Congenital absence of the external carotid artery: Atherosclerosis without a bifurcation

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We report the case of a patient with congenital absence of the external carotid artery in whom we performed a carotid endarterectomy. The radiographic features and operative findings are presented. Four similar cases previously reported in the literature are reviewed. A comment on the pathophysiology of atherosclerosis at the carotid bulb in the absence of a bifurcation and a brief discussion on the possible embryologic explanation of this anomaly are discussed. (J Vasc Surg 2002;35:573-5.)

Multiple anomalies of the carotid artery circulation have been described. Absence of the external carotid artery is extremely rare. In this anomaly, there is a single carotid vessel without an identifiable bifurcation. The branches that usually arise from the external carotid artery are observed arising from the internal carotid artery. A search of the modern literature yielded only four cases that described this anomaly.

Disruption of laminar blood flow at the carotid bifurcation is postulated to play an important role in the genesis of atheromatous plaque. The presence of significant carotid stenosis in the absence of a bifurcation is an unexpected finding and raises the possibility that other local factors may also act as stimuli for plaque formation.

CASE REPORT

A 66-year-old white man was noted to have a right cervical systolic bruit during a routine physical examination. The medical history included hypertension, diabetes mellitus, and heavy tobacco use. Duplex ultrasound scanning results revealed complex plaque that caused a 50% to 79% stenosis of the distal right common carotid artery. The carotid bifurcation was not visualized, and the external carotid artery was presumed to be occluded. On the left side, the proximal internal carotid artery showed a 50% to 79% stenosis with healthy common and external carotid arteries. Both vertebral arteries were visualized and showed healthy antegrade flow.

Angiographic results of the right carotid artery (Fig 1) confirmed high-grade carotid stenosis. The carotid bifurcation and the external carotid artery on this side were not visualized. Distal to the area of stenosis, several vessels were noted to arise from the single carotid artery. To rule out any associated intracranial anomalies, magnetic resonance angiography was obtained (Fig 2). The results confirmed absence of the right carotid bifurcation and revealed healthy intracranial vasculature.

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Competition of interest: nil.

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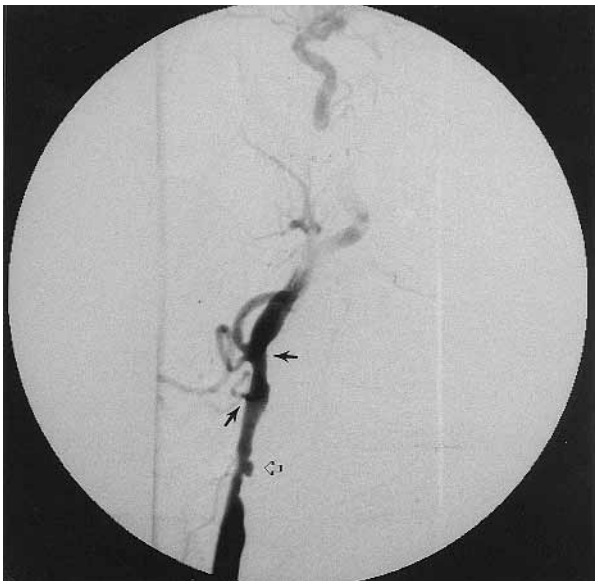

Fig 1. Right carotid arteriogram shows single carotid vessel without bifurcation. *White arrow,* ulcerated plaque at level of C4; *black arrows,* branches arising from single carotid vessel.

An elective carotid endarterectomy was performed. During exploration of the right neck, a single carotid vessel was identified (Fig 3). Branches that usually originate from the external carotid artery were found to arise from the single carotid vessel. An endarterectomy was performed with continuous electroencephalographic monitoring, and a patch angioplasty was used to close the arteriotomy. The patient recovered from the procedure and was discharged 2 days after surgery. Eight months later, the patient underwent an uneventful left carotid endarterectomy.

COMMENT

Congenital anomalies of the carotid artery circulation were first described in the 18th century from cadaveric dissections. More recently, because of the prevalence of atherosclerotic disease in the carotid area and the widespread use of angiographic techniques, further variations have been reported.[1]

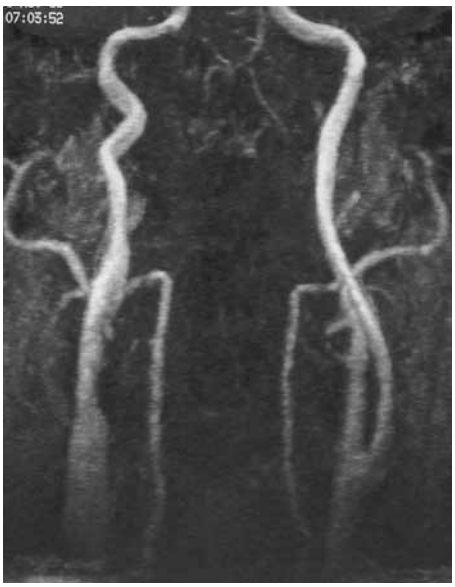

Fig 2. Magnetic resonance angiogram of neck shows healthy extracranial vasculature on left side and single carotid vessel on right, with significant stenosis just distal to expected level of bifurcation.

