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ANATOMIC VARIATIONS

Hypoplasia of the internal carotid artery with associated fenestration and extremely long P1 segment of the ipsilateral posterior cerebral artery diagnosed by MR angiography

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Abstract
We present what we believe is the first report of a patient with unilateral hypoplasia of the internal carotid artery with associated ipsilateral anomalous posterior cerebral artery, extremely long and fenestrated P1 segment that was diagnosed on magnetic resonance (MR) angiography. Careful review of MR angiographic images is important to detect rare arterial variations, and partial maximum-intensity-projection images aid their identification on MR angiography.

Keywords Cerebral arterial variation · Fenestration · Internal carotid artery · Magnetic resonance angiography · Posterior cerebral artery

Introduction

Congenital hypoplasia of the internal carotid artery (ICA) is rare and designated as a Type E anomaly according to Lie's classification [3]. Variations of the posterior cerebral artery (PCA) apart from aplasia of the P1 segment of the PCA are rare, and fenestrations of the PCA have a reported prevalence of only 0.34% [8]. We present what we believe is the first report of a case of hypoplasia of the left ICA with associated unilateral fenestration of the P1 segment and an extremely long P1 segment of the PCA that was diagnosed by MR angiography.

Case report

A 37-year-old woman who experienced multiple transient ischemic attacks (TIAs) transferred from a general hospital to our institution for further evaluation and possible treatment of stenosis of the left ICA. Ten years previously, she had undergone evaluation of subarachnoid hemorrhage of unknown etiology that revealed extreme narrowing of the left ICA, but those images were unavailable. With conservative treatment at that time, she recovered completely from the subarachnoid hemorrhage, but continued to experience TIAs (manifested as mild right hemiparesis and mild speech disturbance) several times per year. She did not have underlined pathology such as cardiac arrhythmia, hypertension, or hyperlipidemia. There was no family history of the ischemic cerebrovascular diseases.

We performed cranial MR imaging and MR angiography using a 3-Tesla scanner (Achieva 3.0T TX Quasar Dual, Philips Medical Systems, Best, The Netherlands). MR imaging showed multiple small infarctions in the left centrum semiovale, indicating borderzone infarctions (Fig. 1). MR angiography was obtained using a standard 3-dimensional time-of-flight (3D-TOF) protocol. Maximum-intensity-projection (MIP) MR angiographic images showed that the left ICA was extremely narrow (Fig. 2), and partial MIP images of the left carotid system (Fig. 3) clearly depicted the ICA anomaly. The ascending pharyngeal artery was markedly dilated due to collateral blood flow, and the left posterior communicating artery (PCoA) was also dilated. Partial MIP images of the vertebrobasilar system (Fig. 4) clearly showed fenestration of an extremely long P1 segment of the left PCA. The artery's redundant course prevented measurement

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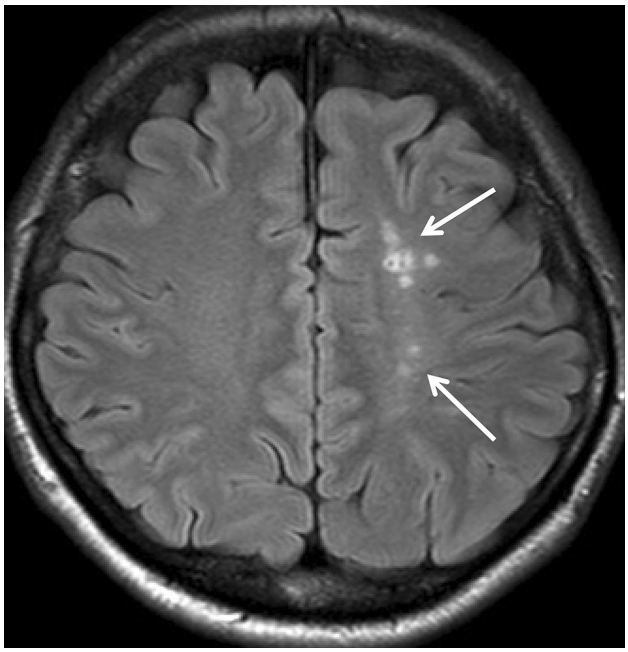

Fig. 1 Magnetic resonance (MR) imaging obtained using a 3-Tesla scanner. Fluid-attenuated inversion recovery (FLAIR) image of the centrum semiovale shows multiple tiny infarctions in the left frontoparietal white matter, indicating borderzone infarctions in the left cerebral hemisphere (arrows)

