Cerebral Arteriovenous Malformation Embolized through Persistent Primitive Hypoglossal Artery

A Case Report

A.K. GUPTA

Department of Radiology Sree Chitra Tirunal Institute for Medical Sciences and Technology; Trivandrum, India

Key words: hypoglossal artery, carotid-vertebral anastomoses, arterio-venous malformation, embolization

Summary

Carotid-vertebral anastomoses are commonly detected as incidental findings. But sometimes these channels are important for the clinical condition of the patient. Here a case of right thalamo-capsular arteriovenous malformation is described where a persistent primitive hypoglossal artery was the only route for embolization of the arteriovenous malformation.

Introduction

Carotid-vertebral anastomoses develop during fetal life between the internal carotid and basilar arterial systems for the supply of the posterior cranial circulation. The channels include trigeminal, hypoglossal, otic and proatlantal segmental arteries. With the development of the posterior communicating artery, these channels regress and disappear but may rarely persist into adult life. The persistent trigeminal artery is the most common anomaly among the four, followed by the hypoglossal artery¹. These channels are detected as incidental findings but may be associated with aneurysms, tumors, subarachnoid haemorrhage and arterio-venous malformations¹⁻⁵. We describe a case of right thalamo-capsular arteriovenous malformation, for which embolization was done through persistent primitive hypoglossal artery.

Case report

This 47-year-old male patient presented with history of sudden onset left upper and lower limb dense hemiplegia associated with loss of consciousness, left focal seizures and left hemisensory dulling. CT scan showed an arteriovenous malformation (AVM) involving the right thalamus and posterior limb of right internal capsule with evidence of haematoma in the same regions. He was managed conservatively after that acute episode. Later digital subtraction angiography showed right thalamic and internal capsular AVM fed by perforating arteries from the right middle cerebral, anterior cerebral and posterior cerebral arteries along with right anterior and posterior choroidal arteries (figure 2A,B).

The AVM showed both superficial and deep venous drainage, but predominant drainage was through multiple superficial cortical veins due to significant stenosis of the straight sinus. The AVM nidus was compact, measured less than 3 cms in diameter without any intranidal aneurysm or venous ectasia. So it was in Spetzler – Martin grade 3. On left common carotid (CCA) injection basilar artery (BA) was filling through a large anastomotic vessel between internal carotid artery (ICA) and intradural vertebral artery. ICA distal to this showed smaller in caliber and poor forward flow compared to

Figure 1 (A) Left common carotid lateral angiogram shows the persistent hypoglossal artery (PHA) arising from internal carotid artery (ICA) and entering through the hypoglossal canal and joining the proximal basilar artery. (B) Anteroposterior view of left ICA with bony landmark show the course of PHA posteromedial to ICA and filling of arteriovenous malformation (AVM).

the right side. The anastomotic vessel arose from the C2 vertebral body level and ascends posterosuperiorly and takes a dorsal loop at the level of basion (within the hypoglossal canal) then joins with the proximal part of the BA (figure 1).

CT angiogram showed that the anastomotic artery is entering through the enlarged left hypoglossal canal (figure 3). Both vertebral arteries at V4 segment and bilateral posterior communicating arteries were hypoplastic (figure 2C,D). Direct arteriovenous fistulae from two posterior cerebral artery perforating feeders were embolized using 50% N-butyl cyanoacrylate. Since rest of the feeders too small for selective catheterization, it was decided to send him for stereotactic radiotherapy for further management.

Discussion

The persistence of a primitive hypoglossal artery is a rare anomaly of Carotid-vertebral anastomoses. Reported frequency of persistent hypoglossal artery (PHA) is 0.02-0.1% which is lower than that of persistent trigeminal artery (0.1-0.6%), the most commonly found carotid-vertebral anastomoses⁶. At 4 mm embryo stage

of development the internal carotid arteries extend from the paired dorsal aorta and anastomoses at three major sites with paired neural arteries in the hindbrain. These sites of communication are named for their relation to the trigeminal ganglion, otic vesicle and rootlets of the hypoglossal nerve forming the trigeminal artery, otic artery and hypoglossal artery. Otic is the first to disappear followed by the hypoglossal and lastly the trigeminal artery¹. At the same stage there are seven transversely oriented arteries in the cervical region (cervical inter segmental arteries), the first of these has been designated as proatlantal intersegmental artery.

PHA originates from the internal carotid artery as a large extra cranial branch at C1 – C3 level and enters the skull through the anterior condyloid foramen (hypoglossal canal) supplying the basilar artery⁷.

Initially the absence of posterior communicating artery was considered as an additional criterion for the presence of PHA. However Agnoli⁸ in a review of 80 cases, reported hypoplastic or angiographically absent posterior communicating arteries on both sides are present in 78% of cases and hypoplastic vertebral arteries in 79% of cases.

