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CASE REPORT

Multiple Intracranial Vascular Anomalies in a Patient with Hypoplasia/aplasia of the Internal Carotid Arteries, Moyamoya Collateralisation, and Aneurysms of the Posterior Fossa Circulation

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ABSTRACT

Bilateral agenesis, aplasia, and hypoplasia of the internal carotid arteries are rare congenital anomalies that are associated with aneurysms of the circle of Willis and dolichoectasia of the vertebrobasilar system. This report of a patient with ruptured posterior fossa aneurysms presenting with subarachnoid haemorrhage illustrates the presence of multiple vascular anomalies, including congenital absence and hypoplasia of the right and left internal carotid arteries, respectively, Moyamoya collateralisation, and posterior circulation aneurysms. The latter ruptured, resulting in symptomatic subarachnoid haemorrhage.

Key Words: *Aneurysm; Carotid artery, internal; Congenital abnormalities; Moyamoya disease; Subarachnoid haemorrhage; Tomography, X-ray computed*

中文摘要

頸內動脈發育不全/不發育合併煙霧病側枝迴圈形成及後顱窩迴圈血管動脈瘤的多發腦血管畸形個例報道

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雙側頸內動脈發育不全，不發育或發育不良是一種少見先天畸形，及與Willis環的動脈瘤及椎基底動脈伸展擴張症有關連。本文報導一例頸內動脈先天右側缺如、左側發育不全，同時合併煙霧病側枝迴圈形成及後顱窩迴圈血管動脈瘤的病人。患者動脈瘤破裂並誘發蛛網膜下腔出血而出現癱狀。

INTRODUCTION

Agenesis, aplasia, or hypoplasia of the bilateral internal carotid arteries (ICAs) are rare congenital malformations,[1] that are associated with aneurysms of the circle of Willis and dolichoectasia of the vertebrobasilar system. Possible causes include secondary regression following a normal initial phase of development or arrest of development at an early embryonic stage. Bilateral aplasia/agenesis of the ICAs is compatible with life when extensive collateral vessels (Moyamoya-like) are established. However, the new haemodynamics predisposes patients to rupture of aneurysms associated with vascular anomalies.[1]

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This report is of a patient with ruptured posterior fossa aneurysms presenting with subarachnoid haemorrhage illustrating the presence of multiple vascular anomalies.

CASE REPORT

A 40-year-old otherwise healthy woman was admitted to the Prince of Wales Hospital, Hong Kong, in 2009 with sudden onset of severe neck pain, radiating to the head and associated with nausea and vomiting. Her blood pressure was marginally elevated. There was no loss of consciousness, prior head injury, or weakness. Urgent unenhanced computed tomography (CT) of the brain showed subarachnoid haemorrhage in the basal cisterns (Figure 1). CT angiogram was performed, which showed 2 small aneurysms (Figure 2), arising from the P1 segment of the right posterior cerebral artery and at the origin of the left anterior inferior cerebellar artery. The entire ICA was absent on the right side and hypoplastic on the left side. These findings were associated with hypoplastic bony carotid canals. Multiple collateral vessels were present in the perimedullary and prepontine cisterns, consistent with Moyamoya collateralisation. The right middle cerebral artery (M1) was absent and the right anterior communicating artery (A1) was aplastic. The anterior circulation was supplied by the posterior fossa circulation via the posterior communicating artery and the external carotid arteries. The patient was treated conservatively for symptomatic relief and blood pressure control.

DISCUSSION

The term agenesis is used when there is total failure of ICA formation in the embryo. Aplasia is used when a precursor is present in the embryo and is represented by a remnant such as a fibrous band. Hypoplasia is used when a small calibre ICA is present.[2] Embryologically, the ICA forms from the third aortic arch and the common carotid artery (CCA) from the ventral aortic root between the third and fourth arches. Lasjaunias and Santoyo-Vazquez have suggested that the ICAs can be divided into 6 segments — cervical, petrous, vertical cavernous, horizontal cavernous, clinoid, and cisternal — which develop from the embryonic arteries.[3] As each segment is independent, an anomaly in a segment may result in segmental agenesis of the ICA. Formation of the ICA is completed at the sixth week of gestation (fourth embryonic week), while the carotid canals form between the fifth and sixth embryonic weeks.[4] As the presence of the ICAs is a prerequisite for the formation of the carotid canals, ICA agenesis results in absence of the carotid canals. In patients with aplasia or hypoplasia, the size of the carotid canals is reduced,[5] which was evident in this patient.

