Received: 29 November 2022 | Revised: 3 March 2023 | Accepted: 5 May 2023

DOI: 10.1002/ccr3.7412

CASE REPORT

Clinical Case Reports Open Access WILEY

Ischemic stroke at first presentation of Takayasu arteritis in a young African male from Kenya, East Africa: Case report and brief literature review

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Key Clinical Message

This case highlights the need for thorough clinical examination to rule out Takayasu arteritis (TA) as a cause of stroke in a young asymptomatic East-African male. Available clinical management guidelines should guide management of TA patients.

Abstract

We present a case of a young, previously asymptomatic East-African Black male presenting with large territory ischemic infarct at first diagnosis of TA. To our knowledge, this is the first published report of a male patient in East Africa with a stroke as the first presentation of TA.

KEYWORDS

cerebrovascular accident, stroke, Takayasu arteritis, vasculitis

1 | INTRODUCTION

Cerebrovascular accidents are quite devastating to those affected and even more so for otherwise young, healthy individuals. It is therefore paramount to appropriately evaluate for possible causes, identify the possible etiology, and put in place strategies to prevent recurrence. These etiologies include cardio embolic disease, hematological conditions, infections of the nervous system, and systemic inflammatory pathologies including vasculitis such as Takayasu arteritis (TA).[1]

The prevalence of cerebrovascular events in the course of TA has been estimated to be between 15% and 20% in the course of the illness with occurrence of ischemic stroke at point of first diagnosis being between 6% and 8% as is seen in our clinical case.[2–4]

Here, we describe a case of a young African male presenting with a catastrophic cerebrovascular event at first presentation. We further give a synopsis of pathogenesis, diagnosis, and management of TA relevant to clinical practice.

2 | CASE REPORT

We present a case of a 25-year-old Black African male who presented to our hospital with a 2-day history of sudden onset right sided weakness and inability to talk.

Informed written consent was sought and obtained from patient's sister and is available upon request from the journal.

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He denied any history of headache, fever, prior trauma, or chest pain. Of note, he had no history of chronic illness, intravenous drug use, cigarette smoking, alcohol use or any personal or family history of hypertension, diabetes or cardiovascular disease. Our patient had been working as a commercial motorcycle rider. General examination revealed a young man who was awake and responsive, not in any respiratory distress with no conjunctival pallor. He had a blood pressure reading of 130/79 mmHg on his right arm and 80/47 mmHg on the left arm revealing an obvious discrepancy between the two arms. Our patient had motor aphasia with right sided cranial nerve 7, 9, 10, 11, and 12 palsy. Motor power of 0/5 on the right upper and lower limb on all muscle groups was noted with normal power on the left. Additionally, we noted hyperreflexia and increased tone on the right limbs with normal global sensation. No cerebellar signs were present. On cardiovascular examination our patient had absent pulses on the left arm, an audible carotid bruit on the left with normal heart sounds without any murmur. A distended bladder with urine retention was present on abdominal examination. Further systemic examination was non-revealing. Working with a diagnosis of a cerebrovascular accident likely due to a large vessel vasculitis, a CT scan of the head was ordered and showed a left sided fronto-temporal hypo density consistent with an ischemic infarct (see Figure 1). Carotid Doppler ultrasound revealed bilateral carotid artery stenosis with 50% occlusion on the right and complete occlusion on the