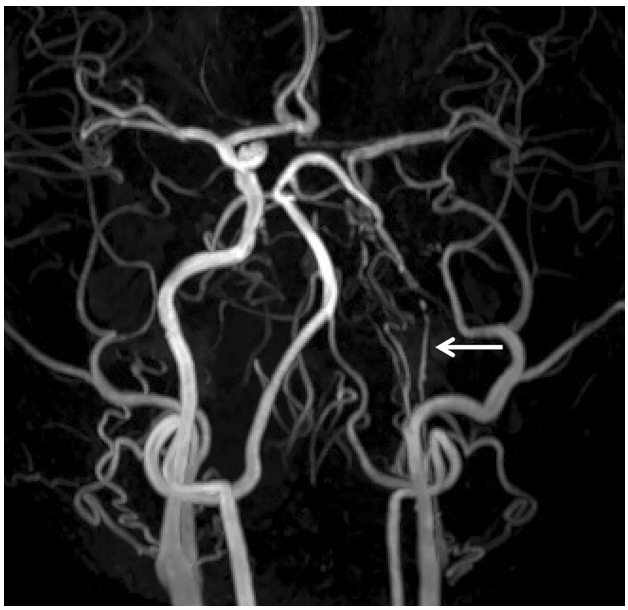

Fig. 2 Antero-posterior (AP) projection of magnetic resonance (MR) angiography shows an extremely narrow left internal carotid artery (ICA, arrow). The diameter of the left cervical ICA is about 1 mm. In contrast, the diameter of the right cervical ICA is about 3.5 mm

of the exact length of the P1 on MR angiography, but we estimated its length as about 30 mm.

The depiction of the left carotid canal in the petrous canal as narrow in computed tomographic (CT) bone images of the skull base (Fig. 5) led us to consider our patient's narrow ICA to represent congenital hypoplasia rather than the manifestation of an acquired lesion.

Discussion

Congenital ICA aplasia/hypoplasia

Lie [3] classified six types of congenital aplasia and hypoplasia of the ICA. Hypoplasia is categorized as Type E. In Type A, which is most prevalent, the posterior communicating artery supplies blood in the ipsilateral middle cerebral artery. In Types B and D, collateral blood flow comes via the anterior communicating artery (Type B) and from the cavernous segment of the contralateral ICA (Type D). Type C represents aplasia of the bilateral ICAs. We previously reported a case of aplasia of the ICA that was unclassified and supplied by the supraclinoid segment of the contralateral ICA [7]. Type F consists of a small arterial network, described as a "rete mirabile", at the extracranial segment.

A narrow ICA is much more commonly acquired than the result of congenital ICA hypoplasia, and the two types are differentiated based on findings of CT of the skull base. The carotid canal appears normal in acquired cases and narrow in congenital hypoplasia (Fig. 5).

Congenital aplasia or hypoplasia of the ICA usually demonstrates no ischemic brain lesions, because collateral blood flow is well developed. However, Nardone and associates [5] reported the cases of two patients with hypoplasia of the ICA who suffered TIAs like those of our patient. In such cases, severe systemic hypotension may cause hemodynamic infarctions in the borderzone of the ipsilateral cerebral hemisphere, as shown in Fig. 1. To prevent future ischemic cerebrovascular events, our patient should be controlled against systemic hypotension and dehydration. If TIAs frequently occur, bypass surgery such as the superficial temporal artery–middle cerebral artery (STA–MCA) anastomosis may be considered.

PCA fenestration and extremely long P1 segment

Variations of the PCA are rare. The vessel is divided into four segments. Its P1 segment is proximal to the junction with the PCoA and described as fetal type when the segment is aplastic or hypoplastic. This most common variation of the artery has a reported prevalence of 10–40% [1]. We recently reported five types of PCA variation diagnosed by MR angiography: fenestration, early bifurcation, and complete duplication of the PCA and accessory (hyperplastic anterior choroidal artery) and replaced PCA [8]. According

