

Takayasu Arteritis Mistaken for Epilepsy: A Case Presenting With Convulsive Syncope

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Abstract

Takayasu arteritis (TA) is a chronic inflammatory disease characterized by granulomatous vasculitis that predominantly manifests as aortoarteritis. This occlusive thromboangiopathy lacks pathognomonic features often resulting in a diagnostic dilemma leading to its under-recognition, misdiagnosis and delayed management. Although neurological manifestations are not uncommon in TA, convulsive syncope as an initial clinical presentation is extremely rare. We report a case of convulsive syncope as a manifesting symptom of TA. A 17-year-old male patient of African origin was referred to us from an upcountry regional hospital with a diagnosis of medically intractable epilepsy for cardiovascular review. He presented with a 28-week history of generalized tonic-clonic seizures followed by loss of consciousness. He denied history of recurrent headaches, fever, visual disturbances, arthralgias, claudication or unintentional weight loss. Physical examination revealed feeble left-sided brachial and radial pulses, elevated blood pressure, differences in blood pressure between arms and left-sided carotid and vertebral bruits. Computed tomography angiogram of his thoracic and abdominal aorta revealed changes suggestive of a diffuse arteritis. Additionally, magnetic resonance angiogram of the brain revealed total occlusion of the left common carotid, left internal carotid, left external carotid and left vertebral arteries. Based on the physical examination and radiological findings, we reached a diagnosis of TA. He was prescribed dexamethasone, methotrexate, acetylsalicylic acid and amlodipine. He had a remarkable recovery and was seizure-free for the last 5 months after discharge. TA may manifest with convulsive syncope mimicking epilepsy. Despite its rarity, presentations of this nature continue to challenge clinicians resulting in

delayed diagnosis with irreversible life-threatening consequences to patients. In view of this, physicians should strive to take detailed history and perform thorough physical examination so as to timely pick the characteristic signs of TA especially in patients presenting with unanticipated symptoms.

Keywords: Takayasu arteritis; Occlusive thromboangiopathy; Granulomatous arteritis; Chronic granulomatous vasculitis; Large vessel vasculitis; Pulseless disease; Convulsive syncope; Takayasu mimicking epilepsy

Introduction

Takayasu arteritis (TA) is a chronic inflammatory disease characterized by granulomatous vasculitis that predominantly manifests as aortoarteritis [1-4]. This occlusive thromboangiopathy has an estimated incidence of two cases/1,000,000 population/year and is more prevalent in Africa, Southeast Asia, and Central and South America [1-4]. The etiology of TA remains poorly understood and likewise its exact pathogenic sequence is yet to be established [1-4]. The disease demonstrates a striking female predilection and commonly but not exclusively presents in the second or third decade of life [1, 3]. Owing to the paucity of distinctive symptoms and absence of a reliable serological marker of disease course, TA often poses a diagnostic dilemma leading to its under-recognition, misdiagnosis and delayed management [1-3].

The hallmark of TA is the granulomatous inflammation that commences from the adventitia and progresses to the intima with resultant segmental stenosis, occlusion, dilatation and aneurysm formation leading to end-organ ischemia [5]. The characteristic proximal arterial involvement usually leaves the brain parenchyma until later stage of disease progression [5]. Takayasu-induced cerebral ischemia may result in a myriad of neurological manifestations including headache, organic confusion, dementia, seizure, meningitis, encephalitis, stroke, visual disturbances and syncope [4-6]. Although between 42% and 80% of patients with TA present with central nervous system (CNS) manifestations, convulsive syncope as the primary presentation is rare [5]. We report a case of convulsive syncope as an initial presentation of TA in a 17-year-old male patient of African origin.

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Figure 1. Computed tomography angiogram of thoracic and abdominal aorta displaying (1) left common carotid artery, (2) origin of left subclavian artery and (3) features suggestive of diffuse arteritis, i.e. irregular narrowing with variable degrees of stenosis, tapering and corrugated appearance.

Case Report

A 17-year-old male patient secondary student of African origin was referred to us from an upcountry regional hospital with a diagnosis of medically intractable epilepsy for cardiovascular review. He presented with a 28-week history of recurrent convulsive syncope attacks which were unresponsive to anti-epileptic drugs. The seizures were generalized tonic-clonic in nature followed by loss of consciousness of variable duration (10 - 30 min). These episodes were often associated with abrupt change of posture particularly sitting upright and on standing. On average he experienced 1 - 3 episodes daily. He denied bladder/bowel incontinence or presence of aura symptoms following or preceding the convulsive episodes respectively. Neurologist's notes revealed post-ictal drowsiness with a transient memory impairment following the attack. There was a negative history of recurrent headaches, fever, visual disturbances, arthralgias, claudication or unintentional weight loss. His past medical history was unremarkable and he denied substance abuse.

On examination, he was alert, oriented and well-kempt. He was pale (conjunctival and palmar) but not jaundiced. Oral examination revealed a lateral tongue bite mark with adequate dentition. His left-sided brachial and radial arterial pulses were feeble, whereas pulses on his right side and in both lower limbs were normal. Left-sided carotid and vertebral bruits were not-



Figure 2. Brain magnetic resonance angiogram showing total occlusion of the left common carotid, left internal carotid, left external carotid and left vertebral arteries.

ed. His blood pressure (BP) was 169/91 mm Hg on his left and 124/82 mm Hg on the right side. He had a pulse rate of 119 beats/min which was regular and of good volume. He was afebrile (37.2 °C) and had a healthy weight (body mass index (BMI) 21.9 kg/m²). Abdominal examination was uneventful with no evidence of renal bruits or aneurysms. The rest of systemic examination was unremarkable.