Patients with congenital ICA agenesis, aplasia, or hypoplasia are usually asymptomatic as cerebral perfusion is usually adequate due to compensatory supply by the collateral vessels.[6] Cerebral perfusion can be maintained by collateral flow from the posterior circulation

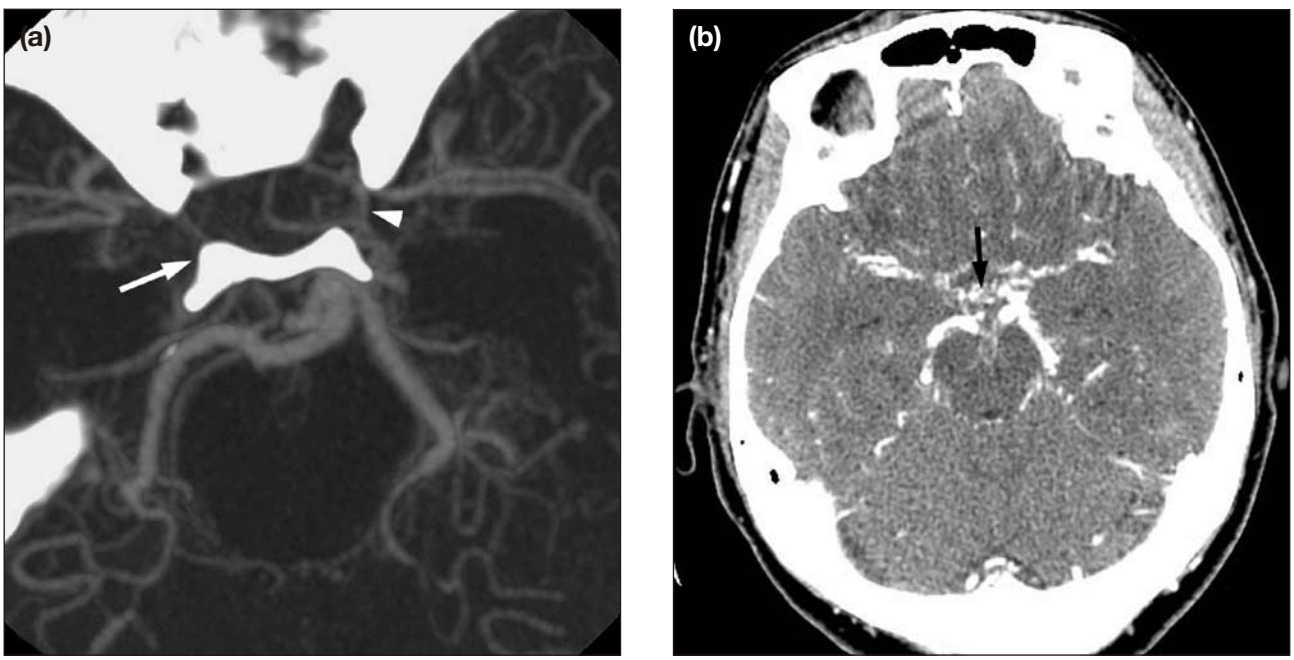

Figure 1. Contrast-enhanced computed tomography of the brain. (a) Axial reconstructed image in maximum intensity projection showing absent internal carotid artery (arrow) and the small-calibre internal carotid artery consistent with hypoplasia (arrowhead); (b) axial image showing multiple collaterals in the prepontine cisterns (arrow) consistent with Moyamoya disease.