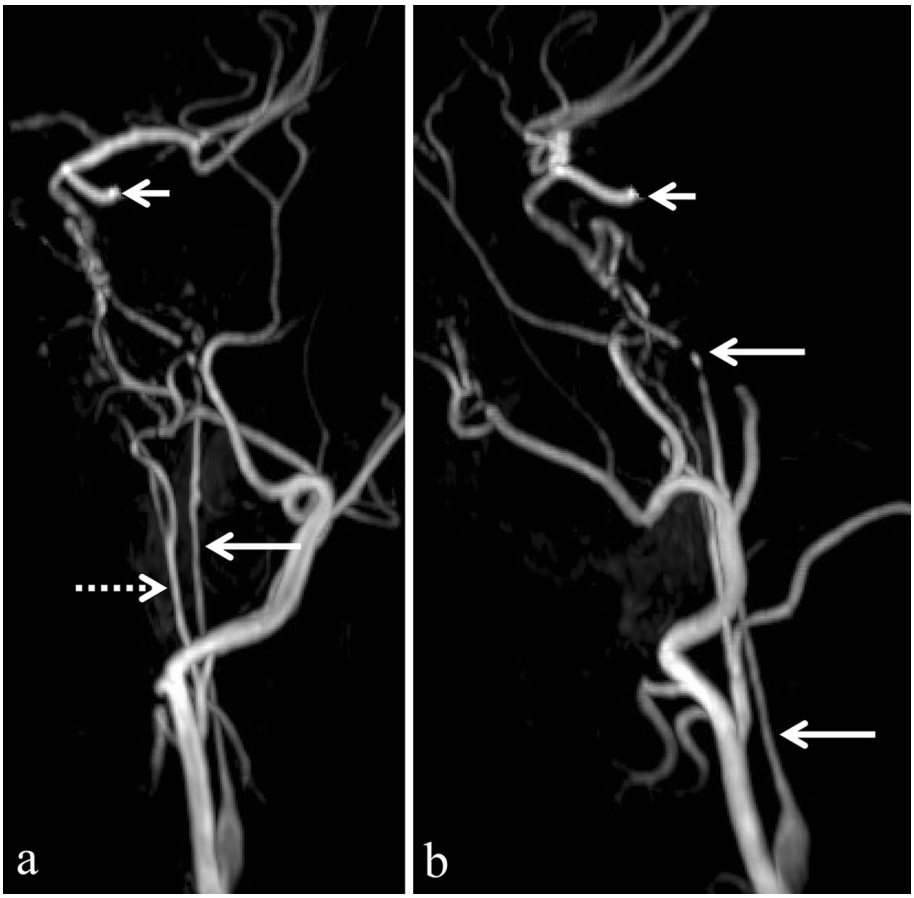

Fig. 3 Antero-posterior (AP) (**a**) and lateral (**b**) partial maximum-intensity-projection (MIP) images of the left carotid system clearly show a narrow left internal carotid artery (ICA, long arrows), dilated left ascending pharyngeal artery (dotted arrow), and large left posterior communicating artery (PCoA, short arrows)

