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Congenital absence of the internal carotid artery diagnosed during investigation of trigeminal neuralgia

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Abstract Congenital absence of the unilateral internal carotid artery (ICA) was found in a patient during MR imaging examination for right trigeminal neuralgia. Magnetic resonance angiography showed complete absence of the right ICA and a large tortuous basilar artery (BA). The source images revealed a deformed right trigeminal nerve resulting from compression by the BA. Computed tomography of the skull base showed absence of the right carotid canal, suggesting agenesis of the right ICA. Longstanding hemodynamic stress may have caused the BA to become extremely tortuous, resulting in the trigeminal neuralgia.

Keywords Absence of internal carotid artery · Tortuous basilar artery · Trigeminal neuralgia · Magnetic resonance angiography

Introduction

Agenesis, aplasia, and hypoplasia of the internal carotid artery (ICA) are rare anomalies [1, 2, 3, 4, 5, 6, 7, 8, 9, 10, 11, 12, 13, 14, 15, 16, 17, 18, 19, 20]. When the ICA is congenitally absent, collateral circulation develops through the circle of Willis from the basilar artery (BA) and the opposite ICA to supply the involved hemisphere. Therefore, neurologic deficits are few; however, these anomalies are frequently associated with cerebral aneurysm [3, 4, 6, 7, 12]. They may be associated with neurovascular compression syndromes such as oculomotor nerve palsy [5], trigeminal neuralgia [13], and spasmodic torticollis [10] in the presence of tortuous vertebrobasilar systems. We treated a patient in whom unilateral absence of the ICA was found incidentally during MR imaging investigation of trigeminal neuralgia.

Case report

A 62-year-old man had a 1-year history of sharp, severe right cheek pain. The trigger zone was at the right nasolabial fold, and the clinical diagnosis was typical trigeminal neuralgia. No neurological deficits were present. He had mild hypertension (140/90 mmHg) and hypercholesterolemia (total cholesterol, 291 mg/dl; normal 130–220 mg/dl). Magnetic resonance imaging and MR angiography were performed to evaluate the trigeminal neuralgia. Conventional MR imaging showed neither cerebellopontine angle tumors nor brainstem lesions. Nonspecific small white matter lesions were the only cerebral abnormalities detected. Magnetic resonance angiography using the three-dimensional time of flight (TOF) technique revealed an extremely tortuous vertebrobasilar system with no detection of the right ICA. The bilateral anterior cerebral arteries were fed by the left ICA. A dilated but faintly visible right posterior communicating artery was identified, but the right middle cerebral artery was not (Fig. 1a–c). The dilated right posterior communicating artery and normal right middle cerebral artery were identified, however, on conventional T2-weighted MR images. The source MR angiography images revealed lateral displacement of the right trigeminal nerve because of compression by both the tortuous BA and right anterior inferior cerebellar artery (Fig. 1c). Subsequent CT of the skull base revealed absence of the right carotid canal, indicative of congenital absence (probably agenesis) of the right ICA (Fig. 2).

