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CASE REPORT

Bilateral Congenital Hypoplasia of the Internal Carotid Arteries

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ABSTRACT

Bilateral congenital hypoplasia of the internal carotid arteries, terminating distally as ophthalmic arteries, was found in a 20-year-old man during investigation for intracranial haemorrhage. The magnetic resonance imaging and digital subtraction angiography findings of this rare congenital malformation are described and common associated abnormalities discussed.

Key Words: *Angiography; Carotid artery, internal; Congenital abnormalities; Magnetic resonance imaging*

INTRODUCTION

Developmental abnormalities of the internal carotid artery (ICA) are rare and associated with a higher prevalence of intracranial aneurysms.[1] These may remain asymptomatic or may present as intracranial haemorrhage or manifestations of cerebrovascular ischaemia. This report presents the imaging appearance of bilateral congenital hypoplasia of the ICAs, continuing distally as ophthalmic arteries.

CASE REPORT

A 20-year-old man presented with acute onset of severe headache and vomiting. Computed tomography (CT) scan of the head showed left temporo-occipital haemorrhage, without subarachnoid component. Other relevant haematological investigations were unremarkable. Subsequent magnetic resonance imaging (MRI) and MR angiography (MRA) of the brain demonstrated markedly attenuated cavernous and petrous segments of both ICAs, with diminished ICA flow signal on MRA source images (Figure 1). The vertebro-basilar system was prominent with tortuous posterior communicating arteries (PCoA) and reforming supracavernous segments of ICAs and its branches bilaterally (Figures 2a and b). Review of the CT scan revealed hypoplastic bony carotid canals at the skull base on both sides, confirming the congenital nature of this abnormality.

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A digital subtraction angiogram was then performed with a view to further evaluating the vascular anatomy and to ruling out any other abnormality not well-depicted by MRA. At angiography, both ICAs were uniformly narrowed in their cervical, petrous, and cavernous segments and were terminating as ophthalmic arteries bilaterally (Figure 3). The supraclinoid ICAs, anterior cerebral artery, and middle cerebral artery were not opacified at the carotid injection. These arteries were instead seen to be filling on the vertebral injection through the prominent and tortuous PCoAs (Figure 4), as also

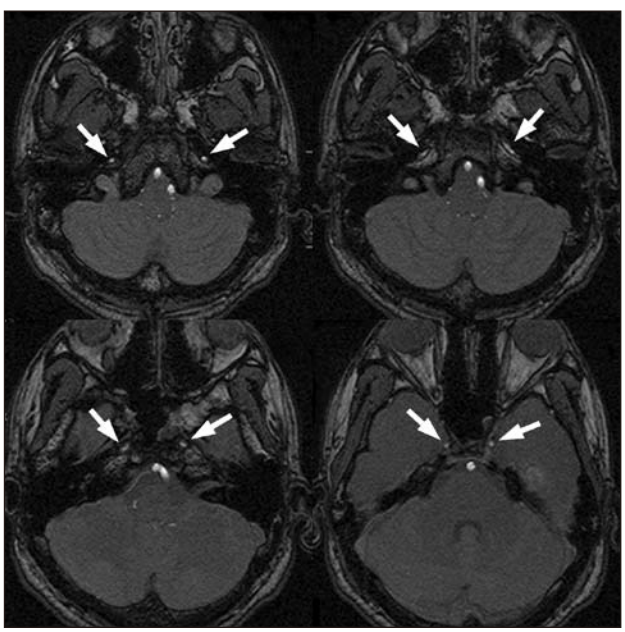

Figure 1. Source images from 3-dimensional time-of-flight magnetic resonance angiogram showing diminished flow-related signal of both internal carotid arteries (arrows) in hypoplastic carotid canals. Also note the prominent vertebro-basilar system.

