ISSN 2005-7881
Journal of Neurosonology 3(1):20-23, 2011

CASE REPORT

Congenital Absence of the Internal Carotid Artery: Duplex Ultrasonographic Findings

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ABSTRACT

Absence of the internal carotid artery (ICA) is an uncommon disorder, occurring in <0.01% of the population. Very few of these cases have reported ultrasonographic findings. A 47-year-old woman with a history of hypertension presented to the hospital with a three-day history of headache. Color-coded duplex ultrasonography revealed a left common carotid artery (CCA) of diminished caliber terminating in the external carotid artery (ECA) with no remnants of a cervical ICA. Brain MRI showed absence of the bony carotid canal, cervical, and petrous portions of the ICA. The MR angiographic findings were consistent with the ultrasonographic findings. The left CCA terminates in the ECA with absence of the ICA. Filling of the left anterior circulation by way of the posterior and anterior communicating arteries is also seen. Color-coded duplex ultrasonography appears to be a sensitive and effective method for detecting absence of the ICA.

Journal of Neurosonology 3(1):20-23, 2011

Key Words: Agenesis, Internal carotid artery, Ultrasonography

Absence of the internal carotid artery (ICA) is an uncommon disorder, occurring in <0.01% of the population.[1] Approximately 100 cases have been reported and very few have had ultrasonographic findings described.[2,3] The ultrasonographic findings of absence of ICA, a rare congenital anomaly, are described in this report.

CASE

A 47-year-old woman with a history of hypertension presented to the hospital with a three-day history of headache. Results of the physical and neurological examinations were unremarkable. Color-coded duplex ultrasonography, performed to evaluate the carotid and vertebral arteries, showed a normal configuration on the right side. There was no significant stenotic flow or plaque formation observed in the right common carotid artery (CCA), external carotid artery (ECA), or ICA. However, the left ICA was not visualized. The left CCA was diminished caliber and terminated in the ECA with no remnants of a cervical ICA observed (Fig. 1) . The brain MRI showed absence of the bony carotid canal, cervical, and petrous portions of the ICA, suggesting agenesis rather than aplasia (Fig. 2).[1] The MR angiographic findings were consistent with the ultrasonographic findings. Imaging of the aortic arch and circle of Willis showed that the left CCA terminated in the ECA with absence of the ICA. Filling of the left anterior circulation by way of the posterior communicating artery (P-com) and anterior communicating artery (A-com) was seen (Fig. 2). There were no other intracranial abnormalities identified.

DISCUSSION

During the carotid ultrasonographic study, the vessel size, orientation, presence of branching vessels, and spectral waveforms

