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

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.

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. 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.

Lie 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. 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
prevalence of up to 34% in patients with ICA absence compared with 4% in the general population, 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.

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.

REFERENCES