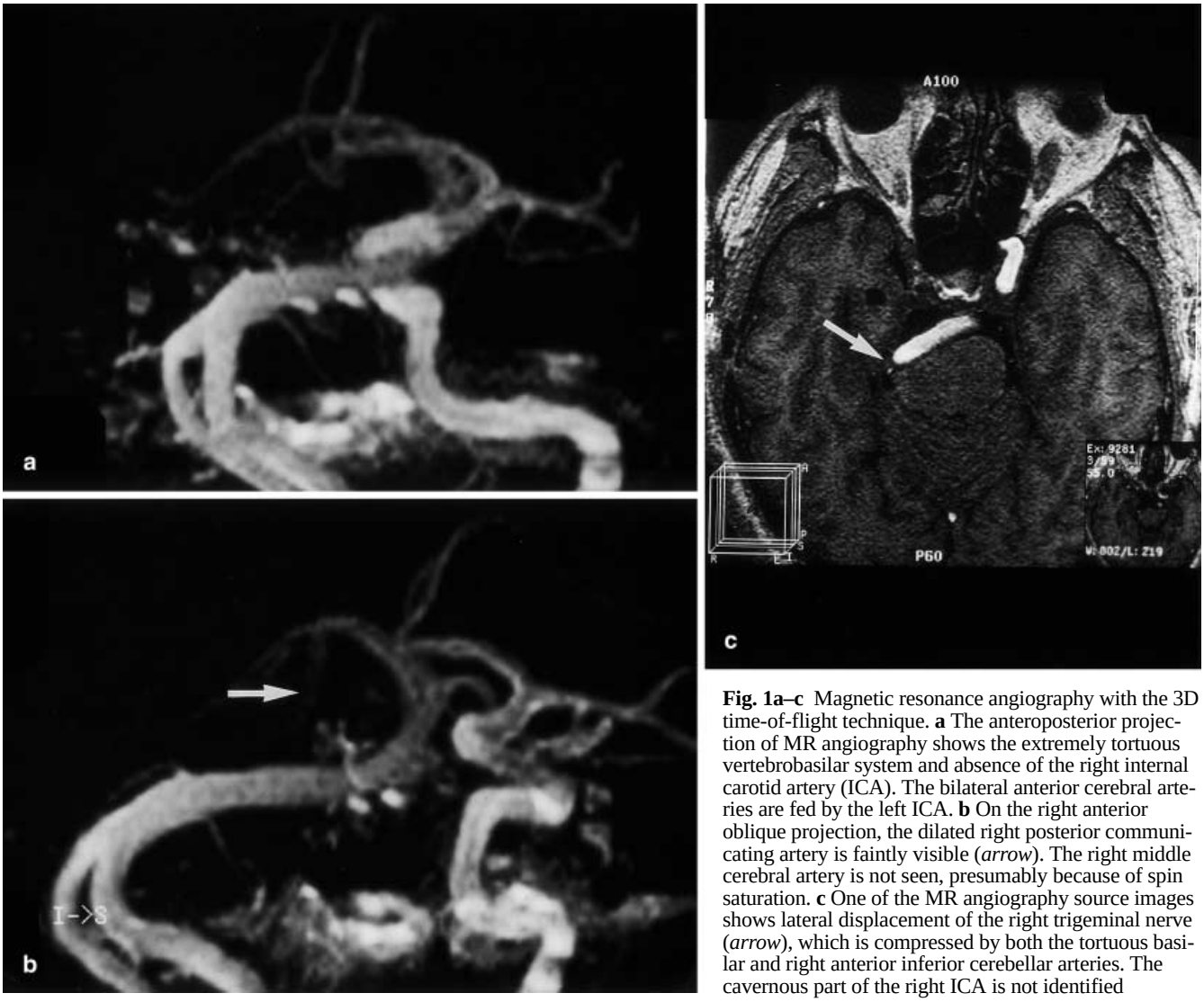

Fig. 1a–c Magnetic resonance angiography with the 3D time-of-flight technique. **a** The anteroposterior projection of MR angiography shows the extremely tortuous vertebrobasilar system and absence of the right internal carotid artery (ICA). The bilateral anterior cerebral arteries are fed by the left ICA. **b** On the right anterior oblique projection, the dilated right posterior communicating artery is faintly visible (*arrow*). The right middle cerebral artery is not seen, presumably because of spin saturation. **c** One of the MR angiography source images shows lateral displacement of the right trigeminal nerve (*arrow*), which is compressed by both the tortuous basilar and right anterior inferior cerebellar arteries. The cavernous part of the right ICA is not identified

The patient was treated conservatively with carbamazepine at a daily dose of 300 mg and infraorbital nerve block, which was effective. After this procedure, complete pain relief was attained.

Discussion

Agenesis involves complete developmental failure of an organ and its primordium. Aplasia refers to the defective development of an organ, although its anlage presumably existed at some time [6]. Since the carotid canal develops secondary to the presence of a fetal ICA, the CT finding of no carotid canal provides evidence for true agenesis rather than aplasia [18]. In the present case, since our patient had no carotid canal according to CT findings, we diagnosed him as having agenesis of the ICA. But since selective cerebral angiography was not performed, we cannot completely rule out the presence of a hypoplastic cervical ICA; therefore, agenesis, aplasia, and some types of the hypoplasia of the ICA may be indistinguishable from each other on CT and MR images. For this reason, we use the term congenital absence of the ICA in this paper [2, 6, 20].

Agenesis of the ICA is a rare anomaly for which the reported incidence is 0.01% [4]. Agenesis of the bilateral ICA is extremely rare [1, 14, 15, 18]. According to Lie [1], unilateral agenesis/aplasia of the ICA is of three types: (a) fetal type in which a dilated posterior communicating artery supplies the middle cerebral artery and the anterior cerebral artery is fed via the anterior communicating artery; (b) adult type in which both the anterior and middle cerebral arteries are fed via the anterior communicating artery; and (c) third type in which the distal part of the ICA is present and is supplied via the intercavernous anastomosis [6, 7, 8, 9, 16]. Our present