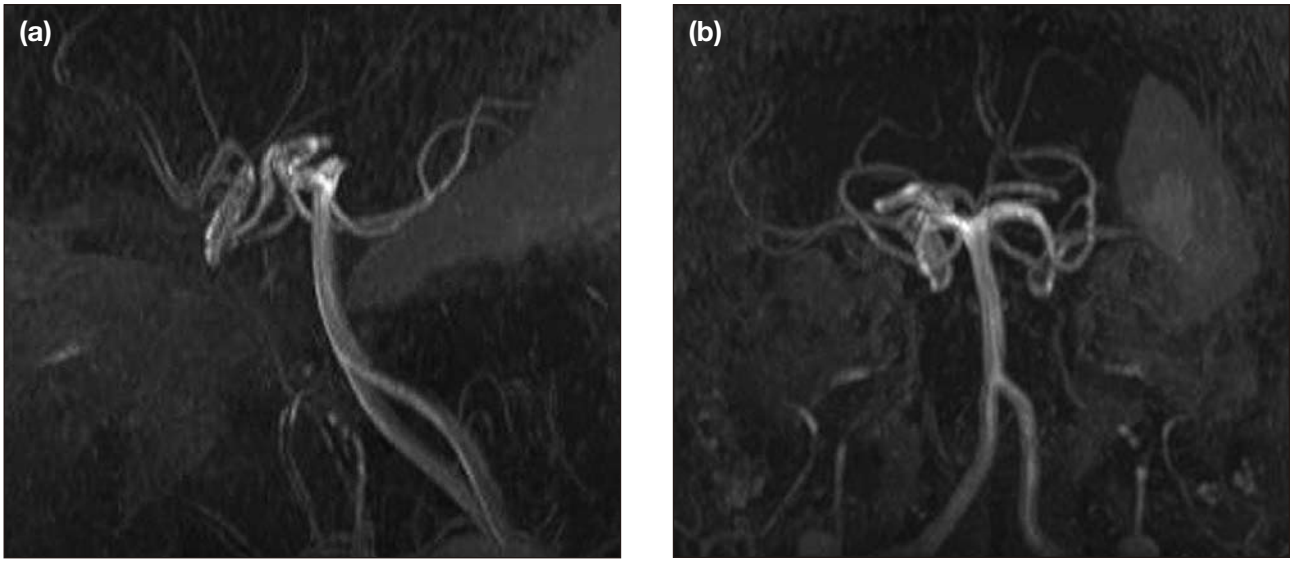

Figure 2. (a) Lateral and (b) frontal projections of magnetic resonance angiogram showing absence of flow-related signal intensity of both proximal internal carotid arteries with prominent vertebro-basilar system.

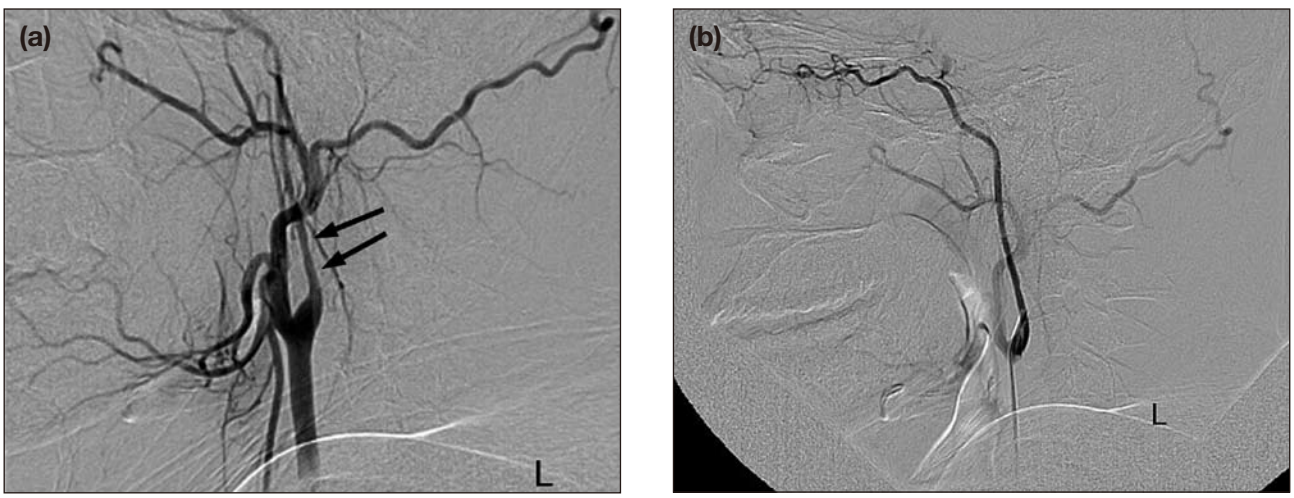

Figure 3. Lateral projection of (a) the left common carotid and (b) the left internal carotid arteriograms showing diffuse narrowing of the left internal carotid artery (arrows) continuing distally as the ophthalmic artery. Supracavernous branches are not opacified. Similar findings were also seen on the right side.

noted on the MRA. A good-calibre anterior communicating artery was present. The external carotid artery (ECA) and its branches were unremarkable bilaterally. No intracranial aneurysm or vascular malformation that might have been responsible for the parenchymal bleed were detected. The patient subsequently recovered and attends regular follow-up.

