The double carotid bifurcation: An almost typical sign in a persistent hypoglossal artery

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Patient: 65 years, female

Clinical History:

This is a report of a patient with an anomalous vascular supply of the vertebrobasilar territory arising from the left internal carotid artery (ICA), which also exhibited a high-grade stenosis.

Imaging Findings:

A 65-year-old woman had suffered repeated drop attacks without any other neurological signs or symptoms. Blood pressure and a single ECG had been normal, but a full cardiologic workup was still pending. On a CCT, no parenchymal brain lesions were detected. Colour-coded duplex ultrasound had shown acoustic shadowing due to a calcified plaque in the left internal carotid artery. Stenosis grading was however specified as 80%. Moreover, kinking or coiling of the left ICA was suspected. A CT angiogram could not be obtained due to multiple severe allergies including contrast media. The patient was scheduled for MR angiography of the supra-aortic arteries, which was performed as a non enhanced 3D time-of-flight multislab sequence from the jugular notch to the carotid siphon. Whole-volume MIPs showed severe hypoplasia of the right and complete absence of the left vertebral artery (VA). Instead, a stout anomalous vessel originated from the left ICA, leading into the basilar artery (Fig. 1). The plaque stenosis of the left ICA was displayed more distinctly on thin-slab MIPs (Fig. 2). Transverse MPRs of the 3D TOF sequence showed a second bifurcation in the course of the left carotid artery (Fig. 3a). The anomalous vessel crossed behind the ICA to enter the hypoglossal canal (Fig. 3b). CT sections of the skull base exhibited extensive calcifications of the anomalous vessel (Fig. 4).

Discussion:

The hypoglossal artery (HA) is one of four anastomoses between the primitive carotid arteries and the paired longitudinal neural arteries of the hindbrain, which later form part of the vertebrobasilar system. They occur around the 29th day of embryonic life, persist for about 1 week, and disappear after normal development of the posterior communicans and vertebral arteries. Those presegmental arteries are denoted after their accompanying nerves as the otic, trigeminal, hypoglossal and proatlantal (C1) arteries in metameric order. Any of these anastomoses may persist through adulthood. The most frequently encountered persistence is that of the trigeminal artery with a reported incidence of 0.1-0.6 %, followed by the HA, which is about ten times less frequent [1]. In the proatlantal artery (PA), and even more the otic artery, literature reports are only sporadic. However, the proatlantal and hypoglossic artery share the extracranial origin, whereas the trigeminal and otic arteries are exclusively intracranial vascular abnormalities. The HA typically originates from the ICA between C1 and C3, although there are exceptions to this rule [2]. The PA, in contrast, originates from the external carotid artery in the majority of cases. Less than 40% have been reported to arise from the ICA [1]. Hence, if a “double carotid bifurcation” is found on imaging studies, a persisting hypoglossal artery is by far the most probable diagnosis. However, the course of the vessel...
through the hypoglossal canal must be ascertained. Duplex sonography studies are rather misleading [3]. Transverse MR or CT thin sections of the skull base usually show a widened hypoglossal canal on the side of the anomaly [4], which occurs unilaterally in almost 99% of cases. In the present case, arteriosclerotic wall calcifications facilitated the identification of the anomalous vessel on a non enhanced cranial CT. Among the four persistent primitive anastomoses, HA is also the one most likely to cause clinical problems [5]. If it is present, the ipsilateral vertebral (VA) and posterior communicating arteries (PCA) are usually hypoplastic or absent. Patent contralateral VAs and PCAs are only present in one third of cases [1]. Thus, the hypoglossal artery becomes the only significant feeder of the posterior circulation. The predominant dependence of the cerebral circulation from the carotid system may have catastrophic consequences in the case of a critical reduction of the carotid blood flow [6]. The proximal ICA stenosis observed in this case may well have been the cause of drop attacks in the presence of transient cardiac arrhythmias. An association of the persistent HA with aneurysms has also been described, not only in the anomalous vessel and the vertebrobasilar region, but also in the anterior circulation [7]. In Moyamoya and quasi-Moyamoya disease, persistent carotid-vertebrobasilar anastomoses have been found much more frequently than in the general population [8]. The significance of this association has still to be elucidated.

**Differential Diagnosis List:** Persistent embryonic hypoglossal artery with ipsilateral ICA stenoses

**Final Diagnosis:** Persistent embryonic hypoglossal artery with ipsilateral ICA stenoses

**References:**


Figure 1

Description: Volume rendered LAO transparency image of the carotid arteries. There is a second bifurcation (2) of the internal carotid artery above the normal bifurcation of the common carotid artery (1), giving rise to the persistent hypoglossal artery. Origin:
Description: Whole volume ap MIP showing an anomalous vessel arising from the left carotid and supplying the vertebrobasilar trunk. Also note the absence of the left and the severe hypoplasia of the right VA. Another intracranial left carotid stenosis may be present at the siphon level. This region is however subject to artifacts. Origin:
Description: Signal void at the origin of the left ICA due to a calcific plaque protruding into the vessel lumen. The persistent HA lies dorsal to the ICA. The filiform vessel parallel to the carotid is actually the right VA, due to an oblique projection plane. Origin:
Figure 3

Description: There is a second bifurcation of the left carotid artery at the C1/2 level giving rise to the hypoglossal artery. \(i\)=internal carotid arteries, \(e\)=external carotid arteries, \(h\)=hypoglossal artery.

Origin:
Description: The left hypoglossal artery crosses the internal carotid dorsally to enter the hypoglossal canal. i=internal carotid artery, h=hyoglossal artery. Origin:
Figure 4

Description: CT of the skull base reconstructed with an edge-enhancing algorithm shows the widened left hypoglossal canal filled by the calcified hypoglossal artery (arrow). Origin: