Bilateral hypoplasia of the internal carotid arteries

Luk Y S, Man E M W, Sy A N L

ABSTRACT

Bilateral hypoplasia of the internal carotid arteries is a rare congenital anomaly. We present the case of a 62-year-old man with this rare condition. The findings from the plain computed tomography and computed tomography angiogram are described in this report. The common collateral pathways associated with bilateral hypoplasia of the internal carotid arteries, the clinical presentation and clinical significance of this condition are also discussed.

Keywords: agenesis, carotid artery, computed tomography, congenital abnormalities, internal

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INTRODUCTION

Congenital anomalies of the internal carotid artery are rare occurrences. This condition is associated with a higher prevalence of intracranial aneurysms. Patients may remain asymptomatic or may present with symptoms of cerebrovascular ischaemia or complications of intracranial haemorrhage.[1] This case report presents the imaging appearance of bilateral hypoplasia of the internal carotid arteries in a 62-year-old man.

CASE REPORT

A 62-year-old Chinese man with a history of diabetes mellitus, hypertension and ischaemic heart disease presented to our hospital with sudden onset of dizziness. Physical examination revealed left-sided ataxia. Plain computed tomography imaging of the brain showed left cerebellar infarct and bilateral hypoplastic carotid canals at the skull base (Fig. 1). Subsequent ultrasonography of the neck arteries revealed a significantly small calibre of bilateral internal carotid arteries. Computed tomography angiogram of the brain showed that both intracranial arteries were uniformly narrowed in their cervical, petrous and cavernous segments. (Figs. 2 & 3) The vertebrobasilar system was prominent, with a prominent right posterior communicating artery (Fig. 4). The external carotid arteries were unremarkable bilaterally. No intracranial aneurysm was detected. The patient was put on aspirin. Gradual improvement was noted in the patient's neurological condition, and he was subsequently discharged.

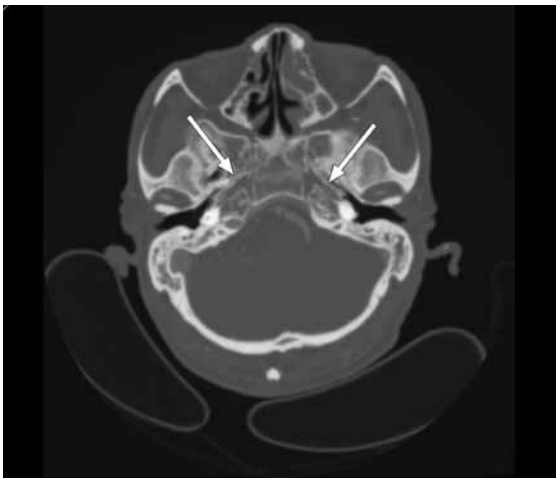

Fig. 1 Axial CT image of the skull base shows bilateral hypoplastic carotid canals (arrows).

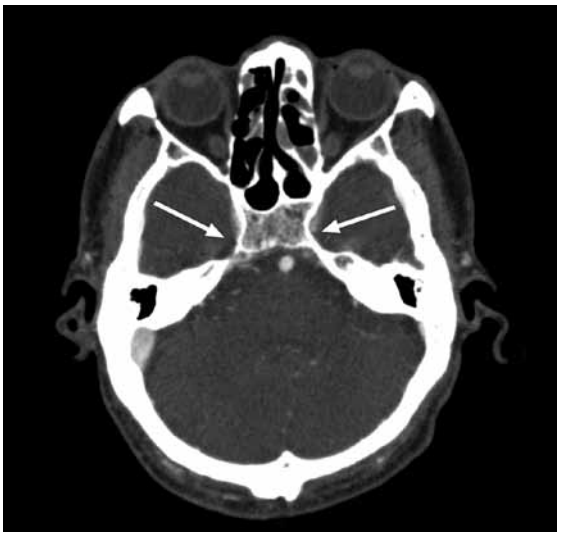

Fig. 2 Lateral projections of CT angiograms show (a) diffuse narrowing of the cervical segments of the left (arrow) and (b) right internal carotid arteries (arrow).

DISCUSSION

Congenital hypoplasia of the internal carotid artery

Department of Diagnostic Radiology, Pamela Youde Nethersole Eastern Hospital, 3 Lok Man Road, Chai Wan, Hong Kong

Luk YS, MBBS
Resident

Man EMW, MBChB, FRCR
Resident

Sy ANL, MBBS, FHKAM
Associate Consultant

Correspondence to:
Dr Luk Yiu Shiobhon
Tel: (852) 6083 2248
Fax: (852) 2794 3241
Email: shiobhon@gmail.com

