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Large Vessel Vasculitis in a Patient with Systemic Lupus Erythematosus Presenting as Takayasu's Pulseless Arteritis

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Abstract

Systemic lupus erythematosus (SLE) with an associated aortoarteritis presenting as an ischemic stroke is a rarity in the medical literature. We report the case of an 11-year-old male presenting with an acute ischemic stroke meeting the criteria for the diagnosis of SLE and findings consistent with an aortitis on imaging but mimicking the diagnosis of Takayasu's pulseless arteritis. Blood and imaging investigations revealed the finding of SLE aortitis following an acute stroke presentation. Thus, it is imperative to note that even though it is infrequent, SLE can be associated with a large vessel vasculitis.

Keywords

Systemic Lupus Erythematosus, Aortoarteritis, Takayasu's Pulseless Arteritis

1. Introduction

It is well recognized that the two common causes of large vessel arteritis are giant cell arteritis and Takayasu's pulseless arteritis [1]. A less well known, cause is SLE especially SLE affecting the aorta and the great vessels arising from the aorta [2]. It is not uncommon for an ischaemic stroke to be caused by SLE but the pathophysiology usually involves vasculitis affecting smaller vessels and or increased stickiness of the blood itself [3].

There is a myriad of etiologies for stroke presentation in the younger population. Strokes in the pediatric groups of patients are relatively rare in which the reported incidence of strokes (ischemic and hemorrhagic) in children under 18 years old was reported to be 1.2 to 13 cases per 100,000 patients [4].

Etiologies of strokes range from cardio embolic phenomena, atherosclerosis

and inflammatory diseases [5]. Vasculitis can lead to aneurysm formation or stenoses of the affected blood vessels which can result in limitation of blood flow to the brain resulting in stroke [6].

In this case report, the patient presented with an acute ischemic stroke in which the etiology was revealed as a lupus arteritis presentation with an associated large vessel vasculitis (aortitis); a rare occurrence as noted throughout the medical literature.

2. Case Report

This is the clinical case report of an 11 year old Afro-Caribbean boy who presented with sudden onset of left upper limb and left lower limb weakness over a period of 2 days. The patient had a background history of a normocytic anaemia which was previously being investigated. Prior to this, there was one hospital admission 2 years ago when the patient presented with arthralgia and chest pain for which no definitive diagnosis was made at the time.

There were also complaints of facial twisting, slurred speech, intermittent diplopia and generalized headaches associated with two isolated episodes of fever.

On physical examination on admission, the boy was noted to have a BP 114/66 mmHg, a tachycardia of 117 beats per minute and he was tachypneic with a temperature of 37.9 degrees Celsius. His oxygen saturation on room air was 97% and his random blood glucose was recorded as 113 mg/dL. There was a grade 2/6 ejection systolic murmur noted in the aortic region with no radiation. Auscultation of the chest and abdominal examination were both unremarkable.

On neurological examination, his Glasgow coma scale score was 15/15 and the patient was oriented to time, person and place. Visual field testing revealed a left incongruous homonymous hemianopia. Fundoscopy was normal. A left upper motor facial weakness was present. Bulbar function and speech were normal.

On limb examination, tone was markedly reduced in the left upper and lower limbs. The power of the left upper and lower limb was 0/5, whilst the power of the right upper and lower limb was 5/5. The patient was hyperreflexic throughout with left ankle clonus and a left up going plantar response. There were no abnormal cerebellar or sensory findings and the gait could not be assessed.

Both radial and the right posterior tibial pulses were palpably absent.

3. Results

CT brain without contrast revealed wedge shaped hypo densities in the right superior fronto-parietal region as shown below in **Figure 1**.

On contrast administration, there was no enhancement of the lesions.

MRI/Angiogram brain demonstrated a large right cerebral infarct involving the right middle cerebral artery territory mainly, plus an absent right internal carotid artery and attenuation of the right middle cerebral artery (**Figure 2** and **Figure 3**).