Received December 23, 2010 / Accepted May 26, 2011

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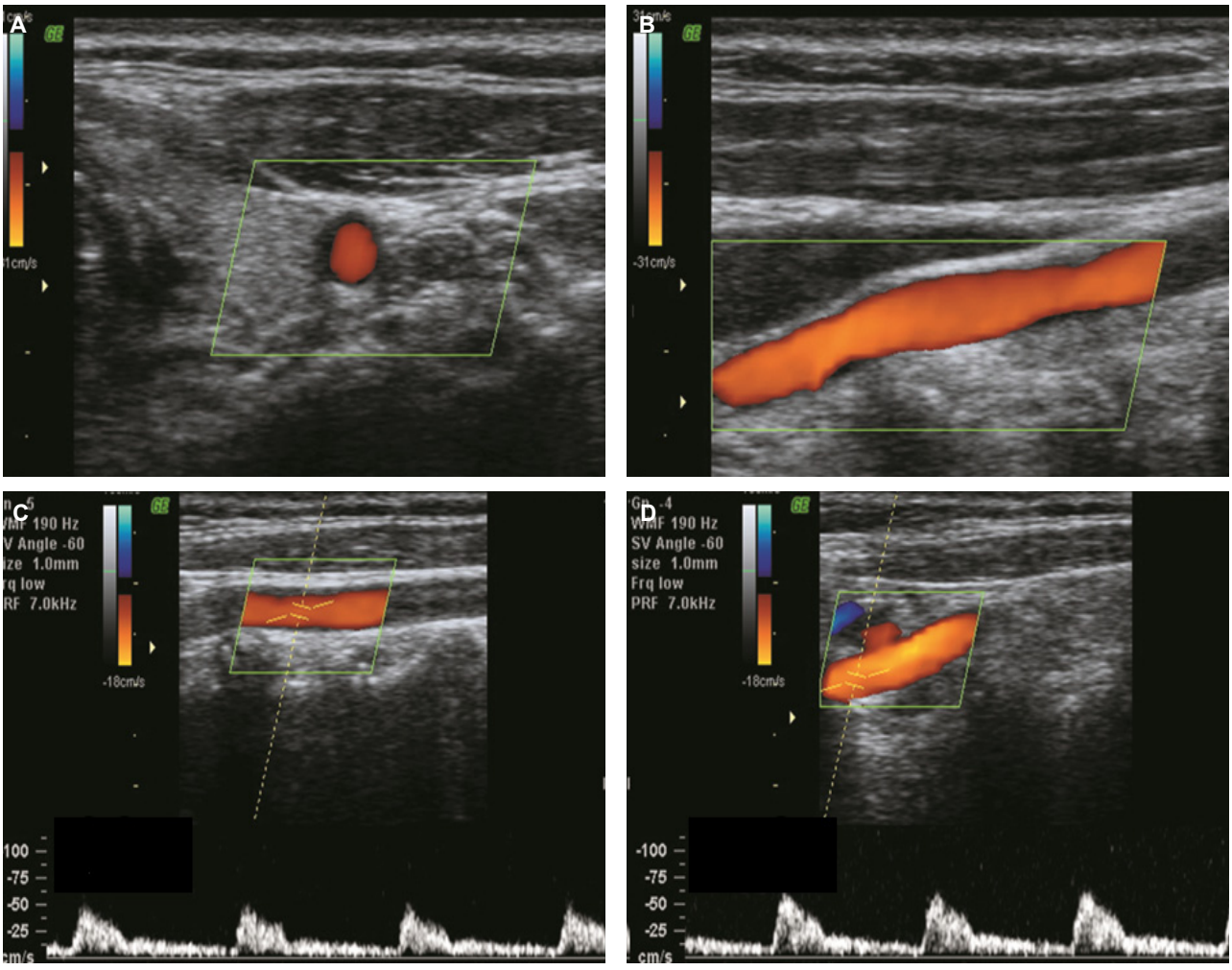

Fig. 1. (A) Color-coded duplex sonography shows the absence of the left internal carotid artery (ICA). (B) The common carotid artery (CCA) of diminished caliber terminates in the external carotid artery (ECA). (C) The CCA demonstrates high-resistance spectral waveforms with sharp systolic peaks and little diastolic flow. (D) The left ECA waveform demonstrates high-resistance characteristics and is indistinguishable from the CCA waveform.

are helpful for differentiating the ICA from the ECA. In this patient, the flow characteristics of the left carotid artery showed a high-resistance flow pattern with low diastolic flow and peaked waveforms. These findings are consistent with an ECA and suggested that the CCA terminated in the ECA. There were no remnants of a cervical ICA observed.

Three types of intracranial vascular anomalies are commonly associated with an absent ICA.[3-5] 1) aneurysms related to the circle of Willis, 2) arterial anomalies including abnormal collateral channels, and 3) dilated vascular channels. These may have clinical significance for carotid surgery, thromboembolic disease, and the prevention of subarachnoid hemorrhage.[2]

There is no complete explanation for agenesis of the ICA. Normally, the ICA is derived from portions of the first and third aortic arches and the paired dorsal aorta. These components unite by way of a proximal process from the capillary network in the walls of the forebrain and midbrain. This union forms a primitive ICA that can be identified around the fourth week of development. Agenesis of the ICA depends on the abnormal regression of the first and third aortic arches.[1,3] It is plausible that mechanical and hemodynamic stresses to the developing embryo may cause this anomaly. There are three types of collateral circulation in cases of unilateral absence of the ICA.[3,4,6] 1) the most common, "fetal" type, which supplies the anterior cerebral artery (ACA) of the affected side via the A-com and middle cerebral artery (MCA) through an enlarged P-com, 2) "adult"

