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Annals of Case Reports

Altaraqji S, et al. Ann Case Report 14: 484.

DOI: 10.29011/2574-7754.100484

Case Report

# Arterial Ischemic Stroke in a Child with Isolated Small Internal Carotid Artery: A Case Report and Literature Review

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**Citation:** Altaraqji S, Terkawi RS, Ali A, Al-Shami R , Mohamed K (2020) Arterial Ischemic Stroke in a Child with Isolated Small Internal Carotid Artery: A Case Report and Literature Review. Ann Case Report 14: 484. DOI: 10.29011/2574-7754.100484

**Received Date:** 24 September, 2020; **Accepted Date:** 29 September, 2020; **Published Date:** 05 October, 2020

## Abstract

**Introduction:** Pediatric Arterial Ischemic Stroke (AIS) has significant morbidity and mortality. Various underlying medical conditions such as congenital heart diseases, hemolytic anemia, collagen vascular diseases, and neuro-metabolic disorders are known to be associated with the development of pediatric AIS. Congenital Hypoplasia of the Internal Carotid Artery (HICA) has been reported to be associated with stroke.

**Patients and Methods:** We report the case of an 11-year-old girl who presented with acute right-sided weakness following an emotionally stressful event.

**Results:** Stroke work up revealed smaller left anterior cerebral artery with otherwise normal laboratory and imaging findings.

**Discussion:** HICA is a rare disorder in which the patient may remain asymptomatic or may present with symptoms due to cerebrovascular insufficiency or compression by collateral vessels. In this case report, the patient had a small left anterior cerebral artery that could explain the etiology of her stroke.

**Conclusion:** Isolated small internal carotid artery could be the underlying cause for the development of pediatric AIS.

## Introduction

The diagnosis of stroke in pediatric patients remains challenging, especially in previously healthy children. Many risk factors that lead to childhood stroke have been identified, including congenital heart diseases, hemolytic anemia, collagen vascular diseases, and neuro-metabolic disorders. Over the years, investigators have confirmed the role of thrombophilia in the predisposition to development of Arterial Ischemic Stroke (AIS). Arteriopathy is another major risk factor that has been identified in approximately 53% of pediatric AIS cases [1]. Congenital Hypoplasia of the Internal Carotid Artery (HICA) is a rare clinical entity.

Less than 200 cases of Internal Carotid Artery (ICA) agenesis or hypoplasia have been reported to date. The development of adequate cerebral collateral circulation usually prevents development of symptoms and may be picked up as an incidental finding during neuroimaging for other indication (s). Headaches, seizures, pulsatile tinnitus, Horner's syndrome, and transient ischemic attacks have been reported in patients with HICA. Posterior fossa brain malformations, hemangiomas, arterial lesions, cardiac abnormalities/aortic coarctation, eye abnormalities in PHACE syndrome, Goldenhar syndrome, and Klippel-Feil syndrome have also been reported in association with hypoplasia or agenesis of ICA.

## Case Report

An 11-year-old girl, not known to have any major medical illness earlier, was brought to the pediatric emergency department of our hospital complaining of sudden-onset Right-sided weakness associated with unilateral facial weakness, aphasia, and dysphagia. These complaints had occurred the night before, just after a stressful

argument with her sister. She also had a mild to moderate headache after the event, but no seizures, trauma, or altered behavior. There was no history of fever, rash, vomiting, diarrhea, or recent immunization. Her past medical history was remarkable for speech delay; hence, autism spectrum disorder was suggested. However, her speech skills improved, and she was doing well at school. She was not taking any medication. Parents were non-consanguineous, and family history was negative for vascular diseases or sudden death. Physical examination revealed an overweight girl with body mass index of approximately 29.4 kg/m$^2$ and high blood pressure of 130/85 mmHg.

