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## CASE REPORT

# Congenital Agenesis of the Internal Carotid Artery Mimicking Suprasellar Aneurysm

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## ABSTRACT

***Congenital agenesis of the internal carotid artery is a rare malformation. Although the condition can be regarded as a normal variant, recognition and knowledge of such an entity is important because it has important implications during carotid and trans-sphenoidal surgery and interventional stenting. Congenital agenesis of the internal carotid artery is also associated with other vascular abnormalities such as cerebral aneurysms. This report is of a patient with congenital agenesis of the right internal carotid artery that was detected incidentally by computed tomography scan.***

***Key Words:*** *Carotid artery, internal; Intracranial arteriovenous malformations; Tomography, X-ray computed*

## 中文摘要

### 擬似蝶鞍上動脈瘤的頸內動脈先天性發育不全

**蕭廣樂、單雅怡、馬嘉輝**

頸內動脈先天性發育不全屬於罕見的畸形病例。雖然這種情況可介定為正常變異，但由於這種病在進行頸動脈和經蝶竇手術以及介入支架植入術時具重要的意義，所以對此病症的認識和理解不可或缺。頸內動脈先天性發育不全亦會與其他血管畸形如顱內動脈瘤相關。本文報告一宗進行電腦斷層造影檢查時偶然發現的右頸內動脈先天性發育不全的病例。

## INTRODUCTION

Congenital agenesis, aplasia, or hypoplasia of the internal carotid artery (ICA) is a rare disorder. The incidence is very low, with the condition occurring in less than 0.01% of the population.[1-4] Around 100 patients have been reported in the English-language literature to date. Acquired causes of ICA narrowing or occlusion must be excluded before making the diagnosis of absence of the ICA. Although many patients with absence of the ICA remain asymptomatic and the condition goes unrecognised, these patients may present later in life with symptoms related to cerebrovascular insufficiency. This report is of a patient with congenital absence of the ICA who presented with visual loss.

## CASE REPORT

A 74-year-old man presented in September 2010 with left-side blurred vision. Initially, he was diagnosed with

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cataract and he underwent surgery. However, the visual acuity in his left eye continued to deteriorate over time. Fundal examination showed a pale left optic disc and he was diagnosed with optic atrophy.

Computed tomography scan, done to exclude a mass lesion along the left optic nerve pathway, showed a fusiformly dilated cavernous segment of the left ICA, suspicious of an aneurysm (Figure 1). The right ICA was not visualised.

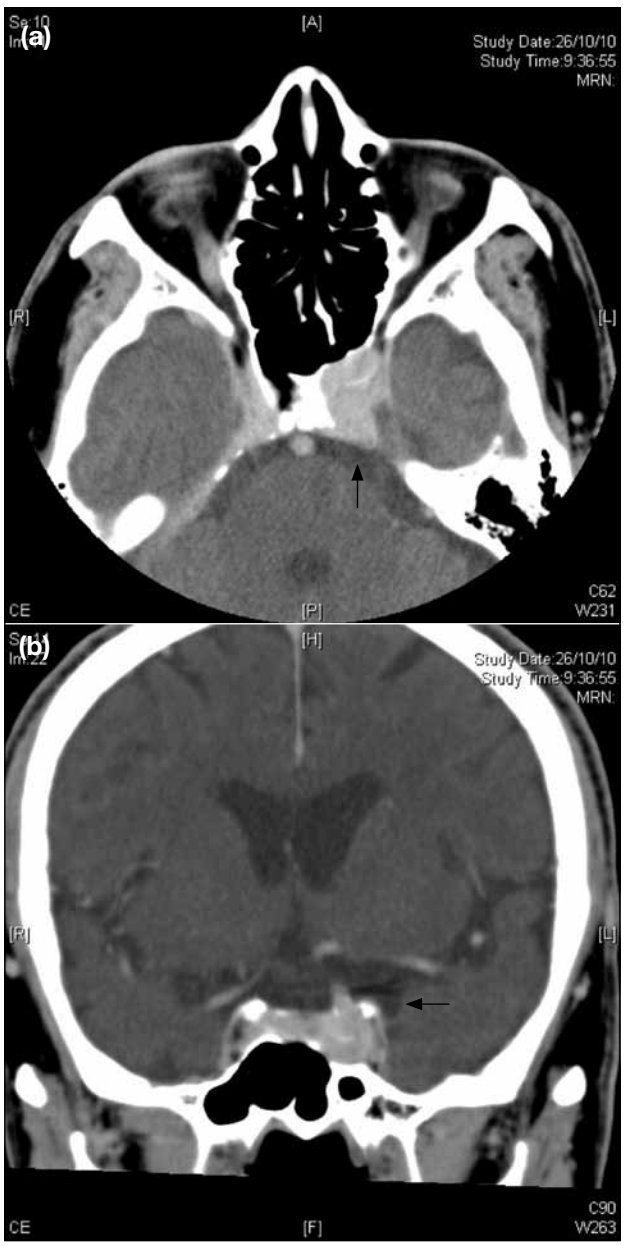


**Figure 1.** Contrast-enhanced computed tomography scans showing (a) an enhancing soft tissue lesion (arrow) at the left cavernous sinus with bony erosion, and (b) coronal view showing the left distal internal carotid artery exiting the lesion (arrow).