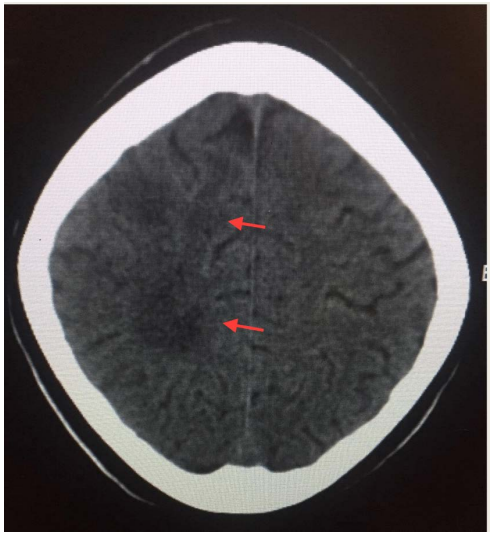

Figure 1. CT brain non contrast demonstrating hypo densities in right superior fronto-parietal region.

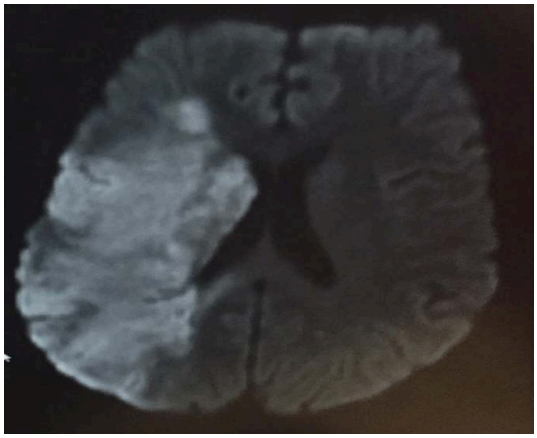

Figure 2. MRI Brain Scan showing large right cerebral infarct.

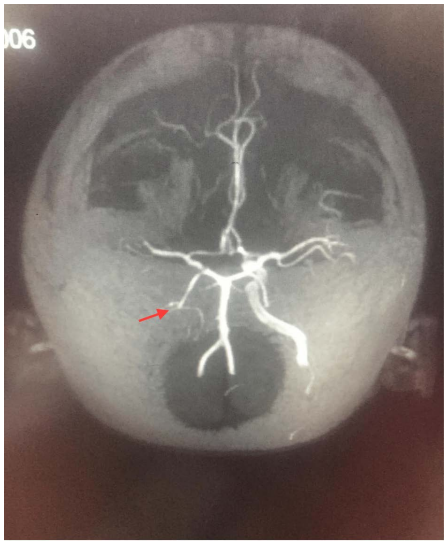

Figure 3. absent right internal carotid and attenuation of the right middle cerebral artery.

CT angiogram showed a filling defect in the proximal right common carotid and abnormalities in the aortic arch consistent with inflammatory disease (Figure 4 and Figure 5).

A normocytic normochromic anaemia was noted with a Hb 8.6 g/dL. There was no renal impairment with a Cr 1.0 and electrolytes all within normal limits. Liver function tests were within normal limits while the erythrocyte sedimentation rate (ESR) was >125 $mmhr^{-1}$ and C-reactive protein was elevated 242 mg/dL.

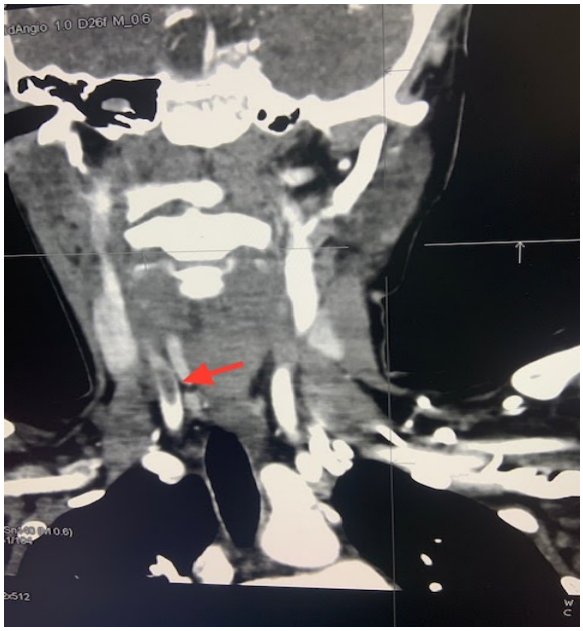

Figure 4. CT carotids showing a central filling defect in the proximal aspect of the right common carotid artery.