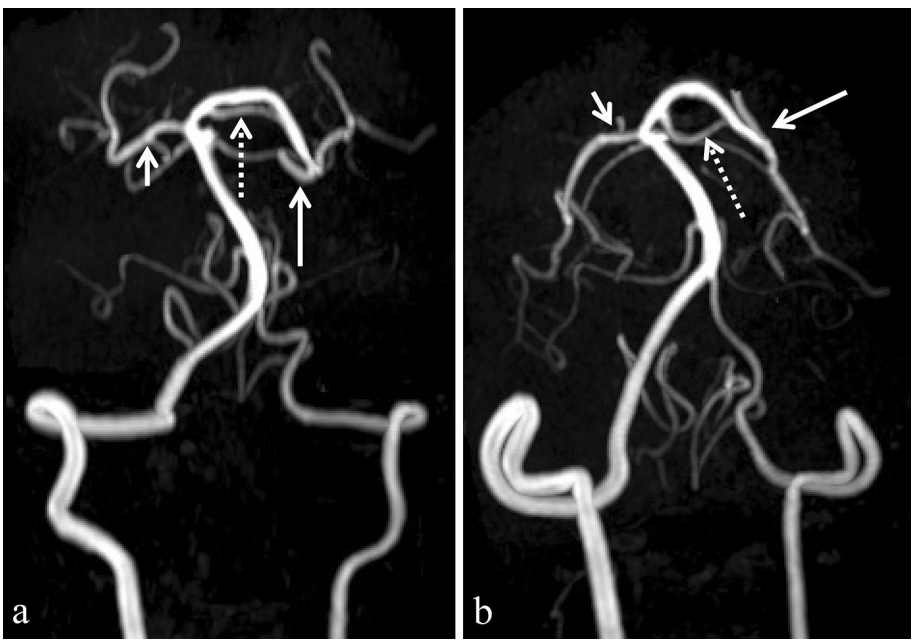

Fig. 4 Two different antero-posterior (AP) projections (**a, b**) of partial maximum-intensity-projection (MIP) images of the vertebrobasilar system clearly show fenestration and an extremely long P1 segment of the left posterior cerebral artery (PCA). The long arrows indicate the junction of the left posterior communicating artery (PCoA) and PCA, indicating the extremely long P1 segment. The dotted arrows indicate fenestration of the left P1. The short arrows indicate the normally positioned junction of the right PCoA and PCA

to our experience, the prevalence of fenestration of the PCA is 0.34%, and it is primarily observed at the P1 segment. We have also reported a case of PCA of duplicate origin with an extremely long P1 segment (22 mm) depicted by MR angiography [4].

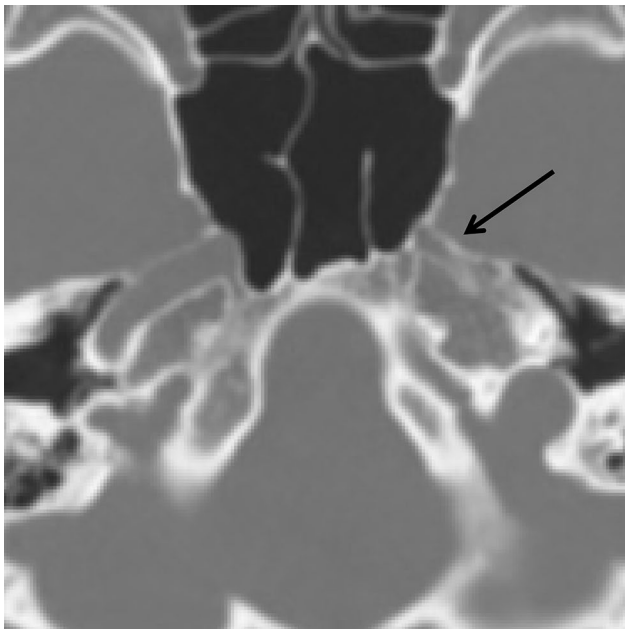

Fig. 5 Computed tomography (CT) bone image of the skull base shows an extremely narrow left carotid canal (arrow)

In examination of adult cadavers, Zeal and Rhoton reported the length of the P1 segment to range between 3.5 and 17 mm [10], whereas that of our patient was extremely long, measuring approximately 30 mm. We previously reported two cases with long P1 segment and extremely long PCoA [9], but the present case did not have a long PCoA.

Variations in the PCA are formed during embryological development. In Padget Stage 2 [6] (embryo of 31 days, 5–6 mm in length), the primitive ICA supplies the entire cerebral region, including the posterior areas through the PCoA, and the PCA is only a branch of its caudal division. The two primitive neural arteries run parallel to each other on the ventral surface of the pons, pass through a plexiform disposition of vessels, and later form the basal artery (BA). In Padget Stage 3 (33 days, 7–12 mm in length), the PCoA arises as the vertebral arteries connecting to the BA and posterior circulation, and the PCA develops fully. Failure of normal arterial regression or fusion during this course may lead to the fenestration of the P1 segment. The textbook description of Lasjaunias, Berenstein, and ter Brugge [2] indicates that the PCoA with the P1 constitutes the caudal division of the ICA with its diencephalo-mesencephalic territory (which includes the cerebellum). It extends to the upper postsegmental portion of the BA (distal to the trigeminal remnant) and influences the pattern of the superior cerebellar artery. The convergent nature or fusion of the BA at the mesencephalic region creates a large range of variations.

Conclusion

We report an extremely rare case of hypoplasia of the ICA with associated fenestration and an extremely long P1 segment of the ipsilateral PCA that was diagnosed by MR angiography. Although we could find no similar case in the literature, these two different variations of the cerebral artery may be related.

Careful review of MR angiographic images is important to detect rare arterial variations, and partial MIP images are useful for their identification on MR angiography.

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Author contributions AU carried out the study design and drafting of the manuscript. AU, TE, and HK performed data acquisition and made a critical review of the manuscript. All authors have read and approved the final manuscript.

Compliance with ethical standards

Conflict of interest We declare no conflict of interest.

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