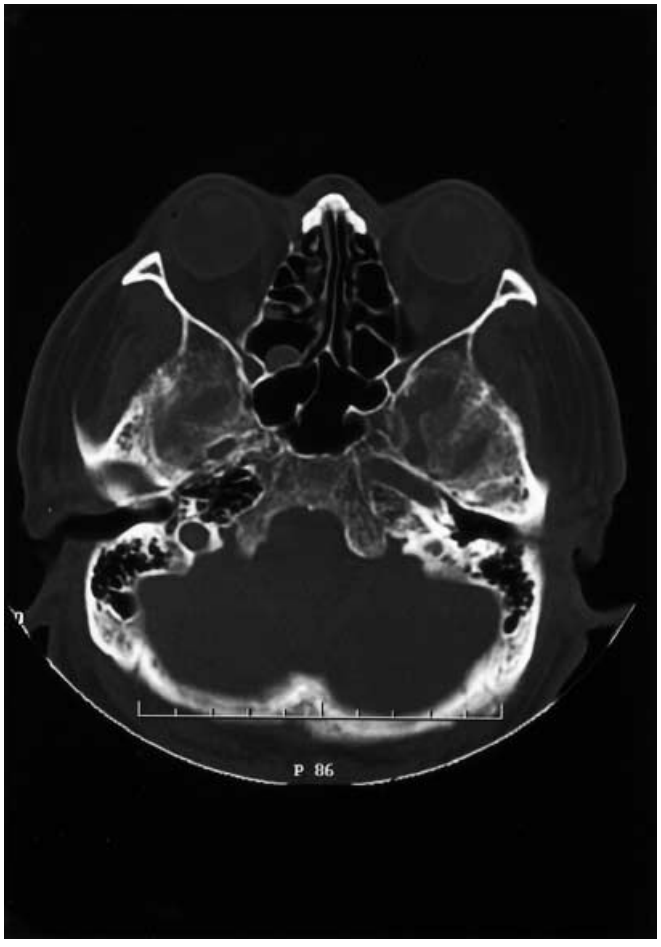

Fig. 2 Bone image of skull base. Computed tomography of the skull base reveals absence of the right carotid canal, suggestive of right ICA agenesis

case was of the fetal type. The primitive ICAs are derived from the first and third aortic arches and the dorsal aortae in the embryo of 3-mm crown-rump length [1]. The third aortic arches play an important role in forming the ICA, and regression or hypoplasia of these arches and dorsal aortae may lead to failure of full development of the ICA [10].

Agenesis, aplasia, and hypoplasia of the ICA are frequently associated with cerebral aneurysms of the circle of Willis, presumably because of increased circulatory stress that derives from altered hemodynamics. Incidence of the cerebral aneurysms has been reported to be 25–35% [3, 4, 6, 7, 8, 12]. The fetal-type anomalies may be also associated with tortuous and ectatic vertebrobasilar systems that lead to the development of neurovascular compression syndromes such as oculomotor nerve palsy [5], trigeminal neuralgia [13], and spasmodic torticollis [10]. Longstanding hemodynamic stress to the vertebrobasilar system causes dilatation and elongation of the vertebral arteries and BAs, resulting in tortuosity of the vertebrobasilar system. Our present patient had additional risk factors for atherosclerosis, such as hypertension and hypercholesterolemia.

Congenital absence of the ICA can be associated with various other abnormalities such as cerebral hemiatrophy [2, 4], epilepsy [8], facial hemangioma and dysplasia of cerebral cortex [11], psychomotor developmental delay with periventricular high-intensity areas [15], hypopituitarism [16], ear malformation and facial palsy [17], congenital Horner's syndrome [19], and persistent trigeminal artery and other vascular anomalies [13, 20]. In contrast to findings in patients with acquired ICA occlusion, ipsilateral cerebral infarctions are not frequently observed in patients with congenital absence of the ICA, because collateral circulation from the contralateral ICA or the vertebrobasilar system, or both, is usually well developed.

The main causes of trigeminal neuralgia are compression of the root entry zone of the trigeminal nerve by vessels and cerebellopontine angle tumors such as epidermoid, meningioma, and schwannoma [21]. Multiple sclerosis plaque in the brainstem also causes trigeminal neuralgia [22]. In most cases, the compressing vessel is the superior cerebellar artery, and the anterior inferior cerebellar artery is the second most common [23]. Tortuous vertebrobasilar arteries also may cause trigeminal neuralgia [24]. In a relatively large study of trigeminal neuralgia (1404 consecutive patients), only 2% of cases involved vascular compression of the vertebral arteries or BAs [25]. In our patient, both the tortuous BA and the right anterior inferior cerebellar artery pressed upon the right trigeminal nerve. To our knowledge, this is the second report of congenital absence of the ICA diagnosed during investigation of trigeminal neuralgia [13]. We stress that the anterior circulation should be carefully evaluated during the interpretation of MR images of posterior fossa neurovascular compression syndromes.

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