Case Report

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Takayasu arteritis presenting with large cerebral infarction in a 39-year-old Syrian woman: a case report

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Introduction and importance: Takayasu Arteritis (TA) is a rare chronic inflammatory disease of unknown etiology that primarily affects large vessels, such as the aorta and its major branches. The disease typically presents with diverse symptoms, depending on the site and degree of arterial lesions. Delayed diagnosis is common, especially in younger populations.

Case presentation: A 39-year-old Syrian female presented with an initial stroke. She had no prior medical history and was otherwise healthy. On examination, she had an absent left radial pulse, a carotid bruit, and muscle weakness. Blood tests showed an elevated ESR and CRP. Computed tomography of the brain revealed a right large cerebral infarction. Multislice computed tomography angiography showed diffuse arterial wall thickening, stenosis, and occlusion of several major vessels, including the left internal carotid artery, right internal carotid artery, and left subclavian artery.

Clinical discussion: The patient was diagnosed with TA based on the American College of Rheumatology criteria. She was treated with prednisolone, methotrexate, and aspirin, and her symptoms improved significantly.

Conclusion: This case highlights the importance of considering TA in the differential diagnosis of ischemic stroke, especially in young patients with atypical presentations. Early identification and management are essential to preclude critical sequelae.

Keywords: hemiplegia, right cerebral infarction, stroke, takayasu arteritis

Introduction

Takayasu Arteritis (TA) is a rare chronic inflammatory arteritis affecting large vessels, predominantly the aorta and its main branches, causing wall thickening, fibrosis, stenosis, thrombus formation, dilatation, or aneurysms of the vessels[1–4]. This rare disease primarily occurs in East and South Asia, as well as Mexico[1], with varying clinical presentations among ethnicities[5]. In general, the initial manifestations include limb claudication, blood pressure asymmetry, heart murmurs, decreased arterial pulse, arterial hypertension, and constitutional symptoms. Lack of specific features, particularly in childhood, causes delayed diagnosis[6,7]. Other manifestations include vascular bruits, particularly affecting the carotids, subclavian, and abdominal vessels, aortic regurgitation resulting from dilatation of the ascending aorta, congestive heart failure due to hypertension, neurological features secondary to hypertension and/or ischemia, including seizures, stroke, amaurosis, and pulmonary vasculopathy, leading to pulmonary arterial hypertension[3,4]. Stroke is a common complication and occurs in 10–20% of patients[8]. Since the stroke is infrequent as the first manifestation and only a few cases have been reported[9], we present a case of a young Syrian female with a large right cerebral infarction as the initial manifestation of Takayasu disease.

HIGHLIGHTS

- Takayasu Arteritis (TA) is a rare chronic inflammatory disease affects large vessels.
- TA etiology still unknown.
- The disease typically presents with diverse symptoms, depending on the site and degree of arterial lesions.
- Diagnosis is based on clinical features and angiographic findings, and the American College of Rheumatology criteria 1990 is commonly used for the diagnosis of TA.

Case presentation

Clinical history and investigations

A 39-year-old married woman arrived at our emergency department after 4 h of a sudden onset of left hemiplegia and severe dysarthria, the National Institutes of Health Stroke Scale (NIHSS) for the patient was 13, which indicates a moderate stroke. Her medical and family history were unremarkable, had not complained of any previous constitutional, neurological, or vascular symptoms, and she did not consume alcohol or

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cigarettes. The physical examination showed an absent left radial pulse, while other peripheral pulses were palpable, carotid bruits could be heard in the neck, and neurological assessment revealed severe dysarthria, left hemiplegia, and muscle weakness (0/5 on the left upper extremity, 1/5 on the left lower extremity). Blood pressure (BP) was 80/50 mmHg in the left arm and 140/50 mmHg in the right arm.

Laboratory tests and radiological investigations

Laboratory results indicated an elevated ESR (31 mm/h) in the first hour (normal range up to 15 mm/h) and CRP (11 mg/l) (normal range up to 5 mg/l). Other laboratory tests were within normal ranges. ECG was unremarkable, and echocardiography revealed mild aortic regurgitation with an Ejection Fraction over 60%. Computed tomography of the brain revealed a right large cerebral infarction (Fig. 1).

