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Case Report

Transient Anarthria and Quadriplegia in a Patient with Basilar Artery Hypoplasia and Coincidental Intracranial Lipoma: A Case Report

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Keywords

Basilar artery · Hypoplasia · Lipoma · Intracranial tumor · Stroke

Abstract

Cerebral ischemia may be rarely associated with a hypoplastic vertebrobasilar system. Intracranial lipoma is also a very rare congenital malformation. We report the case of a 52-year-old woman with vertebrobasilar transient ischemic attack associated with basilar artery hypoplasia and coincidental intracranial lipoma. She presented with sudden-onset dizziness, anarthria, and quadriplegia lasting for about 30 min. The patient's initial blood pressure was measured at 200/120 mm Hg. The magnetic resonance and computed tomographic images showed the absence of an acute ischemic lesion in the brain but revealed a hypoplasia of the basilar artery and bilateral V4 vertebral arteries. A lipoma of 11 mm in long diameter was also found in the quadrigeminal cistern and at the superior vermis. The electroencephalography, transthoracic echocardiogram, 24-h Holter monitoring, and transcranial Doppler ultrasonography, including patent foramen ovale study, were all noted as negative. The patient was treated with oral aspirin 100 mg, atorvastatin 10 mg, and antihypertensive medication. She had no symptom recurrence after the treatment. Our case suggests that hypoplasia of the vertebrobasilar arteries

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can be a predisposing factor for posterior circulation ischemia, especially when additional vascular risk factors coexist.

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Introduction

Basilar artery (BA) hypoplasia is a rare vascular variant with no reliable data regarding its prevalence. It was reported to be associated with a posterior circulation infarct among patients with undetermined or lacunar stroke [1]. Intracranial lipoma is a very rare benign tumor that is believed to constitute less than 0.5% of all intracranial tumors [2, 3]. It is a congenital malformation that is generally accepted to result from an abnormal differentiation of the primitive meninx [4]. Most of these tumors are found to be located at the midline subarachnoid cisterns, most frequently in the pericallosal and quadrigeminal/superior cerebellar regions [4, 5]. 15–55% of the lesions were reportedly associated with other anomalies of the adjacent brain region, including corpus callosum dysplasia, inferior colliculus hypoplasia, vermis hypoplasia, aneurysm, aqueductal stenosis, and mega cisterna magna [3–5].

The presenting case showed transient symptoms of vertebrobasilar insufficiency, which was thus presumably associated with BA hypoplasia. Also, an intracranial lipoma was coincidentally found in the quadrigeminal cistern and the superior vermis. This is the first case report, to our knowledge, of a coincidental vertebrobasilar transient ischemic attack associated with BA hypoplasia and intracranial lipoma.

Case Report

A 52-year-old previously healthy woman presented to the emergency room with sudden-onset anarthria and quadriplegia. During the initial stage of the attack at home, the patient suddenly fell down and felt a sense of being dizzy, then experienced that she could not move any limbs and articulate any words. In the emergency room, the neurologic exam showed severe dysarthria (it was difficult to be intelligible for the patient) and quadriplegia (symmetric weakness of Medical Research Council grade 2 in all limbs). At that time, the patient was alert. Cranial nerve examination was negative, except for a doubtful bilateral facial weakness. Deep tendon reflexes were symmetric and slightly hyperactive in the bilateral limbs, but no Babinski sign was present. The patient's symptoms completely recovered within 10 min after arrival at the emergency department. In the end, it was noted that total symptom duration was about 30 min. When questioned, the patient denied the occurrence of diplopia, loss of consciousness, abnormal movement, headache, hearing loss, or tinnitus. The patient's initial blood pressure was 200/120 mm Hg, but the body temperature, pulse, and respiratory rate were within the normal ranges. The patient had no previous history of a psychiatric illness, drug or alcohol abuse, cigarette smoking, or any other vascular risk factors, including hypertension, diabetes, and hyperlipidemia.

Diffusion-weighted magnetic resonance (MR) image showed a negative finding. However, the computed tomography (CT) and the MR angiography revealed a fetal-type posterior circulation in the right side, and a hypoplasia of the BA and the bilateral V4 vertebral arteries (VAs) (Fig. 1). The diameters of the BA at the mid-pons and VAs 10 mm proximal from the basilar junction were measured based on time-of-flight source images [1]. The diameters of the BA, right VA, and left VA were 1.71, 1.54, and 1.65 mm, respectively.

In particular, the noncontrast CT revealed a hypodense small mass of 11 mm in long diameter (–70 Hounsfield units) in the quadrigeminal cistern (Fig. 2a). The lesion was bright on both T1- and T2-weighted MR images (Fig. 2b, c) but was hypointense on susceptibility-weighted MR image (Fig. 2d). The lesion extended from the quadrigeminal cistern to the superior vermis. Electroencephalography, transthoracic echocardiogram, 24-h Holter monitoring, and transcranial Doppler ultrasonography, including patent foramen ovale study, were all negative. Laboratory tests were all unremarkable, except for mild hyperlipidemia (total cholesterol 214 mg/dL, triglyceride 82 mg/dL, high-density lipoprotein cholesterol 60 mg/dL, and low-density lipoprotein cholesterol 137 mg/dL).