She had affluent aphasia; her speech was hard to understand, but she understood commands. Right upper motor neuron facial weakness was noted, mainly affecting the lower face in terms of deviation of the mouth to the left side. Further, a subtle weakness in right eyelid closure and eyebrow elevation was noted. She was not able to sit by herself. There was severe hemiparesis on the right side with hypotonia and hyperreflexia. Muscle power was 0/5 in the upper and lower limbs. The left side of the body was normal under motor examination, and she was able to write using the left hand to communicate. Sensation was grossly intact on the left side, but the patient denied having sensation on the right side. An urgent head computed tomography ruled out hemorrhagic stroke. Immediate admission to the pediatric intensive care unit was chosen for heparin infusion. As the echocardiography study was normal, heparin therapy was discontinued, and aspirin was initiated. Thereafter, she was shifted to the neurology ward on aspirin prophylaxis to start rehabilitation care.

### Results of Investigations

Complete blood count, complete metabolic panel, fasting lipid profile, and HbA1c were all normal. Thrombophilia workup was conducted, which included thromboplastin time, prothrombin time, international normalized ratio, protein S activity, protein C activity, antithrombin III activity, Factor 2 activity with prothrombin gene mutation, Factor 5 Leiden mutation, D-dimer, fibrinogen, and homocysteine. All these test results were within the normal range. Tests to rule out the possibility of vasculitis and antiphospholipid syndrome were considered too as lupus anticoagulant antibodies, cardiolipin IgG, beta-2 glycoprotein-1 antibodies, and antinuclear antibody; all tested negative, except for cardiolipin IgM that tested weakly positive.

So, later rheumatology consultation was obtained, and the feedback was that this result was not significant, as there was no evidence of vasculitis on head magnetic resonance imaging. Hemoglobin electrophoresis was done to rule out the possibility of sickle cell disease as a risk factor, it revealed no evidence of hemoglobinopathy. Echocardiogram illustrated normal cardiac anatomy and function, with no evidence of intracardiac thrombus. A fundoscopic evaluation was carried out to look for abnormalities in the retinal vessels associated with stroke and carotid artery hypoplasia. The examination was normal.

## Magnetic Resonance Imaging

### MRA Head and Neck

Normal appearance of the great vessels of the neck including both internal carotid arteries was noted. There was no evidence of arterial dissection. The left ICA was developmentally smaller than the right (Figure1). There was a slight attenuated flow through the left middle and A1 anterior cerebral arteries compared with the right (Figure 2).

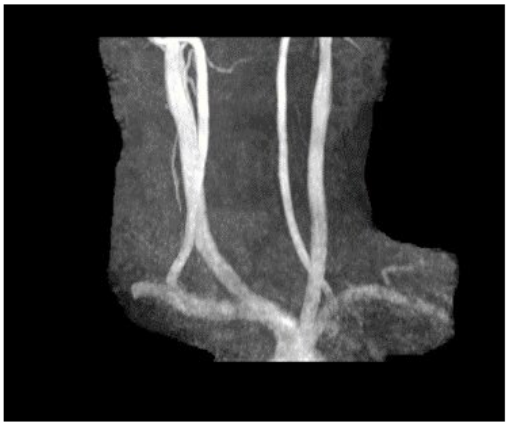

**Figure 1:** MRI flair image showing the left sided MCA stroke.

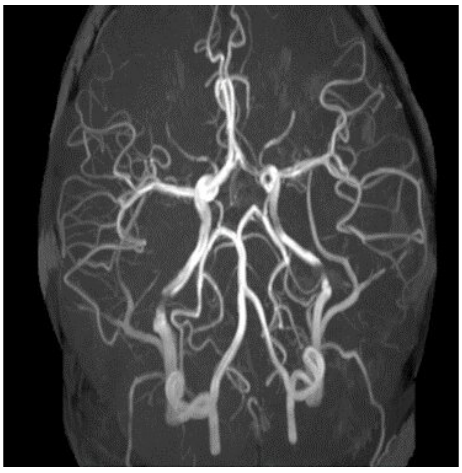

**Figure 2:** MRA showing smaller left ICA.

## MRI Head

It showed evidence of acute infarction within the distal left middle cerebral artery territory with no evidence of brain hemorrhage (Figures 3-5).