DISCUSSION

Congenital absence of the ICA is a rare developmental abnormality, occurring in less than 0.01% of the population.[1] The term 'absence' is commonly used and includes the spectrum of agenesis, aplasia, and hypoplasia, which are postulated to result from insult to the embryo during its early development.[2] In agenesis and aplasia, the ICA is completely absent and is not demonstrable angiographically, whereas a diffusely narrowed ICA is present in hypoplasia, indicating incomplete development. Unilateral ICA abnormalities are more frequent than bilateral ICA hypoplasia or agenesis.[3] Hypoplasia of the cervical, petrous, and cavernous segments of both ICAs, continuing as an ophthalmic artery, as seen in this patient, has rarely been reported in the literature.[4]

In these conditions, 3 major collateral pathways to the anterior cerebral circulation have been described,[3,5] the most common of which is through the enlarged PCoAs, as for this patient. More rarely, there may be an anastomosis between the ECA and ICA at the skull base or a trans-sellar anastomosis from the contralateral

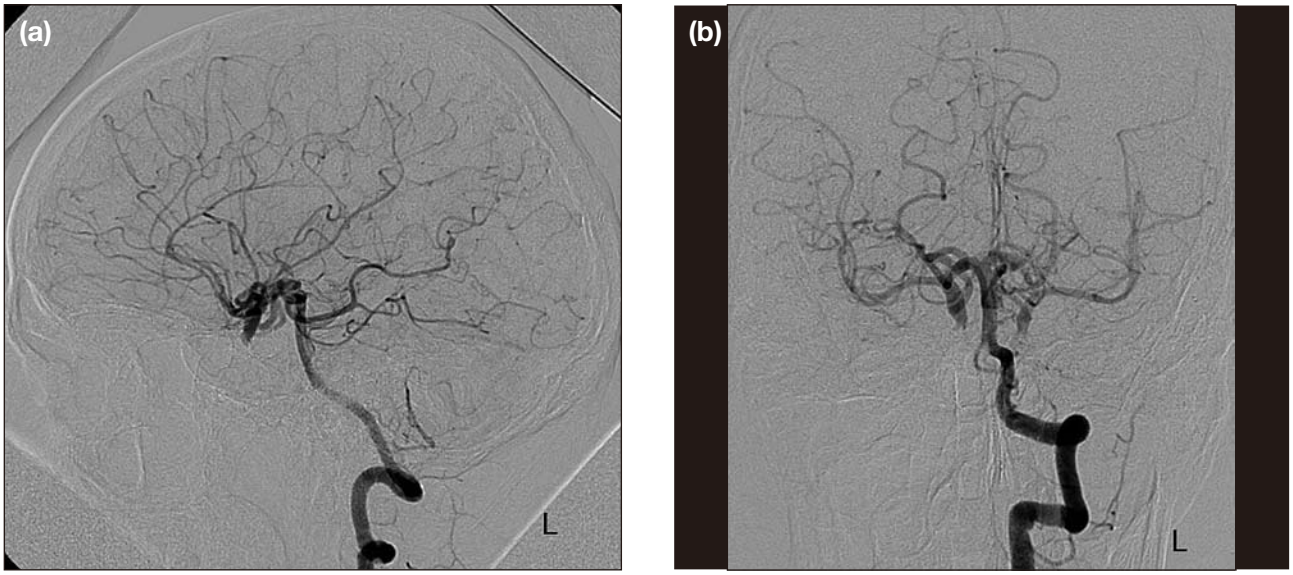

Figure 4. (a) Lateral and (b) frontal projections of the left vertebral arteriogram showing reformation of the bilateral supracavernous internal carotid arteries and their branches through prominent posterior communicating arteries.

ICA, providing collateral circulation. Lie has classified the collateral circulation more descriptively into 6 types.[2] However, this classification does not include the type seen in this patient, suggesting the need for revaluation of the existing classification, as also proposed by Akfirat et al.[4]

Development of the carotid canals at the skull base occurs in the presence of embryonic ICA in early gestation. Demonstration of a small or absent carotid canal therefore indicates a congenital ICA abnormality, enabling differentiation from acquired causes of ICA narrowing, such as chronic dissection or fibromuscular dysplasia.[3,6]

Although many patients with congenital ICA abnormality may remain asymptomatic,[2] there is a high prevalence of intracranial aneurysms of 24% to 34% in these patients compared with 2% to 4% in the general population.[1,7] These aneurysms are thought to result from altered haemodynamics or an underlying genetic defect.[1] No intracranial aneurysms or other vascular abnormalities were detected in this patient that could explain the parenchymal haemorrhage. As the results of other relevant investigations did not show a cause for the haemorrhage, these authors hypothesised that this patient may have coexisting parenchymal vascular abnormalities, probably angiographically occult, which could also result in intracranial haemorrhage. Non-aneurysmal parenchymal or ventricular haemorrhage in patients with ICA hypoplasia has been described previously, and attributed to hypertension.[4] Other presenting symptoms may be related to pressure effects from enlarged collateral vessels, cerebrovascular insufficiency or, rarely, congenital Horner's syndrome.[8]

Congenital hypoplasia of both ICAs is a rare abnormality. Congenital hypoplasia can be differentiated from acquired ICA narrowing by demonstration of hypoplastic bony carotid canals at the skull base. Although many of these patients remain asymptomatic, the increased prevalence of intracranial vascular abnormality is an indication for clinical and radiological surveillance for these patients.[3]

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