Magnetic resonance imaging study was performed to further characterise the lesion, and right ICA agenesis with contralateral ICA hypertrophy was confirmed (Figure 2).

## DISCUSSION

Congenital agenesis, aplasia, or hypoplasia of the ICA was first described by Tode in 1787.[5] However, its exact cause is still unknown. Currently, the condition is thought to represent an insult to the developing embryo with redevelopment of vascular collaterals.[6]

Lie[7] described six collateral pathways. Type A is categorised as unilateral absence of the ICA with collaterals from the anterior and posterior communicating arteries. In type B, the anterior cerebral artery (ACA) and middle cerebral artery (MCA) are supplied by anterior communicating arteries. In type C, there is bilateral ICA agenesis with collaterals from the vertebrobasilar system. In type D, there is unilateral agenesis with collaterals from the contralateral cavernous ICA. In type E, there is bilateral ICA and ACA hypoplasia with both MCAs supplied by enlarged posterior communicating arteries. In type F, there are internal maxillary artery collaterals of external carotid system anastomosis (Figure 3). Some authors include persistent trigeminal artery as an addition type.[8] This patient had a type A anomaly.

Acquired ICA stenosis or occlusion mimicking congenital hypoplasia or agenesis is a common condition in elderly patients. Common causes of acquired ICA stenosis include dissection, atherosclerosis, and fibromuscular dysplasia. Differentiation between a congenital and acquired cause is critical. Demonstration of the absence of the carotid canal at the skull base is diagnostic of congenital agenesis of the ICA (Figure 4).

In this patient, the tortuous collateral from the contralateral side mimicked a suprasellar aneurysm. It is therefore important to exclude this condition before planning any surgical or interventional procedure. Interventional stenting is a common procedure for carotid artery stenosis; before stenting, differentiation of congenital from acquired stenosis must be confirmed.

Most cases of absent ICA are found incidentally. This suggests that the natural collateral development is able to support cerebral perfusion. However, there is an increased risk of cerebral aneurysm formation in association with ICA agenesis, with a reported

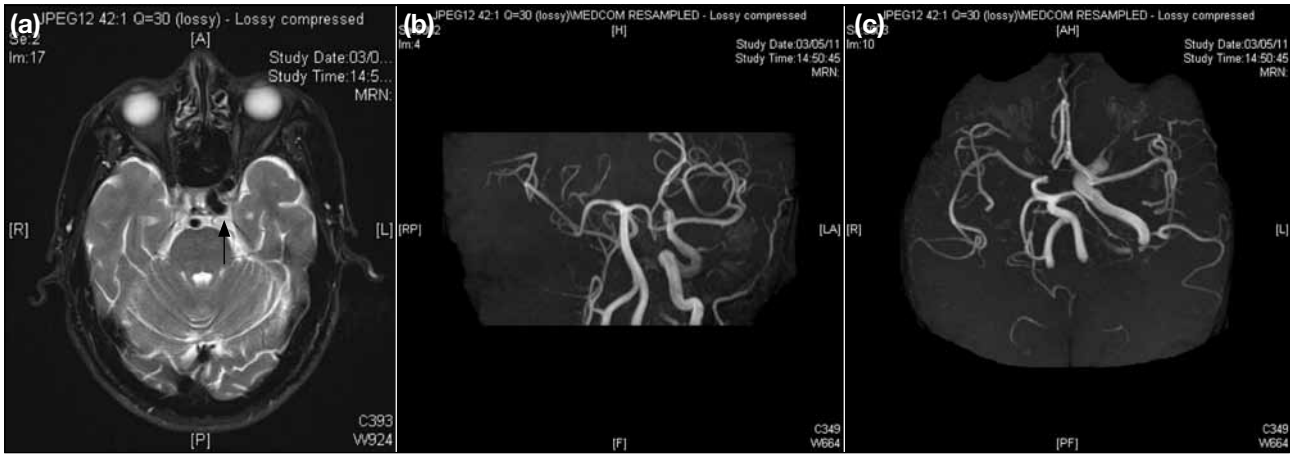


**Figure 2.** (a) A T2-weighted magnetic resonance image showing signal void (arrow) representing the left internal carotid artery, and (b, c) magnetic resonance angiography showing absence of the right internal carotid artery with the right middle cerebral artery supplied by the right posterior cerebral artery.

