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Case Report

Stroke as the initial presentation of Takayasu's arteritis: A case report ☆

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ARTICLE INFO

Article history:
Received 3 December 2019
Accepted 27 December 2019

Keywords:
Takayasu's arteritis
Stroke
Pediatric

ABSTRACT

In this study, we report the case of a pediatric neurology stroke patient who was ultimately diagnosed with Takayasu's Arteritis. Our case describes a 14-year-old Hispanic female with no significant past medical history who presented to an outside hospital for acute onset of confusion and right sided weakness. She was given tissue plasminogen activator (TPA) at the outside hospital and transferred as a stroke alert. Initial The NIH stroke scale (NIHSS) was 12 with primarily right sided symptoms. Physical exam was also significant for asymmetric pulses and blood pressures. Imaging was significant for multifocal stenosis. She was ultimately diagnosed with Takayasu's arteritis and treated with a multidisciplinary approach including pediatrics, neurology and rheumatology. This case represents an important differential diagnosis for pediatric stroke patients including those who have stroke as the presenting symptom of this systemic disease.

Background

Takayasu's arteritis (TA) is a chronic vasculitis that affects the aorta, its major branches, and the pulmonary artery [1,2]. It is most common in women in the second and third decades of life with highest prevalence in Asian countries such as Japan [3,4]. TA is rare in the US with an incidence of approximately 2.6 per million per year compared to 40 per million/year in Japan [5]. The pathogenesis of this disease remains unclear, but current studies suggest an immune-mediated inflammatory process that results in vessel wall fibrosis [6]. This ultimately leads to extensive vessel stenosis which is a major risk factor for strokes. TA has 2 general phases. The first is a systemic phase characterized by generalized symptoms including lethargy, weight loss, fever, myalgia, and arthralgia. The second phase is the "pulseless phase" characterized by vessel stenosis, hypertension, claudication, and neurologic manifestations including headache, dizziness, visual disturbances, transient ischemic attack (TIA), and stroke [7]. It has been estimated that 10%-20% of people with TA have an incidence of stroke [1,8,9]. However, stroke as a first manifestation of TA is rare [10].

Case report

A 14-year-old Hispanic female with no significant past medical history presented to an outside hospital after acute onset

☆ Competing Interests: None.

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https://doi.org/10.1016/j.radcr.2019.12.019

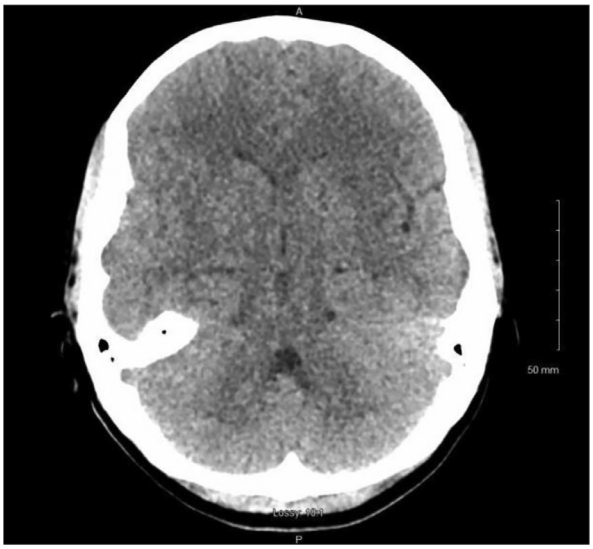

Fig. 1 – CTH showed early signs of left frontal stroke with loss of gray-white matter differentiation.

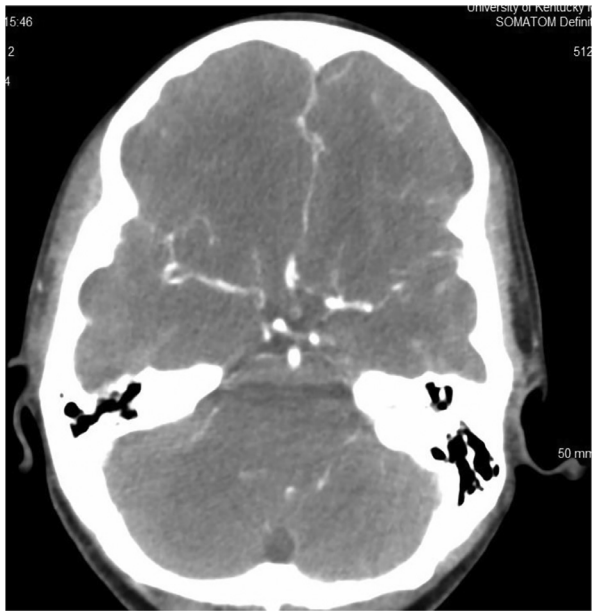

Fig. 2 – Initial CTA H showing distal left M1 stenosis.

of right-sided weakness, right facial droop and confusion at school. She was on no medications and had no known family history of cardiac, cerebrovascular, or rheumatologic disease. Initial NIH Stroke Scale was 12 and Glasgow Coma Scale was 14. CT head was negative for acute findings. MRI head was obtained emergently and showed a left frontal lobe stroke. The patient was within 4.5 hours of onset and had no contraindications to TPA. Therefore, TPA was initiated and she was transferred to our emergency department.

