Persistent Primitive Proatlantal Intersegmental Artery (Proatlantal Artery I) Results in ‘Top of the Basilar’ Syndrome

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Background: Persistent primitive proatlantal intersegmental artery (proatlantal artery I) is an anastomotic channel between the carotid and vertebrobasilar arterial systems. Persistence of this embryonic anastomosis is very rare. The “top of the basilar” syndrome is usually due to occlusion of thalamoperforating arteries by emboli that rise from the vertebrobasilar arterial system. In this case, however, the emboli stemmed from atherosclerotic plaques in the carotid system.

Case Description: A 55-year-old woman was hospitalized with top of the basilar syndrome. A brain scan showed hypodense lesions extending from the mesencephalon to both thalami. Digital subtraction angiography of the cerebral circulation was performed, which revealed a proatlantal artery I on the left side. In addition, atheromatous plaques were seen along the internal and common carotid arteries on the same side.

Conclusions: The occurrence of proatlantal artery I is extremely rare. Top of the basilar syndrome is also rarely encountered. To our knowledge, this is the first report of a patient with both conditions. (Stroke. 1993;24:2114-2117.)

Key Words • carotid arteries • embolism • vertebrobasilar circulation

Persistant caroticovertebral embryonic anastomosis was first named by Padget1 the “persistent primitive proatlantal intersegmental artery.” Later, Lasjaunias et al2 called it the “proatlantal artery I.” This artery is extremely rare.

The occlusion of thalamoperforating arteries results in a clinical state that has different names. It was called the “syndrome of the mesencephalic artery” by Segarra.3 Later, Caplan4 called this syndrome the “top of the basilar.” The occlusion is usually caused by emboli that rise from atherosclerotic plaques in the cervical and intracranial portions of the vertebral artery and the proximal portion of the basilar artery.

In the case reported here, the atherosclerotic plaques situated along the left common and internal carotid arteries were the source of the emboli. These emboli passed through the proatlantal artery I on the left side. As a result, they caused the top of the basilar syndrome.

Case Report

A 55-year-old woman was admitted to the hospital in coma. She had a history of long-standing hypertension and right hemiparesis due to a lacunar infarction that occurred 1 year previously. She had not been using any drugs before admission. On the morning of admission she complained of headache, and she later lost consciousness.

The first neurological examination revealed loss of consciousness. However, she could localize painful stimuli. Her right nasolabial sulcus was slightly indistinct. She was able to move her four extremities. Pupils were equal and miotic. Her plantar reflex was bilaterally extensor. Stiff neck was absent. Cerebrospinal fluid examination revealed no abnormality. Physical examination of the heart, electrocardiogram, and echocardiogram were normal.

Brain computed tomographic scan showed hypodense lesions that started from the basal and medial thalamic regions at the level of the mesencephalon and extended to the thalami, giving a bilobular appearance (Fig 1).

Communication was possible verbally on the 10th day of hospitalization, but cognitive functions such as memory and speaking were still defective. The patient was somnolent and indifferent to her surroundings. Vertical gaze was limited bilaterally.

Doppler ultrasonography showed calcified atheromatous plaques in both carotid arterial systems, with the left internal carotid artery more affected than the right. Digital subtraction angiography demonstrated a vessel rising caudally from the left internal carotid artery. It curved sharply dorsally at the level of Cl to course in the occipitoatlantal space. Here, it proceeded horizontally and dorsally until it joined the vertebrobasilar system (Fig 2). In the anteroposterior view, this anomalous vessel curved around the lateral part of the transverse process of atlas and followed a suboccipital horizontal course to enter the skull through the foramen magnum (Fig 2, right panel). This anomalous vessel was accepted as the proatlantal artery I (Fig 3). The right internal carotid artery showed no abnormality and

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traversed its normal course. There was no anomalous vessel on the right side. Arch aortogram revealed a hypoplastic right vertebral artery and an aplastic left vertebral artery. Atherosclerotic plaques were present along the common and internal carotid arteries on the left side. They probably were the source of the emboli causing the clinical conditions we have described.

The main clinical findings were defective cognitive functions, bilateral upgaze palsy, and somnolence. Motor functions were preserved. These findings supported the diagnosis of top of the basilar syndrome. The patient was discharged with partial recovery after 37 days of hospitalization.

Discussion

In the very early stages of the human embryo, when it is approximately 3 to 4 mm long, various anastomotic channels are present between the carotid and vertebrobasilar systems. These channels start to regress when the embryo is approximately 7 to 12 mm long and disappear when the embryo is approximately 12 to 14 mm long. Rarely, some of the channels fail to regress and persist into adult life. They are known as the persistent primitive proatlantal, hypoglossal, otic, and trigeminal arteries.