Ultrasound of both common carotid arteries demonstrated diffuse annular wall thickening, suggestive for inflammatory changes. Multislice computed tomography angiography revealed the following (Fig. 2):

- Thickening of the aorta and common carotid arteries.
- Lumen stenosis of the left internal carotid artery.
- Occlusion of the right internal carotid artery.
- Annular wall thickening and lumen stenosis of the left subclavian artery.
- Segmental stenosis followed by dilatation of the right subclavian artery.
- Stenosis of the right middle cerebral artery.

Diagnostic strategy

The patient, a 39-year-old woman (under the age of 40), presented with no prior complaints, or medical or family history of any thrombotic diseases. Upon examination, the patient had an absent left-sided pulse, and a 70 mmHg interarm systolic blood pressure difference (greater than 10 mmHg). These findings raised suspicion for TA, which was confirmed by the findings on multislice computed tomography angiography; she was evaluated based on the American College of Rheumatology criteria and met 5 out of 6 criteria (Table 1).

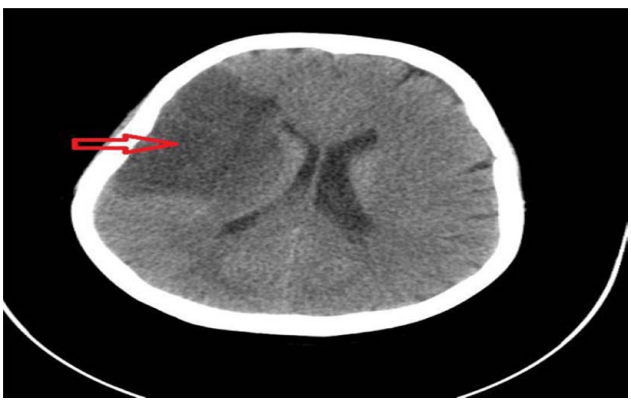

Figure 1. Computed tomography of the brain with right massive cerebral infarction.

The vascular surgery consultation determined that the extent and severity of arterial involvement warranted consideration for surgical intervention after the resolution of the acute phase.

Following diagnosis, literature review, and American College of Rheumatology recommendations; the patient was treated with prednisolone 40 mg/days, methotrexate 10 mg weekly, plus aspirin and physiotherapy. It is worth mentioning that the use of thrombolysis is essential in the management of this patient's case. However, thrombolysis is unfortunately unavailable at our hospital. The patient was discharged from the hospital a week after admission, with regular follow-ups to assess response and adjust the therapeutic plan as needed.

Follow-up the treatment

Re-evaluation after 2 months showed that the patient has improved to this therapy, with no dysarthria, muscle strength increased up to 4/5 in the left lower extremity, and she has regained her ability to walk. Muscle strength increased to 2/5 in the left upper extremity. (ESR) and (CRP) returned to the normal range (Table 2).

Given the patient's positive response to medical management, including prednisolone, methotrexate, and aspirin, coupled with patient refusal of surgery, a conservative approach was considered without the surgery need.

Discussion

TA, also known as pulseless disease, and occlusive thromboaortopathy[3,6], is a rare chronic inflammatory disease that affects all layers of the artery and leads to stenosis and occlusion of its lumen; causing wall thickening, fibrosis, stenosis, thrombus formation, dilatation, or aneurysms of the vessels[1–4], affecting women more than men, typically presenting between 10 and 40 years[6,10]. There are differences in the types of affected arteries between races and countries, leading to different clinical manifestations[5,11]. The most common initial manifestations in Japan are weak or absent pulse, claudication, and aortic regurgitation due to involvement of the aorta and its branches, while hypertension is the most common in India due to renal artery involvement[11]. Its presentation is characterized by two stages, with an early, subacute, 'prepulseless' stage presenting as nonspecific constitutional symptoms, such as low-grade fever, fatigability, weight loss, arthralgia, and myalgia, followed by a late, chronic, pulseless stage presenting with the symptoms of end-organ ischemia due to stenosis of the vessels. However, this classic presentation does not hold true for all patients[4,9,12]. In the chronic stage, clinical presentation depends on the arteries involved. The most common presentation is diminished or absent pulses, associated with limb claudication and blood pressure discrepancies. Other manifestations include vascular bruits, particularly affecting the carotids, subclavian, and abdominal vessels, hypertension resulting from renal artery stenosis, Takayasu's retinopathy, aortic regurgitation resulting from dilatation of the ascending aorta, congestive heart failure due to hypertension, aortic regurgitation and coronary artery disease, neurological features secondary to hypertension and/or ischemia, including seizures, stroke, amaurosis, and pulmonary vasculopathy, leading to pulmonary arterial