His hematological, biochemical, serological and autoimmune tests revealed normal findings except for raised inflammatory markers, i.e. erythrocyte sedimentation rate (ESR) 58 mm/h and C-reactive protein (CRP) 27.6 mg/L. A tuberculin test was negative. Echocardiographic examination revealed normal findings except for a trace aortic regurgitation. Due to a clinical suspicion of TA, we ordered a carotid Doppler ultrasound scan (USS) which showed a complete occlusion of the left common, internal and external carotid arteries. On the other hand, renal Doppler USS revealed bilaterally patent vessels with a spontaneous phasic flow. Moreover, both common and deep femoral arteries exhibited a normal course and calibre with no evidence of intraluminal plaque or stenosis on femoral Doppler USS.

Computed tomography (CT) of brain was negative for stroke or space occupying lesions and electroencephalogram (EEG) ruled out likelihood of a focal cerebral pathology. Furthermore, CT angiogram of his thoracic and abdominal aorta revealed changes suggestive of a diffuse arteritis, i.e. irregular narrowing with variable degrees of stenosis, tapering and corrugated appearance (Fig. 1). Additionally, left common carotid, left internal carotid, left external carotid and left vertebral arteries could not be visualized on the magnetic resonance angiogram (MRA) of the brain (Fig. 2).

Based on the clinical presentation and radiological features, our case fulfilled the American College of Rheumatology criteria [7] for TA. He was counseled and dexamethasone (60 mg per day), methotrexate (10 mg per day), acetylsalicylic acid (75 mg once daily) and amlodipine (10 mg once daily) were initiated on the sixth day of hospitalization. The patient had a total of 14 convulsive syncope episodes in the initial 6 days of hospitalization (i.e. before initiation of steroids) and five episodes in the last 7 days. He was discharged home after 13 days of hospitalization in a considerably stable state. We reviewed him as an out-patient at 3 and 6 months. He had a controlled BP (132/78 mm Hg left-sided and 117/71 mm Hg right-sided) and was seizure-free for the last 5 months (i.e. had two episodes within the first month after discharge).

Discussion

TA is a rare yet the most common large-vessel vasculitis with an unidentifiable cause and undetermined pathogenesis [1-4]. Owing to the involved vascular territory and the degree of disease progression, TA patients vary greatly in their clinical presentation which is collectively characterized by active and quiescent phases [1-3]. Such unpredictability in its presentation and course often makes the diagnosis of this potentially life-threatening condition difficult and late [1-4].

In spite of their relatively late onset, neurological manifestations are not uncommon in TA [4]. Decreased blood flow resulting from steno-occlusive lesions and/or shifting of the blood flow (steal) is responsible for various TA-associated CNS manifestations [4]. Whereas convulsive syncope as an initial clinical presentation is extremely uncommon [8-12], several other neurological manifestations including headache, vertigo, organic confusion, dementia, meningitis, encephalitis, stroke, visual disturbances and intracranial aneurysms have been commonly associated with TA [4, 6].

As presented in this case, the young boy presented with convulsive syncope which was labelled epilepsy and he was prescribed several anti-seizure drugs with no relief. Due to the absence of other clinical manifestations, it took literally 7 months to reach the definitive diagnosis of TA. Despite all the existing challenges attributable to TA diagnosis, we believe that this case is a clear reminder of how fundamental thorough history taking and physical examination are. We are convinced that despite the rare symptomatology, a couple of TA signs including elevated BP, BP discrepancy between arms, unequal pulses and bruits along the neck region were present but missed during the 7 months course of this case illness.

Worth noting is that, a number of medical conditions may present with generalized tonic-clonic seizures, therefore, not every seizure is epilepsy. Unfortunately, we are living in a world where clinicians have lost clinical examination capabilities and rely almost entirely on laboratory and radiological tests to reach a diagnosis [13-15]. As a result, this form of substitution in clinical discipline has led to increased incorrect diagnoses and a more pricey management approach [15]. Nonetheless, it is imperative for clinicians to be aware of the unusual presentations of TA because a timely and appropriate

intervention has the potential to halt the inflammatory process and improve the health-related quality of life.

Despite its rarity, TA may manifest with convulsive syncope mimicking epilepsy. It is necessary for clinicians to understand the dynamic symptomatology of this condition for an early diagnosis and timely management to curb damage accrual and attributable morbidity. Moreover, physicians should strive to take detailed history and perform thorough physical examination so as to timely pick characteristic signs of TA especially in patients presenting with unanticipated symptoms.

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Financial Disclosure

None to declare.

Conflict of Interest

The authors declare that they have no conflict of interest.

Informed Consent

Written informed consent was obtained from the patient's next of kin for publication of this case report and any accompanying images.

Author Contributions

SB and PP took the history and performed the physical examination. FL interpreted the radiological investigations. JM, HJS, NRH, NM, ZM, SB, SAW and MJ participated in clinical management and counseling of the patient during hospitalization. PP wrote the initial draft of the manuscript. All authors reviewed and contributed to the final version of this case report.

Data Availability

The authors declare that data supporting the findings of this study are available within the article.

Abbreviations

BMI: body mass index; BP: blood pressure; CNS: central nervous system; CRP: C-reactive protein; CT: computed tomography; EEG: electroencephalogram; ESR: erythrocyte sedimentation rate; MRA: magnetic resonance angiogram; TA: Takayasu arteritis; USS: ultrasound scan

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