On arrival to our facility, repeat computed tomography head (CTH) showed early signs of left frontal ischemia (Fig. 1). With high suspicion for vascular abnormalities, CT angiogram of head and neck was obtained. computed tomography angiography (CTA) revealed severe stenosis of the brachiocephalic artery, origin of the right common carotid artery, and left common carotid artery which raised concern for a vasculopathy (Fig. 2). NIHSS was 13. She required frequent stimulation to arouse, was disoriented to month and age, obeyed 1 of 2 commands correctly, had minor right facial palsy, no effort against gravity in the right arm and leg, and mild dysarthria. Neurointerventional was consulted and decided against surgical intervention due to suspected Takayasu's with resulting friability of vessels. On general exam, she had discrepancies in blood pressure with measurement of 73/55 mm Hg in the right upper extremity, 117/60 mm Hg in the left upper extremity, 124/60 mm Hg in the right lower extremity, 141/55 mm Hg in the left lower extremity. Other vital signs were within normal limits. She had a right carotid bruit and a faint left carotid bruit. There were also differences in pulses with diminished pulses in the right radial and left dorsalis pedis compared to their counterparts.

With vascular abnormalities on imaging and discrepancies in pulses and blood pressures, rheumatologic workup was initiated. Erythrocyte sedimentation rate (ESR) was positive at 41 mm/hour (normal 0-20 mm/h). Neuroprotective strategies were implemented regarding blood pressure, volume status, electrolytes, and temperature.

Overnight, the patient had worsening in her neurologic exam with decreased responsiveness. CT perfusion showed core infarction involving the left basal ganglia, frontal lobe, and insula (Fig. 3). The following morning, her left pupil became fixed and dilated indicating increasing cerebral edema. Mannitol was given emergently. A repeat CT showed progression of acute infarct, a new midline shift, and partial uncal herniation (Fig. 4). Patient underwent a left hemicraniectomy and a repeat CT showed decompression and minimal brain herniation. Additional imaging was performed to evaluate for large vessel involvement and included: Magnetic resonance angiography (MRA) of head, neck, chest, abdomen, ultrasound of all 4 extremities and renal artery ultrasound (Fig. 5). Imaging revealed stenosis in bilateral common carotids, right vertebral artery, and right brachiocephalic artery. Aorta, renal vessels, and lower extremity vessels were intact and without significant stenosis.

Given the clinical exam, an elevated ESR and the findings on the MR angiogram, the presumptive diagnosis of TA was made. This patient met 3 criteria from 1990 American college of rheumatology (ACR) criteria which is 90.5% sensitive and 97.8% specific for the diagnosis of Takayasu Arteritis. These criteria included age at disease onset <40 years, systolic blood pressure difference >10 mm Hg between arms, and arteriogram abnormalities. Patient was started on high dose Prednisolone for 3 days and then continued on a lower dose. Cyclophosphamide was started on the seventh day of admission. In subsequent days, patient had improvement in alertness and movement of extremities with purposeful movements on the left side. Patient was discharged to an acute rehabilitation facility after 19 days of hospital stay. She was discharged on 60 mg prednisolone daily. Patient is following up with outpatient pediatric rheumatology, pediatric neurology, neurosurgery, and ophthalmology. She continues to work with occupational, physical, and speech therapy. She is also on 5 mg Prednisone daily and receives Cyclophosphamide infusions.

