



## Hypoplastic Ipsilateral Internal Carotid Artery as a Cause of Acute Ischaemic Stroke: A Case Report

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### Abstract

Ischaemic stroke caused by carotid artery disease is generally attributed to atherosclerotic disease of the carotid bifurcation. Non-atherosclerotic disease of the carotid arteries causing ischaemic stroke, though documented, is rather rare. Less frequently, carotid artery dissection, fibro-muscular dysplasia and post-traumatic thrombosis have been described as a cause of stroke. Developmental abnormalities of the intra-cranial carotid or vertebral arteries can result in an incomplete formation of the circle of Willis. However, such developmental abnormalities affecting the extra-cranial carotid arteries is extremely rare and have largely been identified only as an autopsy finding among individuals who remained free of cerebral symptoms throughout their lifetime. Nevertheless, the identification of such maldeveloped extra-cranial carotid arteries in patients with ischaemic stroke offers the clinicians a total new dimension in to cerebral perfusion and stroke aetiology.

**Keywords:** Stroke; Cerebral ischaemia; Carotid stenosis; Atherosclerosis; Carotid hypoplasia

### Introduction

Developmental anomalies of the intra-cranial arteries constituting the Circle of Willis have been frequently documented. However, such developmental anomalies affecting the extra-cranial carotid arteries are extremely rare. Among them, Hypoplasia Of The Internal Carotid Artery (HICA) has a reported prevalence of 0.01% [1,2]. HICA involves the relative and significant narrowing of the extra-cranial internal carotid artery along its entire course outside the cranium without any localized region of stenosis or occlusion. Majority of such HICAs remain asymptomatic and are only identified incidentally during imaging for other indications or during autopsy [3]. Collateral circulation from the contra lateral side internal carotid artery as well as bilateral vertebral arteries result in the great majority of these patients remaining asymptomatic. The reported incidence of ipsilateral ischaemic stroke caused by HICA is extremely rare. This report describes a unique patient who presented with acute ischaemic stroke and was found to have an ipsilateral HICA as the only identifiable cause for stroke.

### Case Presentation

A 53-year-old man was transferred to the emergency department with acute onset weakness of the left arm and leg associated with slurring of speech. The symptoms had been persistent for just over 36 hrs by the time he was taken to a regional hospital and then referred to the tertiary hospital. He had no prior history suggestive of stroke, transient ischaemic attacks or amaurosis fugax. He was a non-smoker and had no history suggestive of ischaemic heart disease or peripheral arterial occlusive disease. He had no diabetes or hypertension nor hypercholesterolaemia. He was not on any long-term medication.

On examination, he had weakness of the left arm and leg (muscle power 3/5). There was obvious slurring of speech but no neurological deficit in the facial musculature. His pulse rate was regular and counted at 76 beats per minute. His blood pressure was 155/90 mmHg.

A non-contrast Computerized Tomography (CT) scan was done which confirmed an ischaemic stroke involving the middle cerebral artery region of the right side. The electro-cardiogram with rhythm strip showed normal sinus rhythm. The trans-thoracic echocardiogram showed normal cardiac structure and function with no evidence of cardiac thrombi or vegetations. A carotid duplex scan was also done to assess the extra-cranial carotid and vertebral arteries. The duplex scan findings showed normal structure and flow on the left side but with a thinned out (2.8 mm) narrow lumen internal carotid artery on the right side. This relative narrowing of the lumen was seen throughout the entire course of the artery in the extra-cranial segment. There was no demonstrable localized

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Received Date: 26 Aug 2018

Accepted Date: 15 Sep 2018

Published Date: 17 Sep 2018

#### Citation:

Gunawansa N. Hypoplastic Ipsilateral Internal Carotid Artery as a Cause of Acute Ischaemic Stroke: A Case Report. *World J Surg Surgical Res.* 2018; 1: 1060.

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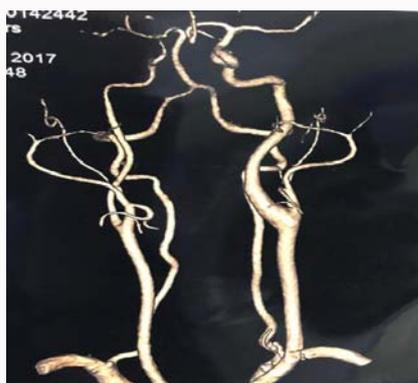


Figure 1: CT Angiogram of the carotid vasculature.

