. Similarly, Dong *et al.*, in 105 patients with Takayasu's arteritis, detected only 9 patients with lower limbs arteriopathy from whom five presented with moderate to severe intermittent claudication [7]. Another large cohort study by Sato *et al.* informed 16% and 33% of patients with lower limbs claudication at the time of diagnosis and then during follow-up respectively while Saadoun *et al.* reported 34% of patients with arterial claudication of lower extremities on his 79-cohort group [9, 10].

Conforming to various reports, the most frequent type of vascular lesion in TA patients seems to be stenosis although aneurysms may also develop. A large series of patients with Takayasu's arteritis showed 93% of stenotic lesions whereas aneurysmal involvement was presented in only 46% [1]. These lesions tend to occur primarily in the ascending aorta, aortic arch and/or its branches while the abdominal aortic bifurcation including iliac arteries is rarely affected [2, 7]. Regarding femoral artery lesions in TA patients, literature has shown that they are even less frequent. [1, 3, 5, 7]. In extensive series published, the percentage of cases with common or superficial femoral artery involvement were between 7-9%. [1, 6].

Patients with abdominal aortic-iliac or femoral involvement can develop severe claudication which could be poorly tolerated. However, since the development of lesions usually is slow and over a long period of time, collateral formation usually occur and, therefore, is not common to present severe ischaemic symptoms (rest pain, ulcerations, or gangrene) which would need an emergency or urgent intervention. Surgical decision making in patients with TA is complex and should be carefully assessed. These patients are frequently young and because of the multi-arterial involvement by the disease itself, they may have significant carotid, renal, cardiac disease or other problems associated that could affect the overall surgical outcome.

Literature agrees that surgical procedures in TA patients should be performed in select clinical scenarios and when the patient is in remission in order to avoid complications (restenosis, anastomotic failure, thrombosis, hemorrhage, infection) due to inflammation [8, 10]. However, the concept of remission is not often clear and still not well defined. Presence of ischaemic symptoms does not always indicate active inflammation of the vessel wall and asymptomatic patients may present active lesions. Taking this fact into consideration, medical therapy with immunosuppression is suggested and recommended during the perioperative period to attain better vascular outcomes [8, 10].

Surgery in patients with TA should be considered only when cardiovascular complications or severe peripheral artery disease symptoms which could affect patient's prognosis or significantly interfere with the patient's lifestyle develop. The optimum revascularization method for these patients is still controversial, the decision as which is the best approach to use remains divided. Some authors still opt for open surgery more often because they consider the recurrence rate of endovascular approaches to be very high. However, long-term large cohort study have reported similar patency rates between endovascular and surgery repair groups suggesting that both methods are safe and efficient [1, 8, 10].

## Conclusion

This paper shows that although uncommon lower limb intermittent claudication may present in patients with TA and should be considered

on young patients as differential diagnosis. A detailed medical history, appropriate physical examination and imaging study are primary elements to achieve a prompt diagnosis and accomplish an adequate and effective treatment. Endovascular approach could be a feasible and alternative treatment option for patients with ischaemic vascular complications.

## Conflicts of Interest

None.

## Author Contributions

Study conception: all authors; Data collection: all authors; Analysis: all authors; Investigation: all authors; Manuscript preparation: all authors; Critical review and revision: all authors; Final approval of the article: all authors.

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