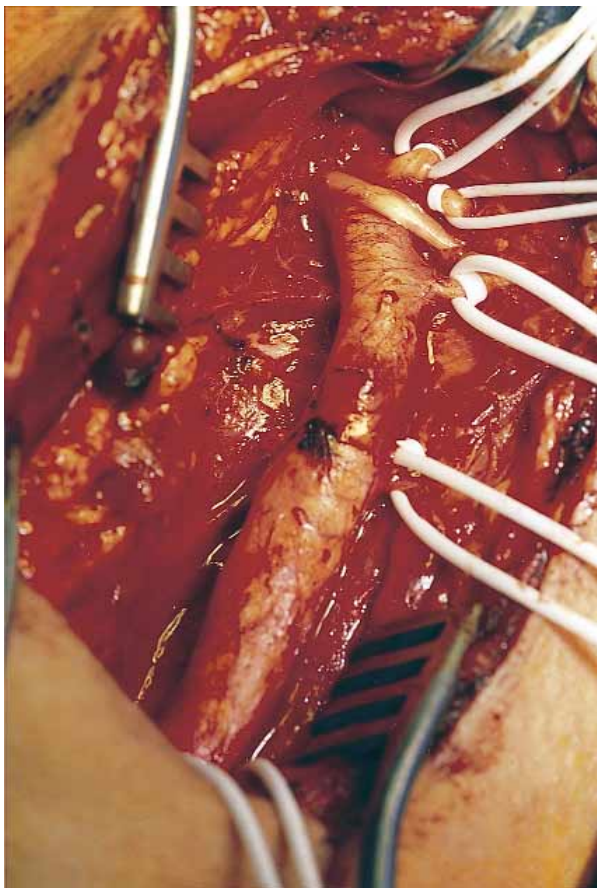

Fig 3. Intraoperative photograph shows absence of carotid bifurcation, branches arising from single carotid vessel (encircled by plastic loops), and hypoglossal nerve crossing the carotid artery.

Absence of the external carotid artery is, however, extremely rare. In 1821, Allan Barns described an internal carotid artery with branches that normally arise from the external carotid artery. An exhaustive search of the modern literature produced only four reports of this condition.[2-5] Possible embryologic explanations for this anomaly have previously been described.[1,4] Briefly, the facial lingual trunk, the middle meningeal artery, and the internal maxillary artery communicate with the dorsal aorta (which ultimately, along with the third aortic arch, will become the internal carotid artery) via the hyoidal artery. The hyoidal artery is believed to regress, and the previously mentioned branches are transferred to the external carotid artery system. As hypothesized by Lambiase et al,[4] lack of regression of the proximal hyoid artery with failure of transfer of its distal branches could explain this rare anomaly.

Important advances have been made in the understanding of the genesis of atherosclerosis. The "response to injury" hypothesis postulates that atheromatous plaque formation results from an excessive, inflammatory, and fibroproliferative response to various noxious stimuli to the endothelium and smooth muscle components of the artery wall.[6] Shear stress, the frictional drag force created by blood flow, is thought to play a pivotal role in this endothelial dysfunction. Shear stress stimulates the release of vasoactive substances, which causes alterations in gene expression, cell metabolism, and cell morphology.[7] Scale models of the carotid bifurcation have shown that early lesions occur in regions of flow separation, low wall shear stress, and departure from unidirectional flow.[8] Pulsatile hemodynamic models have correlated atheromatous plaque location with low oscillating shear stress.[9] In these models, the flow divider at the carotid bifurcation is the main anatomic structure responsible for the aforementioned hemodynamic changes.

Our case illustrates that significant carotid stenosis can occur in the absence of a carotid bifurcation. Furthermore, high-grade carotid stenoses have been found in the cases of absent external carotid arteries described by Lambiase et al[4] and Franklin et al.[5] In these two cases, the anatomic anomaly and the presence of significant stenosis were confirmed during carotid endarterectomy. Interestingly, carotid stenosis as the result of atherosclerotic plaque formation was also found in a case of absent common carotid artery reported by Halstuk and Littooy.[10] In this case, separate origins of the internal and external carotid arteries were noted, along with absence of a carotid bifurcation. In consideration of these reports together, symptomatic high-grade stenosis caused by atherosclerosis at the outer wall of the internal carotid artery was present in four of the six anomalous carotid arteries without a bifurcation or flow divider.

These findings suggest that the presence of a flow divider is not mandatory for the formation of atheromatous plaque at the level of the carotid bulb. Hemodynamic changes can possibly occur in the absence of a bifurcation when tortuosity or changes in vessel diameter are present. These hemodynamic alterations, the presence of baroreceptors at the sinus, and other local factors may, at least in part, be responsible for the predilection of plaque formation in this specific area of the vasculature.

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