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## Case Report

# An unusual case of Takayasu arteritis presenting with persistent cough in a young female patient: A case report <sup>☆,☆☆</sup>

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## ABSTRACT

Takayasu arteritis (TA) is a chronic granulomatous inflammatory arteritis of large vessels. Females aged 20–40 are usually affected and the manifestations can range from asymptomatic disease to major cardiovascular and neurological abnormalities. Herein, we present a case of a 20-year-old female who had a persistent cough as the initial manifestation of an underlying TA. She had a free past medical history. The patient sought medical help multiple times and took many antibiotics with no improvement. Laboratory tests showed a marked elevation in inflammatory markers so the patient was admitted for further investigations. CT scan showed a circumferential mural thickening involving mainly the descending thoracic aorta which is highly suggestive of an underlying chronic granulomatous vasculitis behind this. Our patient was treated with a combination of corticosteroid and azathioprine and showed substantial improvement after 2 weeks. TA can present with various symptoms. Therefore, patients presenting with atypical symptoms and late-onset vascular symptoms need a high degree of suspicion with close follow-up to allow early detection of any complication.

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## Introduction

Takayasu arteritis (TA) is a chronic granulomatous inflammatory arteritis of large vessels with a predilection for the aorta

and its branches, pulmonary and coronary arteries [1]. It is usually detected in young females aged 20–40 with predominance in countries such as India and Japan [2].

Manifestations of TA are due to organ ischemia which can range from asymptomatic diagnoses to major neurolog-

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ical and cardiovascular manifestations [3]. The most common manifestations are claudication pain and fever of unknown origin [4]. The reported incidence of neurological symptoms such as headache, dizziness, visual disturbance or loss of consciousness, stroke, and transient ischemic attack (TIA) varies from 57% to 80% [5]. Many of the symptoms of TA are nonspecific and closely resemble many diseases causing diagnostic dilemmas. It is quite rare for TA to present with cough, dyspnea, and pleural effusion in the absence of vascular symptoms. Here, we report a case of TA in a young female patient who presented with a persistent cough without any vascular symptoms.

This case illustrates the diagnostic challenge faced by our crew when confronted with these atypical manifestations and the need to consider this devastating disease as a possibility.

## Case presentation

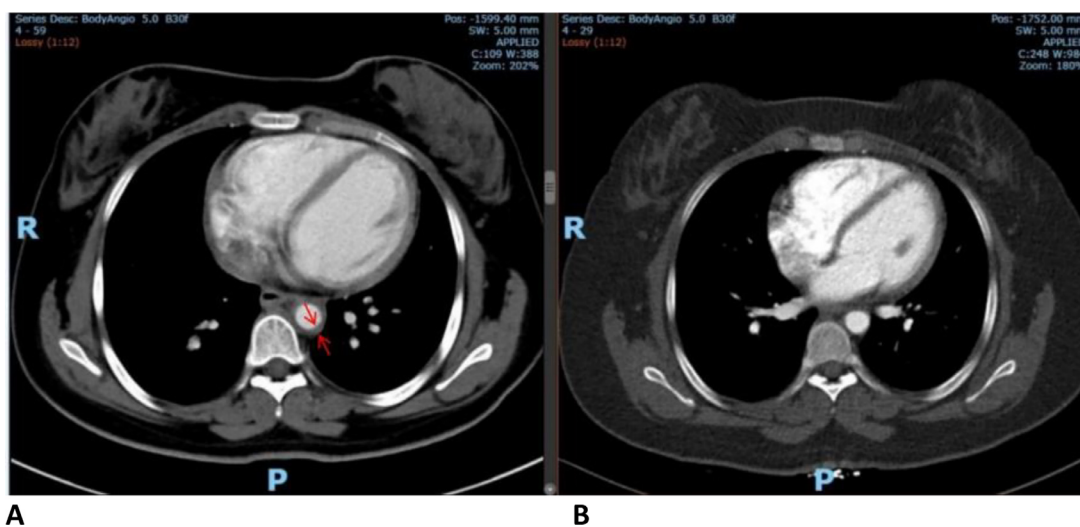
A 20-year-old previously healthy, nonsmoker Caucasian female presented to the local hospital with a chronic cough associated with whitish sputum lasting about 5 months that was associated with a fever of 38.5°C. She also reported that her clothes have become loose although she did not track her weight before visiting us. The patient denied any history of night sweats, blurring vision, bruises, dizziness, headache, nausea, vomiting, or sore throat. Before coming to our hospital, the patient sought medical advice at multiple outpatient clinics and was diagnosed with atypical pneumonia, viral upper respiratory tract infection, and acute bronchitis. She was given multiple courses of antibiotics, antipyretics, and inhalers. However, no improvement was noticed in her symptoms. The patient did not have a past or contact history of tuberculosis. On physical examination, the patient appeared

to be ill and wasted. Her vital signs at presentation were as follows: respiratory rate of 18/min, blood pressure was 110/75 mm Hg with no discrepancy between the right and left arms, pulse rate was 90 beats/min with no radio-radial delay. All peripheral pulses were palpable and the body temperature was 38.5°C. On auscultation, the heart sound was clear with no murmurs or added sounds. Lung sounds were normal with no wheezes or crackles.

