Case Report

A Case of Classical Trigeminal Neuralgia Associated with Bilateral Agenesis of the Internal Carotid Arteries

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Abstract

We describe a case of trigeminal neuralgia caused by compensatory enlargement of the basilar artery in a patient with bilateral agenesis of the internal carotid arteries.

A 46-year-old man experienced sharp, stabbing pain in the left face when washing his face. In imaging examinations, magnetic resonance imaging (MRI) revealed displacement of the left trigeminal nerve due to basilar artery enlargement and leftward deviation, but no brain tumors or other abnormalities. The left internal carotid artery was not visible on magnetic resonance angiography (MRA), but marked enlargement of bilateral vertebral and basilar arteries was seen, along with considerable leftward deviation of the basilar artery. The anterior and middle cerebral arteries constituting the anterior circulation were bilaterally connected with the posterior communicating artery, which was enlarged. Bilateral internal carotid arteries were vestigially interrupted from the bifurcation of the common carotid artery. No carotid canals were apparent on either side on computed tomography (CT). For his symptoms, we managed his pain using carbamazepine, and instructed to follow a low-salt, low-fat diet and to avoid excessive alcohol consumption.

Our case report suggest that imaging examination (e.g. CT, MRA and MRI) are essential when investigating trigeminal neuralgia, in order to visualize the head and neck region and identify any causal vessels and confirm the presence or absence of tumors. In addition, patients with abnormal hemodynamic status such as agenesis of an internal carotid artery or megadolichobasilar anomaly should undergo regular follow-up due to the high risk of cerebral hemorrhage and cerebral infarction.

Keywords :

trigeminal neuralgia;
agenesis of the internal carotid artery;
vertebrobasilar artery;
magnetic resonance angiography;
magnetic resonance imaging

Introduction

Agenesis of the internal carotid artery is a rare disorder with a reported incidence of 0.01% of live human births (1). To date, three cases involving trigeminal neuralgia and agenesis of the internal carotid artery have been reported, but all involved classical trigeminal neuralgia associated with unilateral agenesis of an internal carotid artery (2-4). To the best of our knowledge, no reports have described trigeminal neuralgia with bilateral agenesis of the internal

carotid arteries. We describe a case of trigeminal neuralgia caused by compensatory enlargement of the basilar artery in a patient with bilateral agenesis of the internal carotid arteries.

Case report

A 46-year-old man experienced sharp, stabbing pain in the left face when washing his face. Symptoms resolved after carbamazepine (CBZ) was prescribed at 400 mg/day at another hospital, so he discontinued the medication. Symptoms recurred and he took the remaining medication, but the effects were insufficient and he presented to Nihon

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Fig.1 Basilar artery and left trigeminal nerve in the axial plane on magnetic resonance imaging. Black arrow indicates basilar artery. White arrow indicates left trigeminal nerve.

Fig.3 Agenesis of bilateral internal carotid arteries in the coronal plane on magnetic resonance angiography. Arrows indicate agenesis of bilateral internal carotid arteries.

Fig.2 Basilar and vertebral arteries in the coronal plane on magnetic resonance angiography. Black arrow indicates basilar artery. White arrow indicates vertebral artery.

Fig.4 Anterior cerebral, middle cerebral, and posterior communicating arteries in the axial plane on magnetic resonance angiography. Black arrow indicates anterior cerebral artery. White arrow indicates middle cerebral artery. Arrowhead indicates posterior communicating artery.

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Palpation revealed a trigger zone at the left nasolabial fold. When the trigger zone was touched, he experienced shooting pain persisting several seconds. No hypesthesia was present in the left trigeminal area. Systemic history included hyperuricemia, hyperlipidemia, and hypertension. At our hospital, the head and neck region were imaged using magnetic resonance imaging (MRI), computed tomography (CT), and magnetic resonance angiography (MRA) to investigate potential causes of left facial pain. MRI revealed displacement of the left trigeminal nerve due to basilar artery enlargement and leftward deviation, but no brain tumors or other abnormalities (Fig.1). The left internal carotid artery was not visible on MRA, but marked

enlargement of bilateral vertebral and basilar arteries was seen, along with considerable leftward deviation of the basilar artery. The anterior and middle cerebral arteries constituting the anterior circulation were bilaterally connected with the posterior communicating artery, which was enlarged. Bilateral internal carotid arteries were vestigially interrupted from the bifurcation of the common carotid artery (Figs. 2-4). No carotid canals were apparent on either side on CT (Fig. 5).

Initial treatment comprised CBZ at 600 mg/day. This dose was tapered as symptoms started to improve, reaching 100 mg/day at 6 months after starting treatment. At 12 months after the start of treatment, this lower dose proved insufficient for pain control, so the prescription was increased to 200 mg/day. At present, the patient is taking

Fig.5 Agenesis of bilateral carotid canals in the axial plane on computed tomography. Arrows indicate agenesis of bilateral carotid canals.

CBZ at 600 mg/day and symptoms remain adequately controlled.

The patient showed hypertension and markedly elevated levels of urea nitrogen, uric acid, and triglycerides. Candesartan at 4 mg/day, pitavastatin at 1 mg/day, and allopurinol at 100 mg/day were prescribed for suspected hyperlipidemia, hyperuricemia, and hypertension, respectively, and he was instructed to follow a low-salt, low-fat diet and to avoid excessive alcohol consumption.