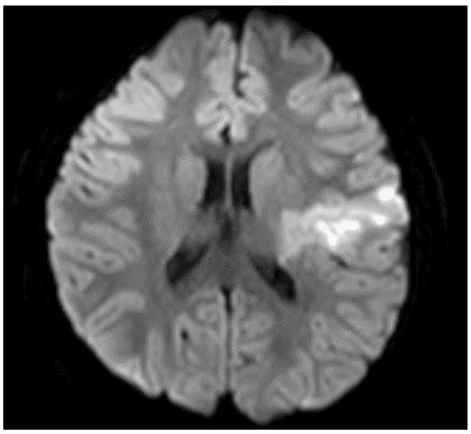

**Figure 3:** Intracranial MRA: demonstrating the small ICA and MCA on the left side.

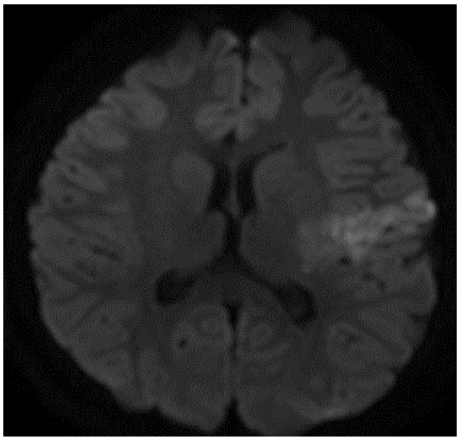

**Figure 4:** MRI head DW.

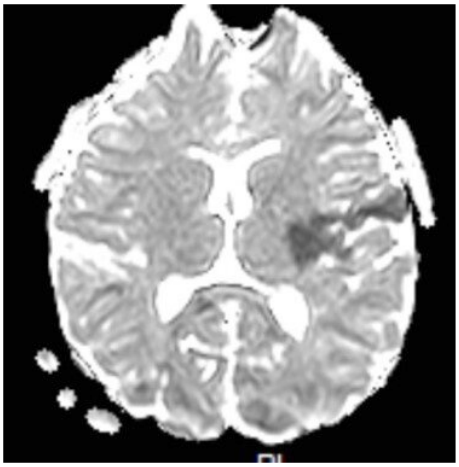

**Figure 5:** MRI head ADS.

**Case Progress**

The patient's speech returned to normal and facial weakness resolved within the first week. Muscle power improved gradually in the limbs and weight-bearing on the right leg was achieved within 2 weeks. She was discharged to a rehabilitation facility with a muscular power of 4/5 in the right lower limb and residual right upper limb weakness. When reviewed in the outpatient clinic 3 months after the stroke, she was walking with a mild hemiplegic gait, she had some weakness of grip and shoulder extension at 4/5 and mild weakness and spasticity in her lower limbs with knee extension and hip abduction at 4/5.

## Discussion

In this case report, we describe a child with an isolated congenital small ICA who presented with a middle cerebral artery ischemic stroke. Arterial ischemic stroke is a compromise in the blood flow leading to brain tissue injury. It is a very rare condition in childhood with an incidence estimated at 5-10 per 100,000 children annually [2]. Diagnosis is made by the presence of focal neurological manifestations with imaging abnormalities. Despite the seriousness and potential persistence of the neurological deficits, AIS diagnosis is commonly delayed in the pediatric age group. This is attributed mainly to the low incidence rate and the possibility of several mimicking conditions for differential diagnoses, such as central nervous system infections, seizures, functional neurological disorders and migraine [2].