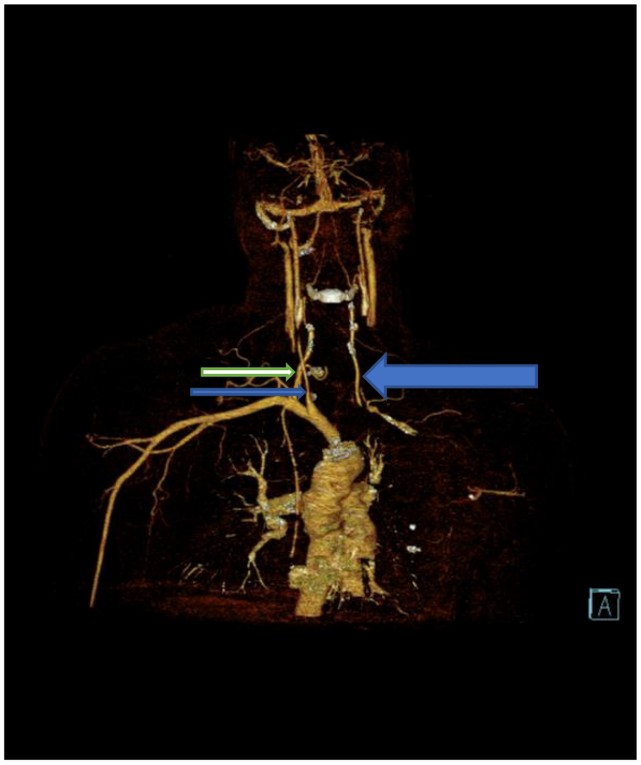

FIGURE 2 CT angiogram reconstruction image shows reduced caliber of the right carotid artery (small blue arrow) as it branches of the brachiocephalic artery as compared to the right vertebral artery branching off the right subclavian artery (white arrow). The large blue arrow shows a fully occluded left carotid artery and subclavian artery with no visible blood flow on angiography with only the left vertebral artery present.

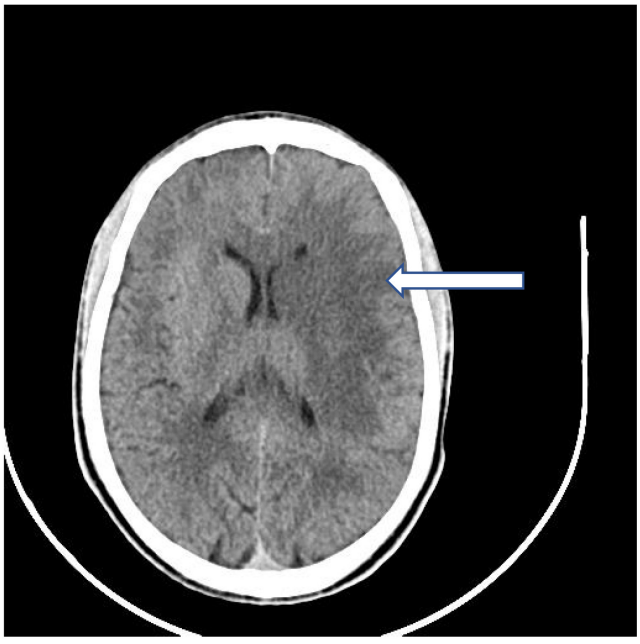

FIGURE 1 Non-contrast CT scan of the head showing a left fronto-temporal hypodensity consistent with an ischemic infarct (white arrow).

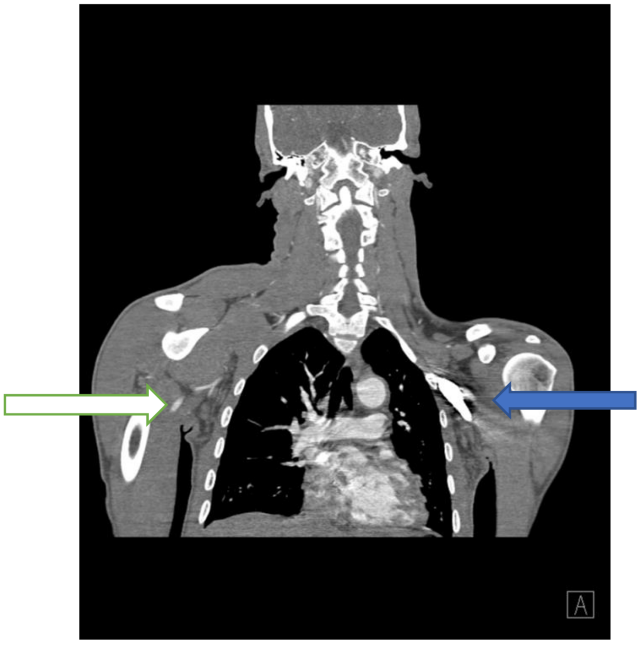

FIGURE 3 Reconstructed CT angiogram image shows visible right axillary artery (white arrow) with absent (occluded) left axillary artery (blue arrow).