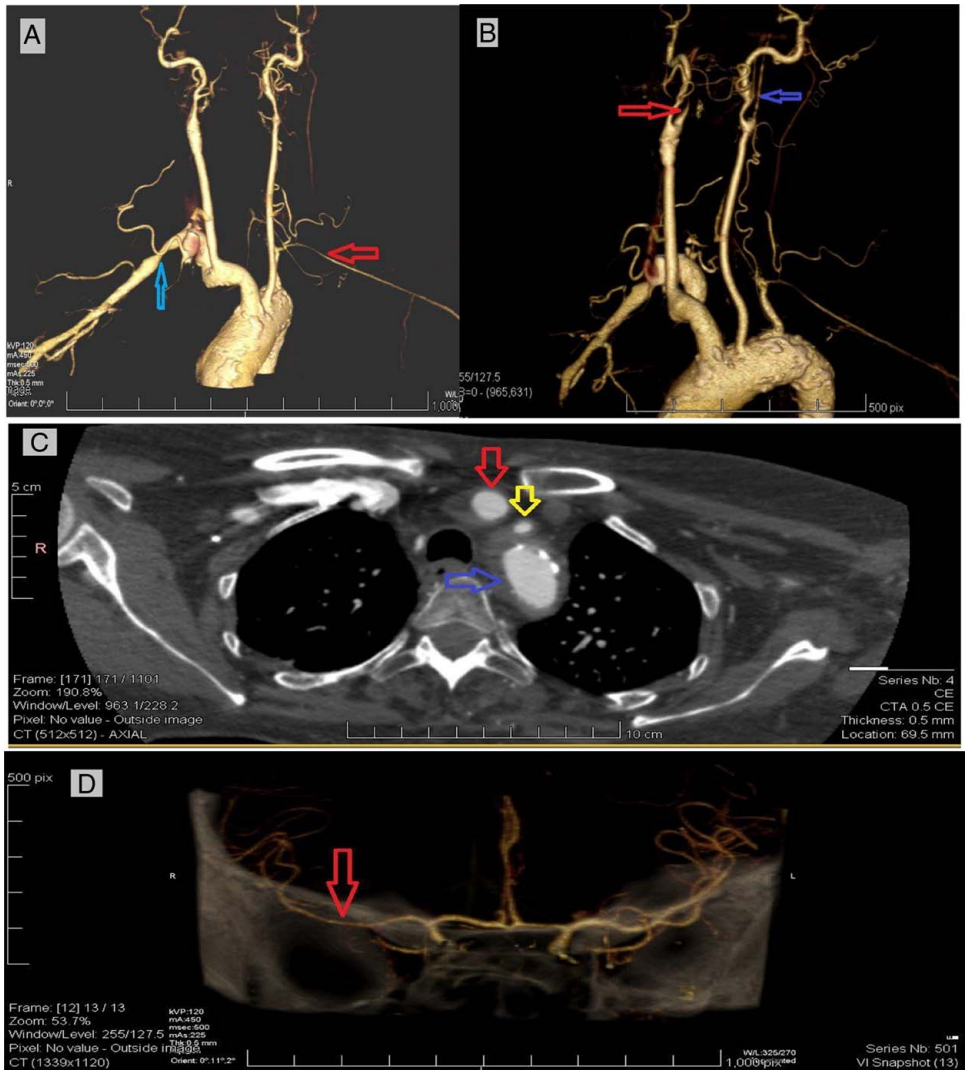

Figure 2. Multislice computed tomography angiography of the infected arteries. A. red arrow; lumen stenosis of the left subclavian artery, blue arrow; segmental stenosis followed by dilatation of the right subclavian artery. B. red arrow; lumen stenosis, and occlusion of the right internal carotid artery, blue arrow; lumen stenosis of the left internal carotid artery. C. blue arrow; walls thickening of common carotid artery, yellow arrow; wall thickening of the brachiocephalic trunk. D. red arrow; stenosis of right middle cerebral artery.

