



PERSISTENT PRIMITIVE HYPOGLOSSAL ARTERY WITH VERTEBRAL ARTERIES AGENESIS AND CEREBELLAR & OCCIPITAL ATROPHY: CASE REPORT.

Radiology

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ABSTRACT

We report a rare case of persistent primitive hypoglossal artery with complete agenesis of bilateral vertebral arteries. MRI showed diffuse cerebellar, posterior parietal and occipital atrophy with only mild atrophy of brain supplied by anterior circulation. On further evaluation with MR Angiography, right sided persistent primitive hypoglossal artery was seen arising from the internal carotid artery and continuing to form basilar artery with agenesis of bilateral vertebral and posterior communicating arteries. Posterior cerebral and cerebellar atrophy seen in this patient can be attributed to the persistent primitive hypoglossal artery being the sole blood supply with no communication between anterior and posterior circulation with absence of bilateral posterior communicating arteries.

KEYWORDS

Primitive Persistent Hypoglossal artery, Agenesis of vertebral arteries, Cerebellar and posterior cerebral atrophy

INTRODUCTION

Four anastomosis exist between the anterior carotid circulation and the posterior vertebrobasilar circulation in embryonic life, with posterior circulation relying almost entirely by blood supply from the anterior circulation. Hypoglossal artery is one of these four anastomosis. With development of the posterior fossa structures, posterior circulation becomes more independent of the anterior circulation and these anastomosis get obliterated [1,2]. Sometimes there is persistence of these anterior to posterior anastomosis in adult life which may be incidentally detected or may have clinical implications. We report a case of persistent hypoglossal artery with atrophy of posterior cerebral structures – cerebellum, posterior parietal and occipital regions.

CASE REPORT

A 57 year old female was referred to the radiology department to undergo MR examination in view of recent behavioural changes. Three-dimensional time of flight (3D TOF) MR angiography images revealed bilateral common carotid artery bifurcation plaques with a luminal narrowing of about 30% to 40% along with agenesis of bilateral vertebral arteries. On further detailed evaluation an anomalous vessel was seen in right hypoglossal foramen. The right persistent primitive hypoglossal artery was seen to arise at the level of C2 vertebra from right internal carotid artery (ICA), continuing as the basilar artery. On evaluation of the circle of Willis, there was agenesis of posterior communicating arteries on both sides. Thus, the posterior circulation being supplied through the persistent carotid-vertebrobasilar anastomosis through hypoglossal artery. Remaining cranial and cervical vasculature was otherwise normal. The T1/T2/FLAIR images of the brain showed severe diffuse atrophy of cerebellar hemispheres and vermis. There was associated diffuse cerebral atrophy in the region of posterior circulation in bilateral posterior parietal regions and occipital lobes.

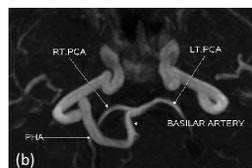
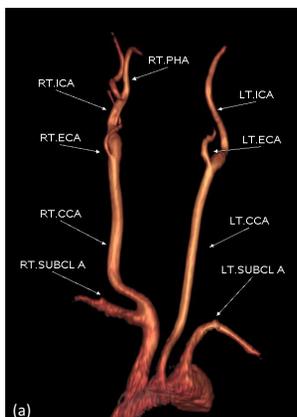


Figure 1: (a) 3D Surface shaded display (SSD) and (b) 3D TOF MIP image in axial plane show persistent hypoglossal artery (PHA) arising from right internal carotid artery and continuing as basilar artery with complete agenesis of bilateral vertebral arteries and posterior communicating arteries.