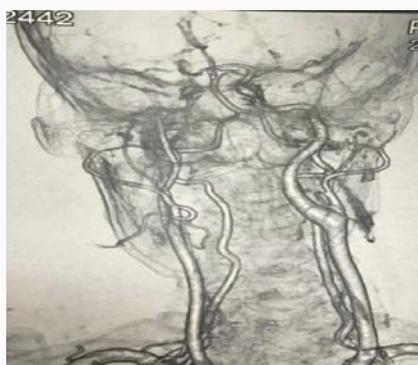


Figure 2: Magnetic Resonance Angiogram of the carotid vasculature.

stenosis or plaque formation within the lumen. The flow velocities were within normal limits with no significant turbulence.

A CT Angiogram (Figure 1) and Magnetic Resonance Angiogram (MRA) (Figure 2) of the carotid vasculature were carried out to better define the carotid artery anatomy. The imaging revealed a thinned out right internal carotid artery throughout its course in to the intracranial segments. The images of the skull base revealed asymmetrically small carotid canal on the right side compared to the left carotid canal. There was no evidence of atherosclerotic disease or carotid dissection. After careful study of the clinical presentation and the imaging, a diagnosis of right sided HICA disease with acute ischaemic stroke was made.

Treatment was started with dual antiplatelets (aspirin and clopidogrel both at 75 mg daily). He was also started on atorvastatin 20 mg daily and ramipril 2.5 mg for blood pressure control. A programmed physiotherapy regime was started and speech therapy rehabilitation was also commenced. Day-05 after admission, the patient was discharged with the above medication to continue physiotherapy and rehabilitation on an out-patient basis.

At 12 months follow-up, the patient remains well and has had no further ischaemic episodes. He has recovered completely from his leg weakness although some residual slurring of speech and some weakness of the left arm (power 4/5) remains.

## Discussion

Developmental anomalies of the extra-cranial carotid arteries are rare with a reported prevalence of 0.01% [1]. As mentioned above, they are mostly detected by incidental imaging or at autopsy. The

incidence of HICA causing ipsilateral ischaemic stroke is extremely rare with reported cases in the literature being around under a hundred cases [4,5]. Among these, approximately half have been reported as bilateral [6].

Lie et al. in his landmark paper on congenital anomalies of the carotid arteries, classified these anomalies in to three basic categories; agenesis, aphasia and hypoplasia [7]. In agenesis and aphasia, the affected internal carotid artery is totally absent in adult life. In Internal carotid hypoplasia, such as was seen in this index patient, it is present but significantly narrowed along its entire course compared to its contra lateral counterpart. The confirmation of diagnosis is by seeing a narrowed carotid canal in the skull base which signifies the absence of a normal sized carotid artery from the developmental stages [8,9]. This finding helps to differentiate HICA from other conditions such as chronic dissection of the artery or fibro muscular dysplasia, where the carotid canal will appear normal in size as it was developmentally normal and acquired the disease later in life. Hence the presence of a narrowed carotid canal signifies a developmental anomaly present from birth, as opposed to an acquired disease of the carotid artery.

HICA can often be associated with other developmental anomalies of the Circle of Willis including aneurysms which may be present in 23% to 45% of patients, compared to 2% to 4% seen in the general population [2,10]. The path physiology behind the formation of such aneurysms is thought to be the asymmetrical hemodynamic with hyper perfusion on the contra lateral side. Once HICA has been identified, the presence of such co-existent anomalies need to be actively looked for and excluded in order to prevent possible cerebrovascular accidents in the future. No other anomalies in the circle of Willis were found in our index patient.

The recommended management of HICA in the absence of co-existent intra-cranial aneurysms remains 'best medical therapy'. This includes the combination of anti platelet drugs, statin therapy and blood pressure control. However, considering the very low incidence of HICA, majority of such treatment evidence comes from anecdotal experience.

## Conclusion

Developmental anomaly of the extra-cranial carotid artery causing carotid hypoplasia is a rare finding usually detected at autopsy. A few cases of such carotid hypoplasia have been described in association with ipsilateral ischaemic stroke. The diagnostic confirmation is based on the demonstration of a comparatively narrowed internal carotid artery along its entire course beyond the carotid bifurcation along with a structurally narrowed carotid canal in the base of the skull. Approximately 1/3 of such patients may have co-existent intra-cranial cerebral aneurysms that may require surgical intervention. In the absence of such co-existent pathology, isolated internal carotid hypoplasia is managed expectantly with 'best medical therapy'.

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