Table 1

1990 criteria for the classification of Takayasu Arteritis.

Criteria	Findings in the patient
Age at disease onset $\leq$ 40 years	39 years old
Claudication of extremities	No complaint
Decreased brachial artery pulse	Absent left radial pulse
BP difference $\geq$ 10 mmHg	70 mmHg interarm systolic BP difference
Bruit over subclavian arteries or aorta	Carotid bruits could be heard in the neck
Arteriogram abnormality	Thickening of the aorta and common carotid arteries.
	Lumen stenosis of the left internal carotid artery.
	Occlusion of the right internal carotid artery.
	Annular wall thickening and lumen stenosis of the left subclavian artery.
	Segmental stenosis followed by dilatation of the right subclavian artery.
	Stenosis of the right middle cerebral artery (MCA)

hypertension[3,4]. Whereas, stroke can occur due to acute ischemia and infarction by thrombosis or, more commonly, embolism of vessels, which has been reported in 10–20% of the patients, out of which 80% had anterior circulation involvement[4,8,13]. However, stroke as initial manifestation is infrequent and reported in 5–8% of patients only, most of them are from East and South Asia and the United States of America (USA)[1,4,7,14]. Thus, the presentation of our patient with large right cerebral infarction as the first manifestation of TA is rare, especially in the Middle East, so reporting our case may increase awareness about this disabling complication of TA, contribute to the limited literature on TA in Arab populations. Our patient met five criteria of the 1990 American College of Rheumatology criteria for the diagnosis of TA[10]. The treatment plan was based on the 2021 American College of Rheumatology recommendations for the treatment of TA, and its goal is to treat the disease and prevent recurrence or critical complications[15]. Despite the involvement of

Table 2

Patient follow-up after medical intervention and physiotherapy.

	Before	After	Notes
ESR	31 mm/h	5 mm/h	Normal range up to 15 mm/h
CRP	11 mg/l	0.3 mg/l	Normal range up to 5 mg/L
Dysarthria	Severe dysarthria	No dysarthria	—
Muscle strength in the left upper extremity	0/5	2/5	—
Muscle strength in the left lower extremity	1/5	4/5	—

the aorta and most of its branches in our patient, the patient improved clinically, with no recurrence occurred in any organ during the 6 weeks following discharge.

Conclusion

TA presents a diagnostic challenge due to its diverse manifestations and geographical variations, but a proper physical examination with good experience may help. This case highlights the importance of considering this rare vasculitis, particularly when confronted with atypical presentations like an initial stroke. Early diagnosis, continuous medical management, along with timely interventions, will halt the inflammation and its progression to the permanent stenotic stage, preventing end-organ damage, and thereby, leading to better clinical outcomes. Further studies are required to improve our comprehension and enhance patient outcomes.

Ethical approval

No ethical approval was needed.

Consent

Written informed consent was obtained from the patient for publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

T.M.: provided medical treatment, wrote and supervise the scientific and academic aspects of the manuscript, and revised it; Z.H.: reviewed the literature, wrote and revised the manuscript, and made grammar and spelling language editing; M.E.: provided medical treatment to the patient; B.A.: provided medical treatment to the patient; J.H.: wrote and revised the manuscript.

Conflicts of interest disclosure

There were no conflicts of interest.

Research registration unique identifying number (UIN)

No registration was needed.

Guarantor

Tareq Muhammad and Zulfiqar Hamdan.

Data availability statement

All data are available.

Provenance and peer review

Not commissioned, externally peer-reviewed.

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