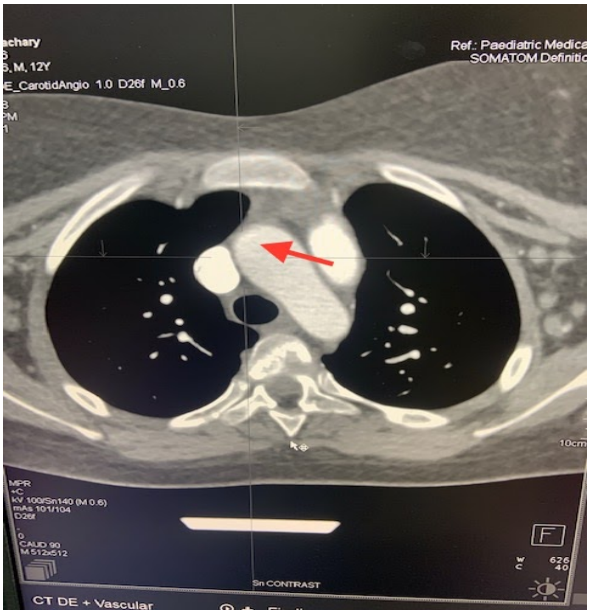

Figure 5. CT chest angiogram demonstrating hypo densities seen in intimal wall of the aortic arch.

Fasting lipid profiles were as follows Total Cholesterol 146 mg/dL, TG 119 mg/dL, HDL 21 mg/dL, LDL 102 mg/dL and VLDL 24 mg/dL. Coagulation studies and thyroid function were within normal limits. Other significant investigations were as follows as shown in Table 1.

ECG showed normal sinus rhythm and transthoracic echocardiogram done on 2 separate occasions demonstrated a normal left ventricle and valves without any evidence of thrombus, vegetation or shunts.

4. Management

Before the results of the autoimmune screen (ANA, dsDNA) were available, the initial thought was a diagnosis of Takayasu's pulseless arteritis.

The patient was treated with methylprednisolone 1 g iv od for 3 days followed by high dose prednisolone on alternating days. He became drowsy whilst his ESR remained > 125 $mmHr^{-1}$ following steroid administration.

Table 1. Blood investigations.

HIV Rapid Test	Negative
Hb Electrophoresis	A1 + A2
Sickle Cell Test	Negative
Urine Cocaine	Negative
Urine Marijuana	Negative
VDRL	Non reactive
Hb A1c	4.6%
HepBsAg	Negative
Anti-HCV	Negative
Rheumatoid Factor	Negative
Blood culture × 4	NBG
Urine Culture	NBG
Urinalysis	NAD
ANA	Positive
Anti-dsDNA	67.19 (Positive)
ANF	126.85 (Positive)
Homocysteine	6.3 (normal)
pANCA	32.76 (elevated)
cANCA	63.83 (elevated)
Anti-thrombin	Not done
Protein C	122 (normal)
Protein S	6.9 (low)
Factor V leiden	2.21 (normal)
Anti-cardiolipin antibodies	negative

Prednisolone was then switched to dexamethasone 4 mg iv tid. However, the boy looked distinctly unwell and his ESR still remained persistently elevated above 125 $mmHr^{-1}$.

There was a new complaint of visual blurring of the right eye. Fundoscopic examination revealed a pale retina and possibly early optic atrophy of the right eye (**Figure 6**).

Intravenous immunoglobulin (IVIG) (0.4 g/kg/day) was then administered for 5 days.

After a small number of days there was slow but steady improvement in the patient's overall condition. Daily physiotherapy and stroke rehabilitation were instituted after which the patient was discharged on oral prednisolone and followed in the outpatient clinic.