left. Further, a CT angiogram showed left subclavian artery occlusion and bilateral carotid artery stenosis with complete occlusion on the left (see Figures 2 and 3). A 2D transthoracic echocardiogram and ECG were normal. His screen for syphilis with VDRL was negative. A CSF GeneXpert and BioFire® meningo-encephalitis panel were also negative. Of note, ESR and CRP were high with values of 85 mm/h. and 30.4 mg/L, respectively. Additionally, lipid profile results returned normal with a negative HIV test by ELISA and antinuclear antibody test. His full hemogram and kidney function tests were normal with a hemoglobin level of 15.4 g/dL. Based on the above clinical findings and tests, a diagnosis of TA was made. Management was initiated with aspirin 75 mg, and our patient pulsed with high dose methyl prednisolone at 1 gm once daily for 3 days. Thereafter a maintenance dose of azathioprine 100 mg twice daily, deflazacort 6 mg twice daily, and physiotherapy was initiated. At 3 months post discharge, he is doing well and has a power of 3/5 on the right lower limb, is able to talk with a slurred speech with no other organ involvement noted.

3 | DISCUSSION

TA, a chronic disorder characterized by inflammation of the aorta or its branches, is also referred to as pulseless disease.[3] It is commonly reported among young female patients of Asian origin. However, a more diverse global occurrence has been described in recent decades mostly among individuals under the age of 40 years.[4] The pathogenic mechanisms behind TA are not fully known. Large and medium sized arteries are considered immune-privileged sites. However, in TA, abnormal activation and infiltration of macrophages and lymphoid cells (CD4 and CD8 T cell) are thought to lead to unchecked granulomatous inflammation of large vessel walls such as the aorta and its branches. Additional postulations suggest activation of matrix metalloproteinase contributes to fibrosis and transmural thickening of large vessels with subsequent stenosis or dilatation.[5] When this occurs in vessels supplying the brain such as the internal carotid artery, it can result in hypo perfusion leading to ischemic cerebrovascular accidents. Elsewhere in the body, hepatic release of acute phase reactants is triggered leading to elevated C-reactive protein.[5]

Clinical presentation is usually non-specific underscoring the need for a high index of suspicion to clinch the correct diagnosis. The highly variable symptoms include fever, malaise, headaches, limb claudication, and hypertension. A classical triphasic pattern of presentation characterized by an initial period of constitutional symptoms such as fever and night sweats in phase I, pain over arteries in phase II and finally a fibrotic phase leading to ischemic symptoms due to critical stenosis of large arteries has been described.[6] However, literature seems to suggest that this picture is hardly seen in routine clinical practice or in recent study reports.[6,7] Due to extensive vascular involvement, TA patients can also present with 20 mmHg systolic blood pressure measurement discrepancy between arms with impalpable pulses on affected limb.[3] Thus, a through history and clinical examination remain a crucial tool in teasing out TA as a possible diagnosis among patients in the at-risk age group.

Clinical and radiological findings are considered the standard for routine diagnosis. As such, several diagnostic criteria have been described.[8,9] One of the most widely used is the American College of Rheumatology (ACR) criteria which has six criteria (see Table 1). Based on the ACR criteria, a diagnosis can be made with a sensitivity of 90.5% and specificity of 97.8% when a patient meets at least three of the six criteria.[8] Our patient met four of the six criteria. More recently in 2022, the ACR and the European League Against Rheumatism (EULAR) endorsed a common classification criterion in the evaluation of patients with a diagnosis of TA[10] (see Table 2).