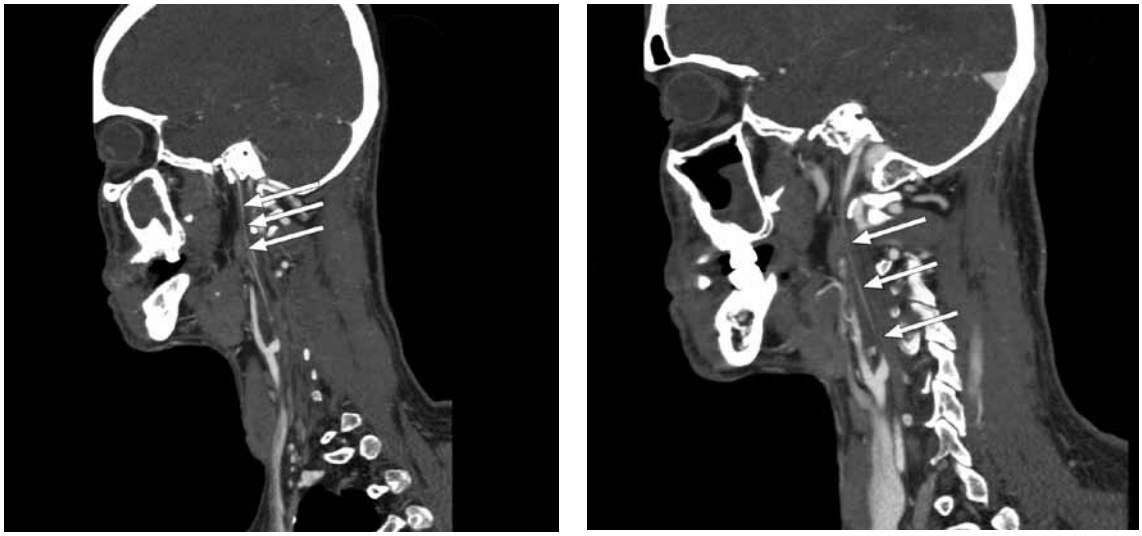

Fig. 3 CT angiograms show significantly narrowed cavernous segment of the bilateral internal carotid arteries (arrows).

is uncommon and occurs in less than 0.01% of the population.[2] About 100 cases have been reported in the literature.[1] The term hypoplasia is part of the spectrum of 'absence', which includes agenesis, aplasia and hypoplasia. In agenesis and aplasia, the internal carotid artery is completely absent, whereas in hypoplasia, development of the internal carotid artery is incomplete.[3] Unilateral absence of the internal carotid artery is more frequently encountered than bilateral absence.[4] A postulated cause of the absence of the internal carotid artery is insult to the embryo during its early development.[5]

Three major collateral pathways to the anterior cerebral circulation have been described. The most common collateral pathway is through the enlarged posterior communicating artery, as was the case for this patient. The other types of collateral pathways are through anastomosis between the external carotid artery and the internal carotid artery at the skull base, or via persistent foetal circulation.[5]

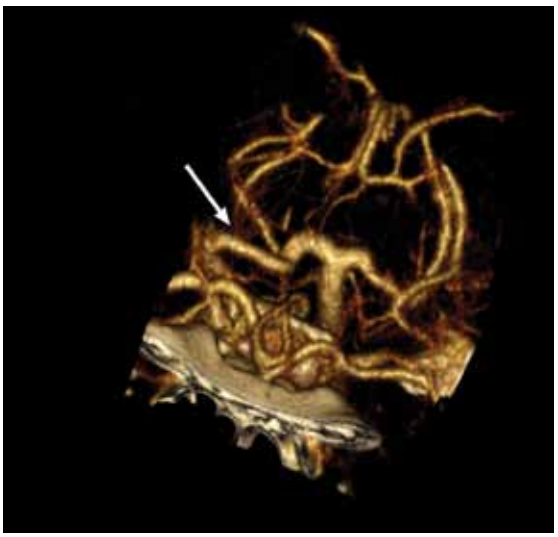

Fig. 4 Three-dimensional CT cerebral angiogram shows a prominent vertebrobasilar system and a prominent right posterior communicating artery (arrow) extending forward to supply the right middle cerebral artery.

Development of the carotid canals at the skull base occurs in the presence of the embryonic internal carotid artery during early gestation. A small or absent carotid canal therefore indicates a congenital internal carotid abnormality, differentiating it from the acquired causes of internal carotid artery narrowing.[4] Chronic dissection, severe atherosclerosis and fibromuscular dysplasia are the acquired causes of a significantly narrowed internal carotid artery.

Most patients with an absent internal carotid artery remain asymptomatic due to the presence of collateral vessels.[1,2] Some patients present with recurrent headache, blurred vision and convulsions.[1] Some may also present with symptoms related to cerebrovascular insufficiency, as the anterior circulation may be dependent on a single carotid artery or the vertebrobasilar system, which may be compromised by atherosclerosis.[5] Absence of the internal carotid artery may be associated with aneurysm, with a high prevalence of intracranial aneurysms of 24%–34%, compared to a 2%–4% prevalence observed in the general population.[5] The association with aneurysms may be related to altered haemodynamics and the result of a developmental defect. Increased regional blood flow, along with congenital defects of the vessel wall and systemic hypertension, are related to the development of cerebral aneurysms.[3,5] Absence of the internal carotid artery may also be associated

with cerebral hypoplasia, and rarely, with congenital Horner's syndrome.[1,6,7]

Recognition of the absence of carotid artery is important when planning carotid endarterectomy, as both cerebral hemispheres may be dependent on a single carotid artery. It is also important to recognise the intercavernous collateral pathway when planning transphenoidal hypophyseal surgery.[5] Hypoplasia of both the internal carotid arteries is a rare anomaly. Although most patients remain asymptomatic, the association with a high prevalence of aneurysm is an indication for clinical and radiological surveillance for these patients.[3] Also, the recognition of this condition is important in thromboembolic disease during carotid or transphenoidal surgery.

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