DISCUSSION

During early embryological development, carotid arteries and longitudinal neural arteries which later develop into vertebrobasilar system provide blood supply to the forebrain and hindbrain respectively. In early phases of development the posterior circulation relies almost entirely from blood supply coming from the anterior circulation through carotid-vertebrobasilar anastomoses. During this early period when the embryo is 4-5 mm in length for a period of about 7 to 10 days, communication is present between the two in form of trigeminal, otic, hypoglossal and proatlantal arteries. These communications or anastomosis with further development of the vertebrobasilar system, when the embryo is 7-12 mm in length, dissolve and posterior communicating arteries develop as a means of communication between the anterior circulation provided by internal carotid arteries and posterior circulation via the vertebrobasilar system [1,2]. The persistence of these anastomosis is a rare phenomenon with persistent trigeminal artery being most common and usually seen as an incidental finding with an incidence of 0.1 to 0.2 % [3]. Persistence of hypoglossal artery is a rare phenomenon being six times less commonly seen as compared to trigeminal artery with the persistent bilateral hypoglossal arteries becoming an even rarer occurrence. To our knowledge till date there have been four case reports of bilateral persistent hypoglossal arteries [4-7].

The persistent trigeminal and otic arteries arise from intracranial internal carotid arteries while hypoglossal and proatlantal artery arise from the extracranial or cervical internal carotid artery. Hypoglossal artery usually arise at C1 to C3 vertebral level [8]. It is associated with hypoplasia or agenesis of ipsilateral or bilateral vertebral arteries. In our case, there is complete agenesis of bilateral vertebral arteries from their origin along with agenesis of both posterior communicating arteries. A “double carotid bifurcation” usually indicates a persisting hypoglossal artery and to confirm the course of the vessel through the hypoglossal canal should be demonstrated [9,10].

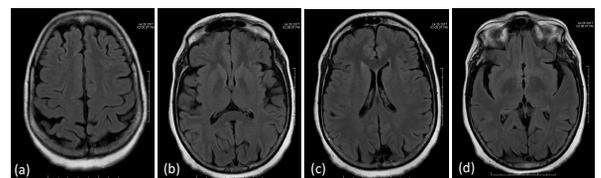


Figure 2: Axial fluid attenuated inversion recovery (FLAIR) images at different levels show diffuse atrophy of bilateral posterior parietal regions and occipital lobes.

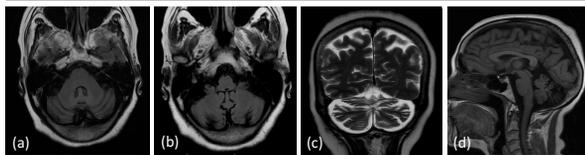


Figure 3: (a,b) Axial fluid attenuated inversion recovery images, (c) coronal T2 weighted and (d) sagittal T1 weighted images show diffuse cerebellar atrophy.

Usually an incidental finding persistent hypoglossal artery is sometimes of clinical significance and is seen to be associated with aneurysm at its junction with basilar artery and its complications such as rupture and hemorrhage. In case of significant narrowing at the carotid bifurcation vertebral artery steal may be seen to provide blood supply to anterior circulation through persistent hypoglossal artery leading to posterior circulation infarcts and other such disease manifestations due to reduced posterior circulation blood supply [7, 11, 12]. In our case, there diffuse atrophy of cerebellum, bilateral posterior parietal regions and occipital lobes supplied by posterior circulation, more than the mild atrophy seen in regions supplied by anterior circulation. These changes can be attributed to the anomalous changes in cerebral vasculature as the posterior circulation in this case is supplied entirely by the persistent primitive hypoglossal artery arising from right internal carotid artery, continuing as basilar artery with complete agenesis of bilateral vertebral artery. No communication is seen between the anterior and posterior cerebral circulation with agenesis of bilateral communicating arteries.

CONCLUSION

Identification of persistent primitive hypoglossal artery is important before carotid endarterectomy or skull base surgery, it being the only supply to posterior cerebral circulation. With the advent of imaging technology, it has become easy to detect such anomalies of the vertebrobasilar system and in view of the clinical significance it becomes important that detailed evaluation be done using either CT Angiography or MR angiography.

ABBREVIATIONS

3D TOF: Three dimensional time of flight; PHA: Persistent hypoglossal artery; MRI: Magnetic Resonance Imaging; ICA: Internal Carotid Artery; ECA: External carotid artery; Subcl: Subclavian; CCA: Common carotid artery; FLAIR: Fluid Attenuated Inversion Recovery.

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