Unlike AIS in adulthood where the risk factors are frequently identified prior to its occurrence, childhood AIS is likely to present with no previously known risk factors. Approximately 53% of pediatric AIS cases are associated with arteriopathy, as described by Amlie-Lefond C et al. [1]. In this study, arteriopathies were categorized as idiopathic focal cerebral arteriopathy in 25% of arteriopathies, Moyamoya in 22%, and arterial dissection in 20%. Very few studies have reported carotid artery stenosis/hypoplasia as a risk factor for AIS in children. Congenital hypoplasia or agenesis of the ICA was reported as an isolated risk factor for AIS in a few publications both case reported on adults with congenital hypoplasia who developed arterial stroke without concomitant risk factors [3,4]. Fibromuscular dysplasia of the intracranial artery was described in 1997 as a cause of reversible childhood ischemic stroke in an 8 year old child [5].

However, many other publications have reported hypoplasia of the carotid artery together with other risk factors. For instance, a recent case report described that a 7-year-old boy with factor S deficiency and HICA presented with AIS [6]. Another case report of a 7-month-old infant with a similar clinical scenario, had positive results for IgG anti-cardiolipin and anti-beta-2 glycoproteins I

antibodies test, besides having ICA agenesis [7]. Sfaihi L et al. has also described an infant with hereditary spherocytosis and carotid artery hypoplasia, who developed AIS [8]. Incidental finding of ICA hypoplasia without stroke has been described in association with saccular aneurysms, the study identified ICA hypoplasia as a risk factor for saccular aneurysms in three patients. The authors recommended doing intracranial MRA as part of the initial evaluation of patients with ICA hypoplasia [9]. The overall risk of developing stroke in patient with isolated ICA hypoplasia has not been studied before because of the rarity of the condition. Our case report demonstrates that, given the possibility of recurrence, evaluation of the anatomy of the carotid arteries should be part of the investigation plan for arterial ischemic stroke.

## Conclusion

Isolated small ICA could be an underlying cause for the development of pediatric AIS. Detailed evaluation of the carotid arteries should be part of the evaluation after AIS in children.

## Acknowledgement

We acknowledge the help of Dr Roxana Gunny senior attending radiologist.

## References

1. Amlie-Lefond C, Bernard TJ, Sebire G, Friedman NR, Heyer GL, et al. (2009) Predictors of cerebral arteriopathy in children with arterial ischemic stroke: results of the International Pediatric Stroke Study. Circulation 119: 1417-1423.
2. Amlie-Lefond C (2018) Evaluation and Acute Management of Ischemic Stroke in Infants and Children. Continuum (Minneap Minn) 24: 150-170.
3. Li S, Hooda K, Gupta N, Kumar Y (2017) Internal carotid artery agenesis: A case report and review of literature. Neuroradiol J 30: 186-1891.
4. Tsai MJ, Lin WC, Kuo CH, Chou HF, Yang RC (2007) Acute midbrain infarction in a child with intracranial carotid artery hypoplasia and aberrant cerebral vasculature: a case report. J Child Neurol 22: 465-470.
5. Zurin AA, Houkin K, Asano T, Ishikawa T, Abe H (1997) Childhood ischemic stroke caused by fibromuscular dysplasia of the intracranial artery--case report. Neurol Med Chir (Tokyo) 37: 542-545.
6. Gupta R, Aulakh R, Tiwari A (2019) Arterial Ischemic Stroke in a Child with Internal Carotid Artery Hypoplasia and Protein S Deficiency. J Pediatr Neurosci 14: 52-54.
7. Melit LE, Marginean CO, Simu I, Bucur G (2019) Acute ischemic stroke in a 7-month-old infant, risk factors, and diagnosis peculiarities: A case report. Medicine (Baltimore) 98: e17864.
8. Sfaihi L, Boukedi A, Aloulou H, Kammoun T, Mnif Z, et al. (2010) Hypoplasia of the internal carotid artery: a rare cause of cerebral ischemic stroke in children. Arch Pediatr 17: 1657-1660.
9. Zink WE, Komotar RJ, Meyers PM (2007) Internal carotid aplasia/hypoplasia and intracranial saccular aneurysms: series of three new cases and systematic review of the literature. J Neuroimaging 17: 141-147.