The patient was treated with oral aspirin 100 mg and atorvastatin 10 mg. Blood pressure was constantly measured at more than 140/90 mm Hg, and so amlodipine 5 mg was prescribed. The patient had no symptom recurrence after the admission.

Discussion

The patient's symptoms were presumed to be caused by transiently reduced blood flow in the vertebrobasilar system because they are characteristic for vertebrobasilar insufficiency, including drop attack, dizziness, dysarthria, and quadriplegia. However, she had no atherosclerotic steno-occlusive lesion in the vertebrobasilar system. Besides, no cardioembolic source was found in the heart studies.

Instead, the hypoplastic vertebrobasilar artery could be postulated to have been the culprit lesion for the patient's symptoms [6, 7]. It is possible that a trigger condition (such as emotional stress) can lead to a paradoxical constriction of the vulnerable hypoplastic artery and a subsequent transient hemodynamic insufficiency in the corresponding vascular territory [8]. Furthermore, the hypoplastic artery is not only prone to collapse but is also more susceptible to prothrombotic and atherosclerotic processes than normal-sized arteries because of a decreased flow capacity [9]. Previous cases have shown the possibility that a hypoplastic BA may lead to a posterior circulation ischemia [6, 7]. In particular, when additional atherosclerotic risk factors coexist, the stroke risk further increases [10]. A recent study also suggested that a BA hypoplasia is associated with posterior circulation stroke in lacunar or undetermined stroke [1]. In addition, a fetal-type posterior circulation, though always accompanied by a BA hypoplasia, does not contribute to the infratentorial region because the tentorium cerebelli blocks the leptomeningeal connection [11].

Another peculiar finding in our case is an incidentally found intracranial lipoma, a very rare congenital malformation. It is usually asymptomatic, like in our case, except for rare cases, such as sylvian fissure lipoma associated with epilepsy [3]. Interestingly, an intracranial lipoma is frequently accompanied by other anomalies of the neighboring structures. The etiology is unclear, but it was suggested that the lipoma might lead to a mechanical interruption or a focal perfusion insufficiency, disturbing the normal growth of adjacent neural structures [12]. Callosal dysgenesis has thus been the most frequent anomaly, since intracranial lipoma most frequently occurs in the interhemispheric pericallosal region [2, 4]. Besides, arterial abnormalities, such as aneurysms near the lipoma (e.g., aneurysm of the middle cerebral artery near the sylvian fissure lipoma), have been reported [3, 13].

However, the BA hypoplasia is not neighboring to the quadrigeminal lipoma in our case. Thus, it is likely to be an incidental finding irrelevant to lipoma. On the other hand, a case of associated anomalies distant from the lipoma was previously reported in the literature [3]. The case with an intercerebellar fissure lipoma had not only a vermis hypoplasia near the

lesion but also multiple distant intracranial anomalies, such as a saccular aneurysm in the right cavernous internal carotid artery and a callosal dysplasia [3]. The explanation for this finding is still unknown. Indeed, the lipoma might have secreted some factors inhibiting the growth of the associated structures [13]. Therefore, the hypoplastic artery in our case might be another abnormality associated with the intracranial lipoma, although this appears unlikely, since it has not been reported in the literature so far.

Conclusions

Our case showed hypoplasia of the BA and bilateral V4 VA presumably causing a vertebrobasilar transient ischemic attack, as well as a coincidental lipoma in the quadrigeminal cistern. This case suggests that hypoplasia of the vertebrobasilar arteries can be a predisposing factor for posterior circulation ischemia, especially when additional vascular risk factors co-exist.

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Statement of Ethics

The authors have no ethical conflicts to disclose.

Disclosure Statement

The authors have no conflicts of interest to declare.

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Author Contributions

All authors met the criteria for authorship and have approved the contents of the text. K.-O.J., D.-H.H., and J.-H.P. did the clinical assessment of the patient and treated her. K.-O.J. and S.-J.L. contributed to the study concept and wrote the first manuscript. All authors participated in the data interpretation and the writing of the manuscript. S.-J.L. and H.J.K. revised the text.