Figure 2 (A) Left PHA injection shows the connection to proximal basilar artery. (B) Left ICA injection lateral view shows the filling of AVM via perforators. Note the non-opacification of posterior communicating artery. (C,D) Right and left vertebral artery injections show the hypoplastic distal vertebral arteries.

The characteristics which constitute the criteria for identification of the PHA are described as follows: a) the PHA arises from the cervical part of the internal carotid artery at the C1-C2 level, b) together with the XIIth cranial nerve enters the posterior cranial fossa via the hypoglossal canal, c) the basilar trunk arises from the branch

of PHA and d) the ipsilateral vertebral artery may be hypoplastic⁹. In our case it exactly followed the typical course of a PHA. The ipsilateral ICA was smaller and bilateral vertebral vertebral arteries were hypoplastic distally.

Although conventional angiography has been considered as the reference standard for the

Figure 3 A) Plain CT of base of skull shows the enlarged left hypoglossal canal. B,C,D) Contrast enhanced CT scan shows the PHA coursing posteromedial to ICA and entering through the hypoglossal canal to join the basilar artery.

detection and evaluation of vascular structures, there are diagnostic alternatives with different advantages. CT angiography is a noninvasive examination, which has the advantage of multiplanar demonstration of the PHA by the use of

3D reconstruction techniques. Therefore, CT angiography provides excellent anatomic localization of PHA in all its parts and depicts clearly its entrance to the hypoglossal canal⁶. In our case CT angiography showed the course of the

artery through hypoglossal canal, not via foramen magnum, which is typically followed by proatlantal arteries.

MR imaging and MR angiography in particular is able to sufficiently demonstrate the PHA, without any contrast medium. MR angiography, by using a maximum-intensity-projection (MIP) program is able to show the complete carotid-vertebral anastomoses without superimposition of other vessels^{6,10,11}.

Both CT and MR angiography are diagnostic alternatives, which have already been reported in the literature in cases of PHA detection and evaluation of certain pathologic conditions. Thus, stenosis due to atherosclerotic plaque or aneurysm formation can be depicted by 3D CT or MR angiography^{12,13}.

Since the presence of PHA may be completely asymptomatic, it may appear as an incidental finding in a cerebral angiogram performed for another diagnostic purpose. However, its identification is clinically important for patient management, as for planning carotid endarterectomy or skull base surgery¹⁴. The existence of PHA has also been related with increased incidence of intracranial vascular abnormalities and atherosclerotic disease^{12,13,15-21}. The later may present as a plaque extending

from the distal carotid artery into the internal carotid artery (ICA) or as an isolated stenosis near the PHA takeoff. The group of persistent primitive arteries, in which PHA belongs to, is implicated in cases of moyamoya and quasi-moyamoya disease as well^{22,23}. Although some reports suggest higher frequency of intracranial arteriovenous malformations and aneurysms in the presence of a PHA, no statistically significant difference has been found⁶. PHA may provide pathway for cerebral embolism.

In contrast to PHA, the pro atlantal inter segmental artery curves sharply dorsally to course in the occipitoatlantal space like vertebral artery and then passes through the foramen magnum. Hypoglossal artery has a more vertical course than the proatlantal artery and lacks the horizontal sweep of vertebral and proatlantal artery²⁴. This carotid-vertebral anastomosis is extremely rare. There were only six reported angiographic demonstrations until 1976²⁵.

This case was unique in the sense that the access to the AVM was possible only through the PHA as the V4 segment of both vertebral arteries and posterior communicating arteries were hypoplastic. To the best of our knowledge, there is no report of AVM embolization through the PHA.

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Dr A.K. Gupta
Professor and Head
Department of Radiology
Sree Chitra Tirunal Institute
for Medical Sciences and Technology
Trivandrum, Kerala
695011 India
E-mail: gupta@sctimst.ac.in

EDITORIAL COMMENT

In their paper of "cerebral arteriovenous malformation embolized through persistent primitive hypoglossal artery – a case report" and Al. are reporting a very rare situation where a brain AVM is associated to an arterial embryonic anomaly. This type of anomaly is often associated to aneurysms or to different diseases like the PHACE syndrome.

It is unfortunate that there is still historical reference being made and not updated concerning the otic embryonic artery. As discussed the past 20 years by ourselves and others it is likely that this artery does not exist. It is absent of the evolution of species, never seen, persistent or enlarged in any disease in human.

It evident today that trigeminal hypoglossal and proatlantal arteries are the only three arterial systems, that follow actually regroupment of the cranial peripheral nerves (V2-V3-Vm and VII, XI-X-XI-XII and C1-C2). V1, III, IV and VI involving the infero-lateral trunk. A large range of predictable of variations is associated to the persistence of hypoglossal-ascending pharyngeal systems, AICA and PICA. They have been extensively illustrated and commented in the literature and text books.

Pierre Lasjaunias

Hôpital de Bicêtre
Service de Neuroradiologie Diagnostique
et Thérapeutique
Le Kremlin - Bicêtre, France

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