Padget and Lasjaunias et al identified and named these vessels, and their terminology is still used. Arteries named by Padget “persistent primitive proatlantal intersegmental artery” and “persistent primitive first cervical intersegmental artery” are named by Lasjaunias et al “proatlantal artery I” and “proatlantal artery II,” respectively.

The type I proatlantal artery rises from the caudal, internal, or external carotid artery and ascends to the level of the occipitoatlantal (proatlantal) space without passing through the transverse foramen of any cervical vertebra. Here, it joins the fourth segment of the vertebral artery. The type I proatlantal artery takes a sharp curve dorsally and courses above the transverse process of the atlas horizontally and suboccipitally. It enters the skull through the foramen magnum, respectively.

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The type II proatlantal artery rises from the external carotid artery and joins the third segment of vertebral artery below the first cervical vertebra. Since it passes through the transverse process of the atlas, its dorsal curve is lower than that of type I. It also enters the skull through the foramen magnum.1,2,5-7

The type I proatlantal artery should be differentiated from a more frequently seen persistent hypoglossal artery. Many criteria have been established to differentiate the two arteries. Analysis of well-documented cases reveals the following differentiating features5,7,9-11:

1) The hypoglossal artery usually rises from the internal carotid artery at the level of the C1 or C1/2 interspace. The proatlantal artery I rises at the level of the C2 or C3 interspace. Thus, the hypoglossal artery leaves the internal carotid artery at a higher level than the proatlantal artery I. (2) The proatlantal artery I curves sharply dorsally to course in the occipitoatlantal space. The suboccipital horizontal course of the proatlantal artery I and the vertebral artery is identical. However, the proatlantal artery I does not pass through the transverse process of any vertebra. The hypoglossal artery has a more vertical course than the proatlantal artery I. It lacks the suboccipital horizontal sweep characteristic of the vertebral and proatlantal arteries and never extends as far posteriorly as the proatlantal artery. (3) The proatlantal artery enters the skull through the foramen magnum, as does the vertebral artery. The hypoglossal artery enters the skull through the hypoglossal canal. While it passes through the hypoglossal canal, it displays a small dorsal curve. This small curve must not be confused with the horizontal suboccipital course characteristic of the proatlantal artery I, which is related to the transverse process of C1.

In the case reported here, the anomalous vessel rose from the internal carotid artery opposite the lower end of C2 (Fig 2, left panel). It ascended to the level of the transverse process of C1. Then, it curved sharply dorsally to course in the occipitoatlantal space (Fig 2). This suboccipital horizontal sweep is characteristic of proatlantal artery I. It also extended farther posteriorly than would be expected of a hypoglossal artery. Thereafter, it entered the skull through the foramen magnum to join the vertebrobasilar system.

Proatlantal arteries type I and type II are rarely seen. Since they are asymptomatic, they are usually discovered incidentally. In many of the reported cases, the persistent artery is on the left. The vertebral artery is aplastic on the same side and hypoplastic on the other side in most cases. Thus, the posterior cerebral circulation is supplied by one of these persistent arteries. Occlusion of these arteries results in ischemia of areas supplied by the vertebrobasilar system. However, no other case has been reported until now, except a patient with vertebrobasilar system insufficiency resulting from an ulcerative lesion of the internal carotid artery.10

Since the proatlantal artery was first described by Padget,1 27 cases have been reported up to the present. In 1985, Sato et al11 reported 13 cases with proatlantal artery I.5,6,11-13 In 1988, Sato et al16 reported 8 cases with proatlantal artery II.16-19 In addition to these, 5 cases with proatlantal artery III8,10,20,21 and 1 with proatlantal artery II have been reported. A total of 18 cases with proatlantal artery I and 9 cases with proatlantal artery II have been reported thus far.

Our patient is the first to present with a top of the basilar syndrome. Top of the basilar syndrome was first described as the syndrome of the mesencephalic artery by Segarra3 in 1970. The main manifestations are loss of cognitive functions, bilateral vertical gaze palsy, and somnolence. However, motor functions are preserved. Later, Caplan4 called it top of the basilar syndrome. Basically, the lesion is in the area supplied by thalamoperforating arteries. These arteries rise from the mesencephalic artery, which is the continuation of the basilar artery. Top of the basilar syndrome frequently results from emboli rising from atherosclerotic plaques in the cervical and intracranial portions of the vertebral artery or the proximal portion of basilar artery.

In this case, emboli probably rose from the left common and internal carotid arteries, passed to the vertebrobasilar system through the proatlantal artery I, and occluded the thalamoperforating arteries, thus resulting in top of the basilar syndrome.

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