Using the new criteria, our patient also met the threshold for classification as TA with a total score of 9 points. The diagnostic cutoff is 5 points.[10] Our case highlights an unfortunate occurrence where the patient had already developed a cerebrovascular event at diagnosis, seen in up to 6%–8% of patients at presentation and up to 20% in the course of the illness.[2,11]

TABLE 1 The 1990 American College of Rheumatology criteria for classification of Takayasu arteritis (TA).[8]

1.	Age of 40 years or younger at disease onset
2.	Claudication of the extremities
3.	Decreased pulsation of one or both brachial arteries
4.	Difference of at least 10 mmHg in systolic blood pressure between arms
5.	Bruit over one or both subclavian arteries or the abdominal aorta
6.	Arteriographic narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the upper or lower extremities that is not due to arteriosclerosis, fibromuscular dysplasia, or other causes

Note: A diagnosis of TA can be made with a sensitivity of 90.5% and specificity of 97.8% when a patient meets at least three of the six criteria.[8]

TABLE 2 2022 American College of Rheumatology/European League Against Rheumatism classification criteria for Takayasu arteritis (TA).[10]

Absolute requirements	
Age of less than 60 years at time of diagnosis	
Evidence of vasculitis on imaging	
Additional clinical criteria	
Female sex	+1
Angina or ischemic cardiac pain	+2
Arm or leg claudication	+2
Vascular bruit	+2
Reduced pulse in upper extremity	+2
Carotid artery abnormality	+2
Systolic blood pressure difference in arms of greater than or equal to 20 mmHg	+1
Additional imaging criteria	
Number of affected arterial territories (select one)	
One arterial territory	+1
Two arterial territories	+2
Three or more arterial territories	+3
Systemic involvement of paired arteries	+1
Abdominal aorta involvement with renal or mesenteric involvement	+3

Note: Sum the scores for 10 items, if present. A score of 5 or more points is needed for the classification of TA.[10]

Two major rheumatology organizations namely EULAR and ACR have published guidelines on management of TA. However, the quality of evidence available to support these recommendations has remained low.[12,13] Treatment is multidisciplinary with both medical and surgical interventions needed. Systemic glucocorticoids form the backbone of medical treatment of TA patients. Additionally, disease modifying anti-rheumatic drugs such as azathioprine and methotrexate are concomitantly used to allow for tapering of steroids and minimize glucocorticoid associated side effects. Lastly, biologic agents such as tocilizumab and anti–tumor necrosis factor inhibitors are recommended in disease relapse. Surgery on the contrary is usually indicated in critical vessel stenosis or to repair arterial aneurysms.

Even with significant advances in the diagnosis and treatment of TA, the rate of complications prevails with reported rates as high as 50%. Some of these complications include ischemic cerebrovascular accidents as experienced by our patient, aortic regurgitation with associated heart failure, and end-stage renal disease. As expected, patients with more extensive disease at the time of diagnosis have higher complication rates.[14,15] Similarly, patients with a more progressive course, that is, with few or no event-free periods also experience higher rates of complications.[16] Recent studies have found that the male sex, presence of ongoing inflammation with elevated CRP levels, and those with carotidynia are more likely to experience relapses.[17,18]

4 | CONCLUSION

Although TA remains an uncommon diagnosis, it is important to consider it in the differential diagnosis of young African patients presenting with cerebrovascular accidents. Early diagnosis and appropriate management will be essential in reducing complication rates and improving outcomes.

AUTHOR CONTRIBUTIONS

Christopher Owino: Conceptualization; writing – original draft; writing – review and editing. **Betty Sirera:** Conceptualization; writing – original draft; writing – review and editing. **Felix Tarus:** Visualization. **Beryl Ganda:** Supervision. **Chrispine Oduor:** Supervision; writing – review and editing. **Abraham Siika:** Supervision; writing – review and editing.

FUNDING INFORMATION

There was no funding or financial assistance for this manuscript.

CONFLICT OF INTEREST STATEMENT

The authors declare that they have no conflicts of interest.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

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How to cite this article: Owino C, Sirera B, Tarus F, Ganda B, Oduor C, Siika A. Ischemic stroke at first presentation of Takayasu arteritis in a young African male from Kenya, East Africa: Case report and brief literature review. *Clin Case Rep*. 2023;11:e7412. doi:10.1002/ccr3.7412