Two weeks later the boy appeared better. His peripheral pulses returned and the ESR started to trend downward. One month later, he was walking with help but left arm function was still very much impaired. He remains on a reducing dose of oral prednisolone.

5. Discussion

There is a paucity in the literature of reported cases demonstrating patients diagnosed with SLE having an associated large vessel vasculitis. The classic large vessel vasculitides include giant cell arteritis and Takayasu's arteritis [1]. In one case report, there was an associated large vessel vasculopathy in a Japanese patient with a background diagnosis of SLE and temporal arteritis [7]. We found a similar case to ours where there was a patient with a Takayasu's arteritis presentation followed by an associated diagnosis of SLE [8]. In this reported case, the patient was a 24 year old Indian female who initially presented with clinical symptoms and signs consistent with a diagnosis of Takayasu's pulseless arteritis. Three years later she then represented with rash, arthralgia, non scarring alopecia and other clinical features of SLE [8]. In our case, the patient similarly presented originally with clinical features more consistent with a diagnosis of Takayasu's pulseless arteritis but with a much more persistently elevated ESR. In

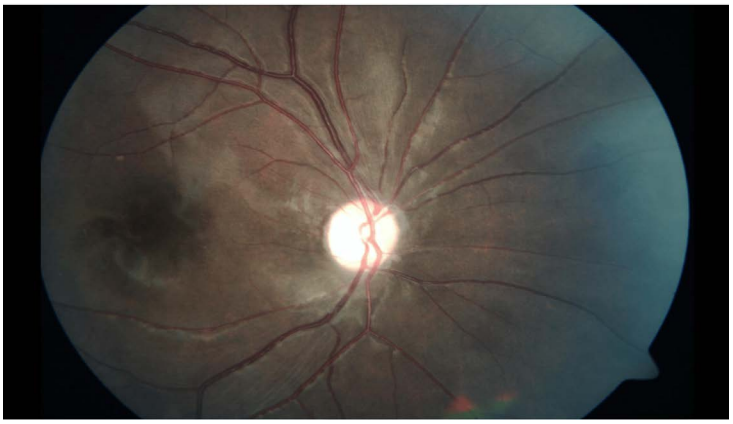

Figure 6. Retinal image demonstrating pale retinal background.

our patient, the diagnosis of SLE was made on the same admission and based on the previous complaints of arthralgia plus the positive autoimmune blood investigations.

Of note, there have been reported cases of non specific aortitis involving the aorta and its main branches with non specific symptomatology unfulfilling the diagnostic criteria for SLE [9]. Aortoarteritis is more prevalent in Japan and Afro-asian countries and has been noted to rarely be associated with SLE [10].

Another differential diagnosis worthy of mention includes a study by Wichremasinghe *et al.* [11]. They reported a small number of patients with emboligenic aortoarteritis where thrombi formation occurred from a transient form of focal aortoarteritis involving the medial elastic tissue of the aorta. This led to a potential etiology for an ischemic stroke in the younger population. None of their patients had a diagnosis of SLE. It is plausible that our patient could have had thromboembolism from inflammatory disease of the aorta and one of its main branches giving rise to the middle cerebral occlusion and possibly the right ophthalmic artery as well.

The boy presented with what appeared to be a good fit for the diagnosis of Takayasu's pulseless arteritis. The absent pulses, involvement of the aorta and great vessels and raised ESR were well in keeping with this diagnosis [12]. However, Takayasu's pulseless arteritis is seen primarily in Asian women and is not associated with lupus antibodies [8] [13]. Furthermore, there have not been any reported cases of Takayasu's pulseless arteritis in the Caribbean population plus SLE is a relatively common disorder in people of Afro-Caribbean descent [14] [15]. Our patient met the criteria for the diagnosis of SLE based on the EULAR/ACR scoring scale [16].

6. Conclusion

Even though the occurrence of SLE associated with a large vessel vasculitis is an unusual occurrence in the medical literature, the incidence is important to note as it can impact on the outcome of patients' morbidity and mortality.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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Abbreviations

SLE: Systemic lupus erythematosus

ESR: Erythrocyte sedimentation rate