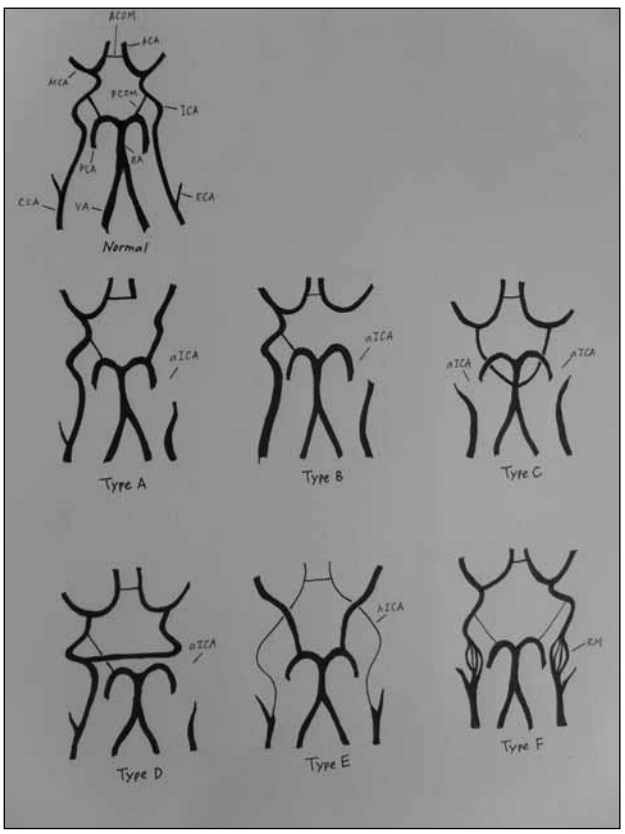


**Figure 3.** Absence of the internal carotid artery and types of collateral blood flow.
Abbreviations: ACA = anterior cerebral artery, ACOM = anterior communicating artery, MCA = middle cerebral artery, PCOM = posterior communicating artery, ICA = internal carotid artery, ECA = external cerebral artery, BA = basilar artery, PCA = posterior cerebral artery, CCA = common carotid artery, VA = vertebral artery, aICA = agenesis internal carotid artery, hICA = hypoplastic internal carotid artery, RM = rete mirabile (collaterals).

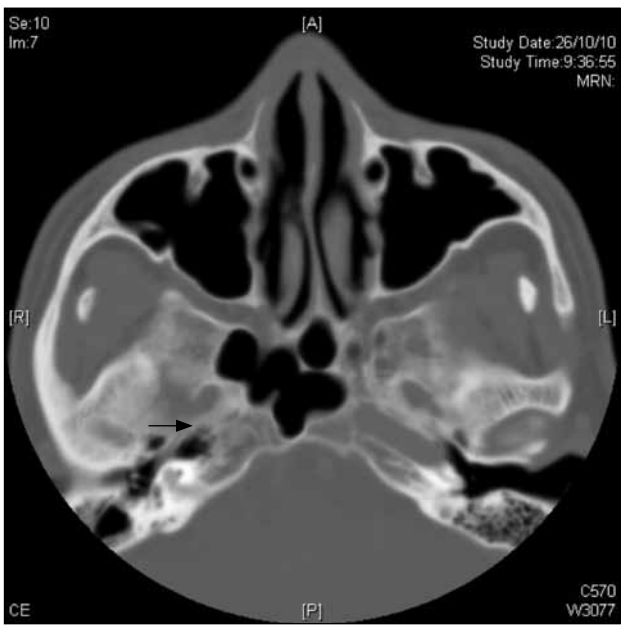


**Figure 4.** A plain computed tomographic image of the skull base with bone window showing the absent right carotid canal (arrow). The left carotid canal is well visualised.

prevalence of up to 34% in patients with ICA absence compared with 4% in the general population,[4,9] so some authors suggest that these patients require long-term follow-up. There is also an association with atherosclerosis and thromboembolism, probably related to cerebral haemodynamic disturbance.[1-4,9]

This report is of a patient with absent ICA. Although uncommon, this condition is important because of the

associated vascular and parenchymal disorder. Absence of the carotid canal at the skull base is the diagnostic feature that allows differentiation from the more common acquired stenosis or occlusion.

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