Discussion

The International Classification of Headache Disorders (ICHD) -II by the International Headache Society divides trigeminal neuralgia into two types: classical and symptomatic. Classical trigeminal neuralgia involves classic pain symptoms either caused by neurovascular compression or of unknown etiology. Symptomatic trigeminal neuralgia is caused by structural lesions such as meningioma, schwannoma, or other tumors. The most common cause of trigeminal neuralgia is neurovascular compression (72.0-90.8%), followed by structural lesions (5.1-10.7%). The etiology is unknown in 5.3-13.2% of cases(5-8). According to the ICHD-II, classical trigeminal neuralgia is defined by pain that: is sudden; persists for between 1 s and 2 min at one more points in the area of innervation of the trigeminal nerve; is sharp, stabbing, and of severe intensity; originates from a trigger point or is caused by trigger factors; follows a particular pattern of onset in each individual; and occurs in the absence of obvious clinical neurological deficits. Conversely, symptomatic trigeminal neuralgia is characterized by paresthesia of the trigeminal nerve area. Classical

trigeminal neuralgia is caused by cerebrovascular compression of the root entry zone, where the myelin sheath at the cerebellopontine angle changes from central to peripheral. The root entry zone is vulnerable to mechanical compression, so persistent neurovascular compression can cause demyelination. This allows action potentials in the trigeminal nerve to transmit incorrect information to distal pain nerves, resulting in pain(9). The present patient displayed characteristics of classical trigeminal neuralgia, but the blood vessel involved was the enlarged and curved basilar artery. The superior cerebellar artery is the blood vessel most often responsible for classical trigeminal neuralgia, followed by the anterior inferior cerebellar artery (10). Few cases involve comparatively large cerebral blood vessels such as the vertebrobasilar artery, as in the present patient. In a review of 1404 cases of classical trigeminal neuralgia, only 31 (approximately 2%) involved vertebrobasilar arterial compression of the trigeminal nerve(11). The present findings suggest that use of MRI, MRA, and CT to investigate trigeminal neuralgia may be effective for identifying the presence of brain tumors and blood vessels compressing the root entry zone of the trigeminal nerve.

Trigeminal nerve compression caused by dilatation of the vertebrobasilar artery trunk has been observed in patients with both trigeminal neuralgia and agenesis of the internal carotid artery (Table 1) (2-4). The elongated, tortuous, and dilated basilar artery and resulting hemodynamic state found in the present patient is known as a megadolichobasilar anomaly(12). Patients often become aware of a megadolichobasilar anomaly due to symptoms arising from compression of the cranial nerve and brainstem by the abnormally enlarged, curved basilar artery. Features of megadolichobasilar anomaly include congenital abnormalities and hemodynamic changes due to atherosclerosis, cerebral arteriovenous malformation, and cerebral aneurysm. The features of megadolichobasilar anomaly in the present patient comprised hemodynamic changes due to agenesis of the internal carotid arteries and arteriosclerosis due to hyperlipidemia and hypertension. Careful follow-up is required due to the risks of cerebral hemorrhage and cerebral infarction caused by further enlargement of the same area with age, hyperlipidemia, and poor blood pressure control.

Agenesis of the internal carotid artery is rare and few reports have described bilateral agenesis(13-27). Possible causes of agenesis of the internal carotid artery include

Table 1 Summary of 4 cases of internal carotid artery agenesis associated with trigeminal neuralgia

Source	Age/Sex	Trigeminal divisions involved/Side	Agenesis of ICA	Anamnesis	Compressing vessel	Therapy methods
Fukuda M(2)	69/F	3 rd /L	L	Nothing	Elongation and dilatation of L VA	MVD
Uchino A(3)	62/M	2 nd /R	R	Hypertension Hyperlipidemia	Large tortuous BA	CBZ Infraorbital nerve block
Li F(4)	27/M	3 rd /L	L	Unknown	Ecstatic and tortuous BA	Local anesthesia Radiofrequency thermocoagulation
This case	46/M	2 nd /L	Bilateral	Hypertension Hyperlipidemia Hyperuricemia	Large tortuous BA	CBZ

M male, **F** female, **L** left, **R** right, **ICA** internal carotid artery, **BA** basilar artery, **VA** vertebral artery, **CBZ** carbamazepine, **MVD** microvascular decompression.

congenital defects resulting from abnormalities occurring during embryonic development, acquired conditions during infancy such as trauma to the head and neck, and arteritis-related occlusion. The internal carotid arteries begin to develop in the embryo at a crown-rump length of 3 mm, forming from the first and third aortic arches (forming the distal and root portions, respectively) and the dorsal aortae between them (forming the intermediate portion). At a crown-rump length of 4 mm, the distal portion divides into an anterior division that will later become the anterior cerebral, middle cerebral, and anterior choroidal arteries and a posterior division that will later become the posterior communicating and posterior cerebral arteries. At a crown-rump length of 12–14 mm, the dorsal aortae between the third and fourth aortic arches atrophy and disappear, and the carotid canal is formed (25). Congenital defects are thought to result from abnormalities in this process. The present patient had no clear history of arteritis or trauma to the head or neck during infancy, no indications of inflammation on blood testing, and no appearance of the carotid canals on CT. Agenesis of the internal carotid arteries was thus considered congenital.

Conclusion

1. Although bilateral agenesis of the internal carotid arteries was found, symptoms of trigeminal neuralgia were unilateral.
2. MRI, MRA, and CT are essential when investigating

trigeminal neuralgia, in order to visualize the head and neck region and identify any causal vessels and confirm the presence or absence of tumors.

3. Patients with abnormal hemodynamic status such as agenesis of an internal carotid artery or megadolichobasilar anomaly should undergo regular follow-up due to the high risk of cerebral hemorrhage and cerebral infarction.

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