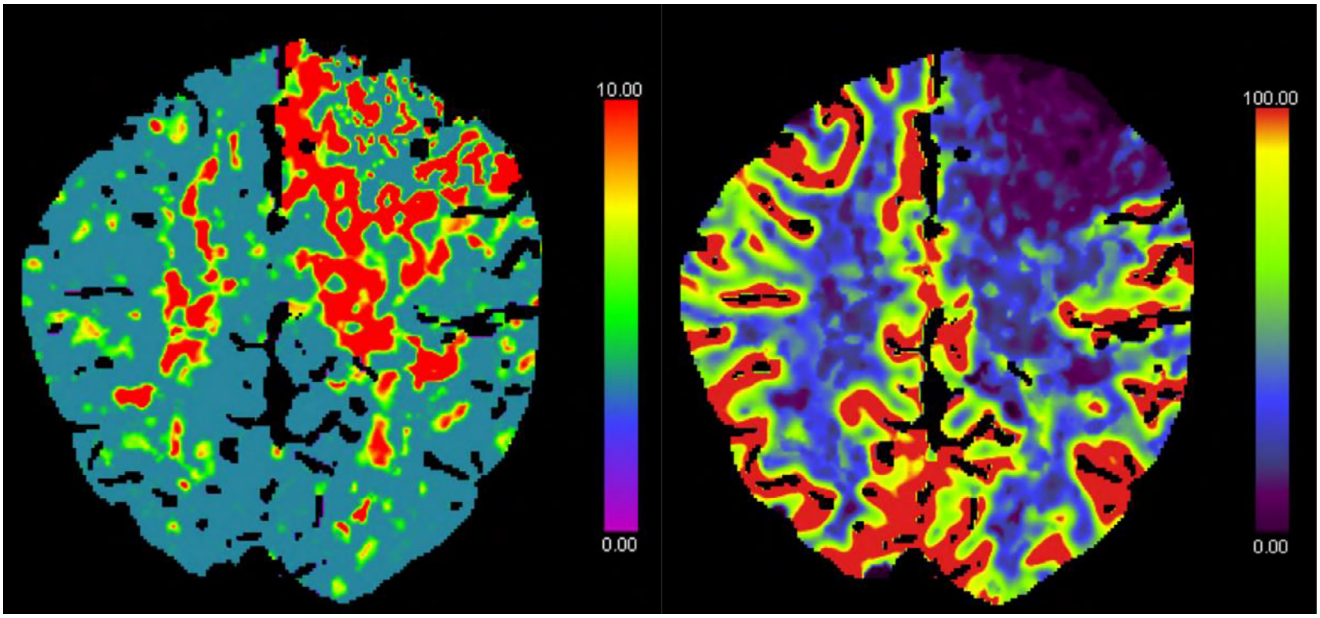

Fig. 3 – Mean transit time and cerebral blood flow perfusion CT scans showed core infarction involving the basal ganglia, frontal lobe and insula.

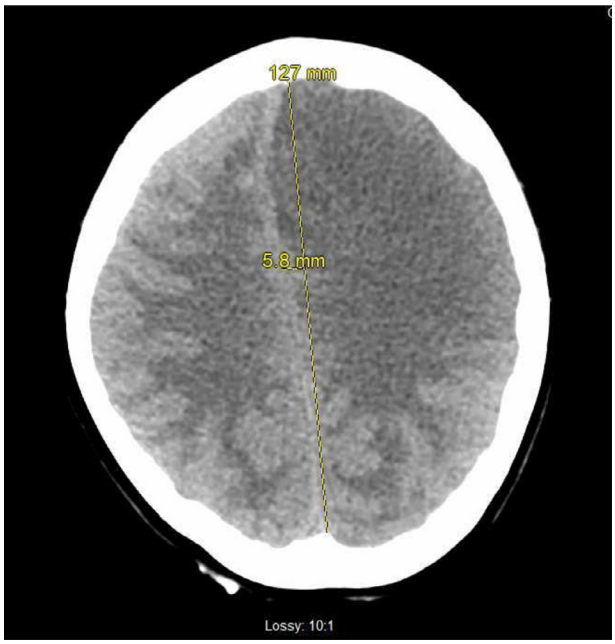

Fig. 4 – CT showed progression of acute infarct with evolution of edema and left to right midline shift.

Fig. 5 – MRA Neck of right carotid showed focal areas of stenosis.

Discussion

TA is a chronic inflammatory arteriopathy of unknown origin [11]. The pathology of this condition results in stenotic changes in the vessel lumina and thrombus formation [6]. The prevalence of stroke in patients with TA has been estimated to be around 10%-20% [1,8,9] and should not be overlooked by physicians. Cerebrovascular complications can be a major cause of morbidity and mortality in patients with TA [12]. Therefore, developing a standardized approach towards complications such as stroke is necessary to prevent morbid events and premature deaths.

In addition, it is recommended that young stroke patients with nonspecific symptoms such as fever, malaise,

dizziness, abnormal pulses, and discrepancy in blood pressure be worked up for TA. In this particular case, the patient had nonspecific findings prior to hospitalization that could have aided in a timelier diagnosis. Symptoms included blurry vision, dizziness, joint pain, and fever. Unfortunately, the diagnosis was made only after the patient suffered a stroke. With severe stenosis on imaging, abnormal pulses and discrepancy in blood pressure, TA was suspected and rheumatologic workup was initiated.

In conclusion, stroke as the initial presentation of TA is rare [10]. It is important to consider TA in stroke patients who may have dyslipidemia and/or hypertension but lack other conventional risk factors for stroke [1,13]. It has been hypothesized that the pathophysiology of stroke might be different in TA and further investigation about the mechanism of stroke in TA is warranted [14].

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