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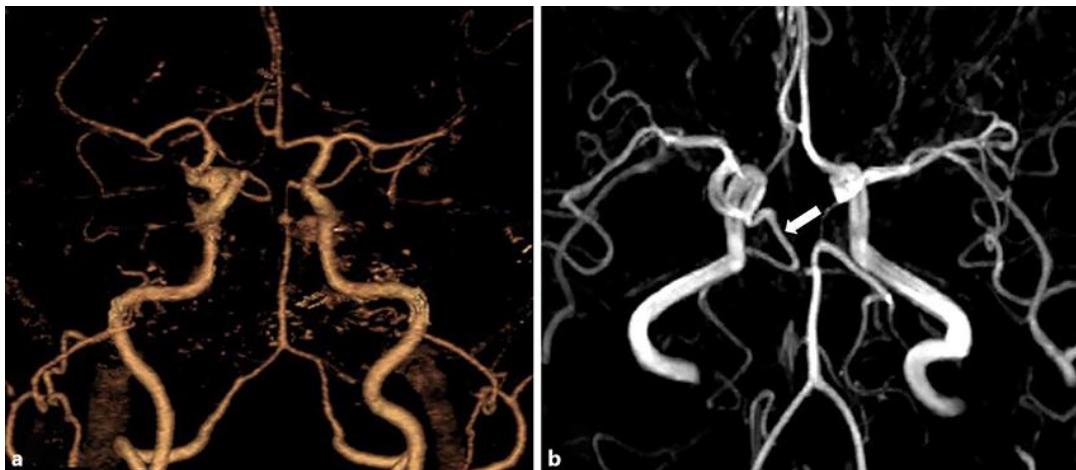


Fig. 1. The images of intracranial brain arteries on computed tomographic (a) and magnetic resonance angiography (b). The images show a fetal-type posterior circulation on the right side (arrow on b) and a continuously reduced diameter from the bilateral V4 vertebral arteries all through the basilar artery.

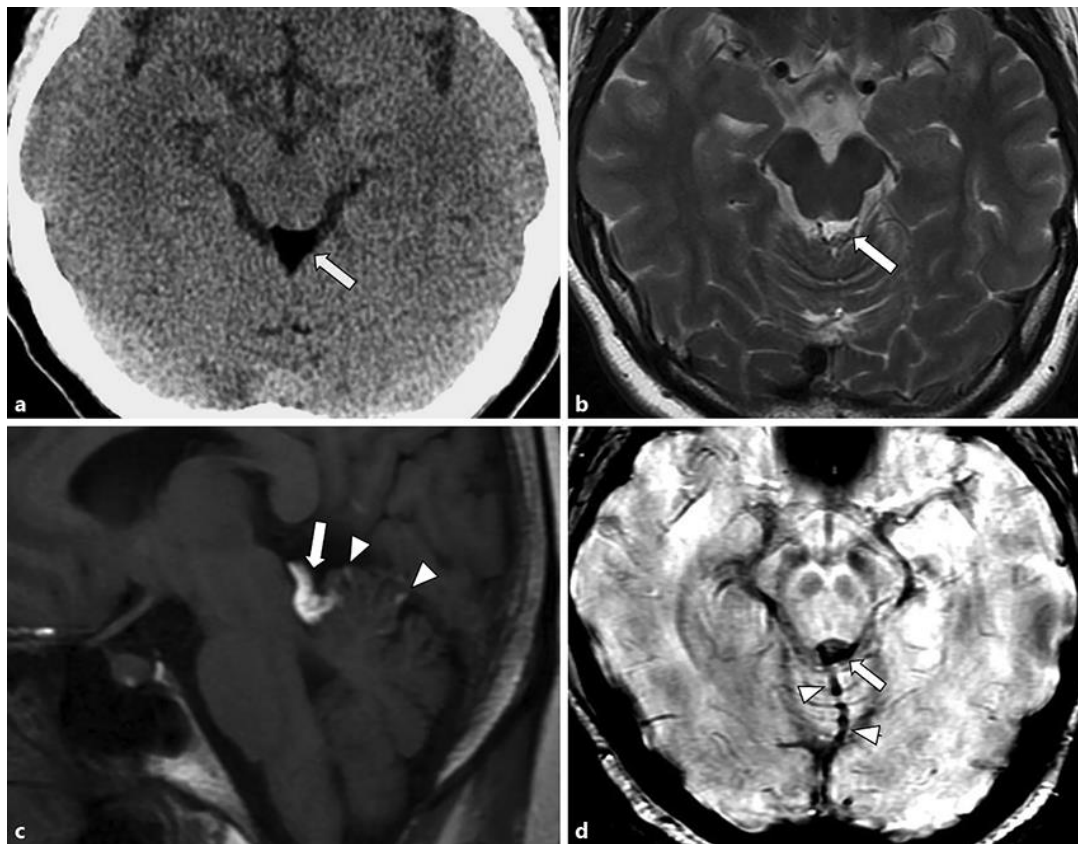


Fig. 2. Images of the lipoma. Noncontrast computed tomographic axial image shows the hypodense small mass of 11 mm in long diameter (–70 Hounsfield units) in the quadrigeminal cistern (a). The lesion is bright on both T2-weighted axial (b) and T1-weighted sagittal (c) magnetic resonance images but is hypointense on susceptibility-weighted axial image (d). The lesion extends from the quadrigeminal cistern to the superior vermis (arrowheads on c and d).