Preliminary investigations showed markedly elevated inflammatory markers: erythrocyte sedimentation rate (ESR) was 110 mm/h (0-29 mm/h), C-reactive protein (CRP) was 122 mg/dL (negative < 0.9 mg/dL), platelets count was  $662 \times 10^9/L$  ( $150-400 \times 10^9/L$ ), and hemoglobin was 9.2 g/dL (12.1-15.1 g/dL). The patient was admitted to the hospital for further investigations.

Electrocardiographic (ECG) was normal. Additionally, a 2D echocardiogram was done and showed normal left ventricular function (Ejection Fraction: 60%) with no regional wall abnormality. Abdominal ultrasound was also normal. The patient had a negative history of COVID (no history of infection or vaccination). A bronchoscopy was not done. At this point, the case was a dilemma between lymphoma and vasculitis. Therefore, a computerized tomography (CT) scan with intravenous contrast (IV) contrast of the chest, abdomen, and pelvis was done to evaluate the arteries and lymph nodes. CT of the thorax revealed circumferential mural thickening involving mainly the descending thoracic aorta with no areas of appreciable stenosis. These findings were highly suggestive of aortitis and an underlying chronic granulomatous vasculitis, TA, behind these symptoms (Fig. 1A).

She was started on IV methylprednisolone (250 mg) for three days during the admission then she was discharged on prednisone (20 mg) per os (PO) 3 times per day and azathioprine (50 mg) PO 1 time per day for 6 months. The patient showed significant clinical improvement within 2 weeks of treatment initiation.



**Fig. 1** – Thoracic CT scan with IV contrast/arterial phase shows circumferential mural thickening involving mainly the descending thoracic aorta (red arrows) (A), with no areas of appreciable stenosis. The other image shows the complete resolution of the previously noted mural thickening after six months of treatment (B).

The patient was followed up for 6 months with detailed clinical and laboratory assessments. Her lab tests showed a significant decrease in ESR 30 mm/h (0–29 mm/h) and CRP 5 mg/dL (negative < 0.9 mg/dL). Chest CT angiography was performed and showed complete resolution of the previously noted mural thickening (Fig. 1B).

## Discussion

TA was first described by an ophthalmologist, Dr Mikito Takayasou in 1908 [6]. TA is a recognized, but rare form of large vessel vasculitis with high prevalence in countries such as India and Japan [3,7]. TA is also known as the “pulseless disease” or “aortic arch syndrome” and is thought to be of autoimmune origin [4]. It is 10 times more common in females than in males of 10–40 years of age [7]. There are 6 cases of TA per 1000 people worldwide. However, the prevalence of TA in Palestine is yet unknown [3].

TA can be divided into 2 phases: the initial prepulseless systemic phase and a later occlusive phase [4,8]. In the initial prepulseless phase, patients present with constitutional symptoms, fever, and arthralgia [8]. On the other hand, in the occlusive phase, patients present with limb claudication, neurological symptoms, and hypertension [4].

We report the case of a patient with an unusual initial presentation. She had a chronic cough with whitish sputum for 5 months. A literature search revealed 1 case of TA that presented with pulmonary edema due to heart failure [4]. A high index of suspicion is needed to identify the atypical presentations of TA. It is crucial to diagnose this disease early and to initiate treatment upon diagnosis. Our case illustrates the possibility of having a long initial phase before the onset of vascular symptoms.

Angiography was considered the gold standard imaging study to evaluate the disease extent [9]. However, the noninvasive tests have been gradually surpassing angiography in diagnosing TA [9]. Vessel wall edema and thickening are better indicators of early vascular involvement than narrowing [9].

Treatment of TA is challenging. Recent studies support the use of steroids and immunosuppressive therapy (azathioprine, methotrexate, cyclophosphamide) in treating TA [8]. For critical vessel narrowing, surgical revascularization techniques can be offered [9].

Our patient responded to a combination of methylprednisolone and azathioprine. Her condition markedly improved after 6 months of follow-up.

## Conclusion

TA can present with various symptoms. Patients presenting with atypical symptoms and late-onset vascular symptoms need a high degree of suspicion with close follow-up to allow early detection of any complications.

## Ethical approval and consent to participate

Approval was obtained from the Institutional Review Board of An-Najah National University.

## Consent for publication

Written informed consent was obtained from the patient.

## Authors' contributions

YA and SM: Wrote the manuscript. MM and AA: Performed the radiological examination. RH: Management and follow-up of the case. All authors read and approved the final manuscript the final manuscript.

## Patient consent

Written informed consent was obtained from the patient.

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