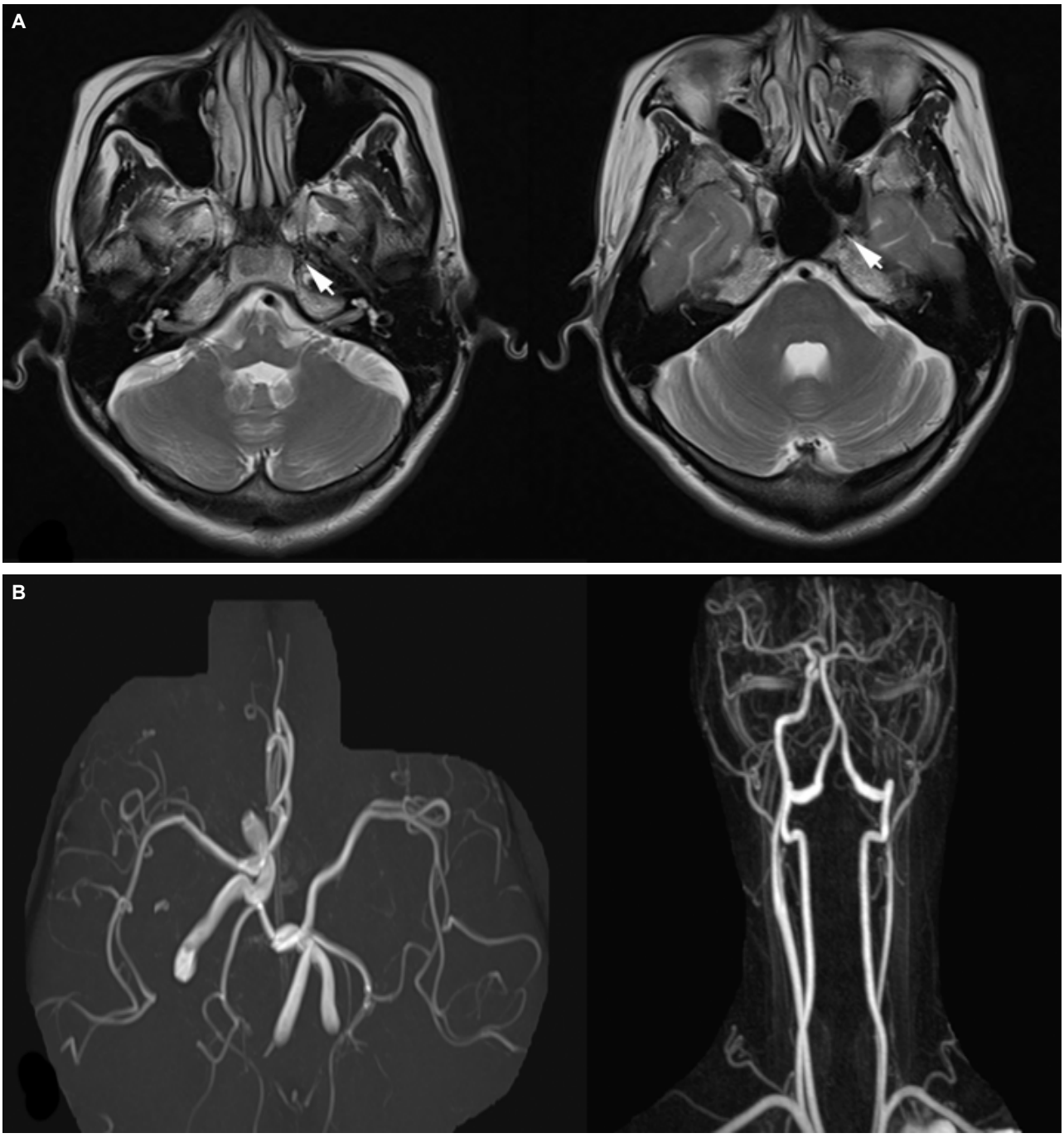

Fig. 2. (A) Brain MRI shows absence of the bony carotid canal and petrous portions of the ICA (arrow), suggesting ICA agenesis. (B) MR angiography reveals the left CCA terminating into the ECA with absence of the ICA. Filling of the left anterior circulation by way of the posterior communicating artery and anterior communicating artery is also seen.

type which supplies the ACA and MCA of the affected side through a patent A-com, and 3) the least common type, which supplies the ACA and MCA of the affected side from the contralateral ICA through anastomotic vessels.

The findings of the current case add to the growing understanding of the embryologic development of the carotid arteries. In addition, color-coded duplex ultrasonography appears to be a sensitive and effective method for detecting absence of the ICA.

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