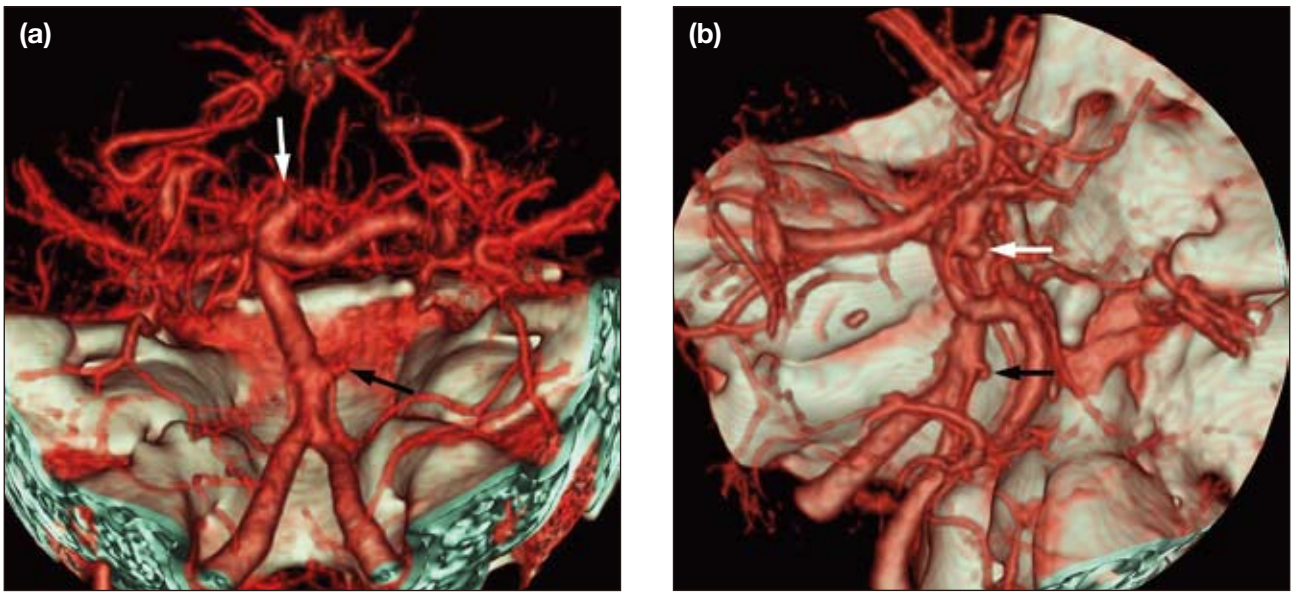

Figure 2. Three-dimensional volume-rendered computed tomography angiography images. (a) A 2-mm saccular aneurysm at the origin of the left anterior inferior cerebellar artery (black arrow), and multiple collaterals constituting Moyamoya appearance (white arrow); and (b) 2 small (2 mm) aneurysms in the posterior circulation, 1 at the P1 segment of the right posterior cerebral artery (white arrow) and another at the origin of the left anterior inferior cerebellar artery (black arrow).

through the posterior communicating arteries, or by a persistent foetal circulation or, transcranially, through the branches of the internal maxillary artery.[7] Dilatation of the vertebrobasilar system, anterior circulation, or external carotid system results in adequate collateral blood supply in the agenetic segment. In unilateral agenesis of ICA, the predominant supply is through the contralateral ICA.[6] In patients with bilateral agenesis of the ICAs, the supply is from the vertebrobasilar system.[2] In this patient, the supply was predominantly from the posterior circulation via the posterior communicating artery and partly from the external carotid system.

Some patients with ICA agenesis or hypoplasia may present with symptoms of cerebrovascular insufficiency, such as transient ischaemic attacks, hemiparesis, or hypotensive episodes in adulthood. Occasionally, patients may have complications of subarachnoid or intracerebral haemorrhage related to the presence of aneurysms or arteriovenous malformations. The explanation for development of flow-related aneurysms is accounted for by the haemodynamic stress on the cerebral arteries as a result of increased flow through the collateral channels. Two aneurysms were present in the posterior circulation in this patient. The prevalence of cerebral aneurysms in the general population is 2% to 4% compared with 24% to 34% in patients with ICA agenesis.[2,6-8]

Aneurysms in the anterior communicating artery are most commonly described in the literature,[7,9] followed by the basilar artery,[10] posterior communicating artery, posterior cerebral artery, and the first part of the subclavian artery. Other associated conditions are cerebral arteriovenous malformations, hypopituitarism, neurofibroma, interruption of the aortic arch, aberrant origin of the ophthalmic artery, CCA agenesis/hypoplasia, basilar artery aplasia with nasopharyngeal angiofibroma, corpus callosum agenesis,[11] and congenital temporomandibular joint ankylosis.

In conclusion, this report is of a patient with ruptured posterior fossa aneurysms presenting with subarachnoid haemorrhage. On careful evaluation of CT angiography, the aneurysm was found to be related to a complex spectrum of vascular abnormalities, including congenital aplasia of the right ICA, hypoplasia of the left ICA, and associated Moyamoya collateralisation. CT angiography is the imaging technique of choice for patients presenting with an acute cerebral vascular event, as it provides a useful guide to the management of the condition, and may reveal unexpected cerebral vascular abnormalities.

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