

Posterior Circulation Positional Transient Ischaemic Attacks due to Persistent Primitive Hypoglossal Artery with Redundancy

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Abstract

A persistent primitive hypoglossal artery was the sole identifiable stroke risk factor for a patient presenting with positional, posterior circulation ischaemic episodes. Diagnosis was made by magnetic resonance arteriography and verified by conventional angiography. Tortuosity and coiling of the hypoglossal artery was also present. Dynamic transcranial Doppler sonography was normal and SPECT brain scanning revealed an area of posterior hemisphere hypoperfusion. Redundancy of the primitive hypoglossal artery is the postulated mechanism of cerebral ischaemia.

Introduction

Four different primitive arteries connect future anterior and posterior circulations in the embryonic state.¹ The trigeminal, otic, hypoglossal or proatlantal arteries normally regress and are replaced by the posterior communicating arteries but may persist in adult life. These four arteries may persist with differing frequencies and varying complications such as cerebral aneurysms,² cranial nerve palsies³ and as a route of anterior to posterior circulation embolism.⁴ Brain ischaemia and infarction have recently been described with such primitive anastomoses without any other cause of embolism or in association with carotid stenosis at the presumed donor emboligenic site.⁵⁻⁷ Hypoglossal arteries are said to be present in 0.05% of cerebral angiograms and have been reported to be associated with aneurysms,^{8,9} arteriovenous malformations,¹⁰⁻¹² moyamoya disease¹³ and Arnold Chiari malformation.¹⁴ Cerebral ischaemia has been described in one case.¹⁵ The mere presence of the congenital anomaly alone is insufficient to imply causation. A relatively young patient, free of cerebrovascular risk factors is presented with a hypoglossal artery with the additional abnormality of extensive coiling (redundancy) of the vessel in the neck.

Case report

A 48 year old right handed, white woman reported a sudden onset of bilateral visual impairment lasting several minutes, followed by intermittent shaking on both sides of her body lasting about 2 hours. Episodes of the body becoming "spastic" every 4-5 minutes were observed by her husband, lasting about 5 minutes at a time. He described the attacks as the arms and legs extending, and her lifting off the bed. The neck would become very stiff and she would arch her spine consistent with opisthotonic posturing. She was totally

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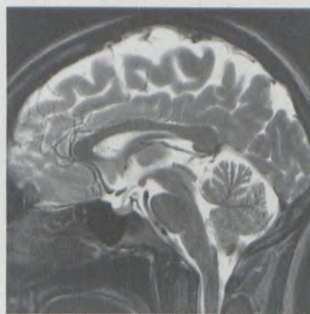
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conscious during these episodes. Pins and needles were perceived on both sides but more on the right. Numbness of the tongue, partial deafness of the right ear and dizzy spells were other frequent intermittent symptoms. A second similar attack occurred one week later. Of note is that turning her head to the left for more than 1-2 minutes invariably provoked dizziness. Looking down, such as when going down stairs and turning her head to either side would provoke dizziness. Tinnitus was invariable. No headache, weakness, diplopia or imbalance was associated. Her past history was notable in that she was diagnosed to have had a right sided "Bell's palsy" (of sudden onset) on the 20 July 1995 which lasted about 2 weeks. She had no cerebrovascular risk factors. General and neurological examination was normal save for mild dysmnnesia and visuospatial impairment, the latter tested with the Rey Complex Figure Test. The clinical assessment at time of first visit included a differential of posterior circulation ischaemia, partial seizures or cardiac dysrhythmias.

The basic cerebrovascular relevant blood screen, chest radiograph, electrocardiogram, electroencephalogram, computerised tomography brain scan and cardiac investigations including cardiac echogram were normal. The magnetic resonance imaging (MRI) brain scan showed a very small hyperintense focus in the left frontal region of dubious significance. An abnormal signal void due to a carotid basilar anastomosis was detected (Figure 1). This anastomosis was confirmed on time of flight magnetic resonance (MR) angiography as a very dilated tortuous hypoglossal artery with an aplastic right vertebral and hypoplastic left vertebral artery (Figure 2). Selective angiography confirmed the MR angiogram findings (Figure 3). The single photon computed tomography (SPECT) brain scan showed

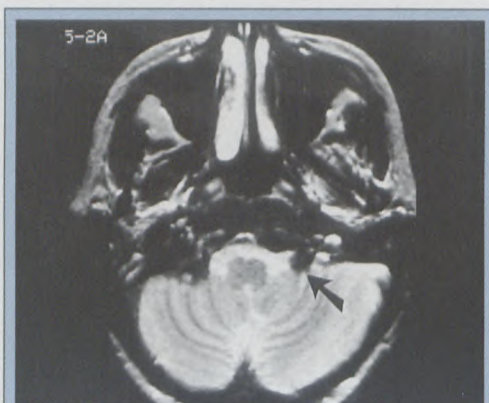


Figure 1: A T2 weighted axial scan demonstrates a signal void from the carotid basilar anastomosis (arrow).

left hemisphere hypoperfusion to visual inspection and semiquantitative analysis (Figure 4). The asymmetry index was calculated as 22%, significantly different to the normal controls (n=5) with a mean value of 4.2% for the parieto-occipital region (standard deviation ± 2.7 , range 0 - 9.5).



Figure 2: MR angiogram demonstrates the tortuous left hypoglossal artery (arrow). Note the aplastic right vertebral artery (arrowhead).

The cerebral vasomotor response was tested with intravenous Diamox without any further abnormalities noted on the SPECT scan. Transcranial Doppler sonography including dynamic studies with head rotation and flexion was normal. Specifically basilar artery velocity was unchanged with head rotation to the right and left for 2 minutes each. She was treated



Figure 3: Selective left vertebral angiogram confirms the tortuous left hypoglossal arterial anastomosis (arrow).

with Aspirin 150 mg daily and presently remains well at 2 months follow up.

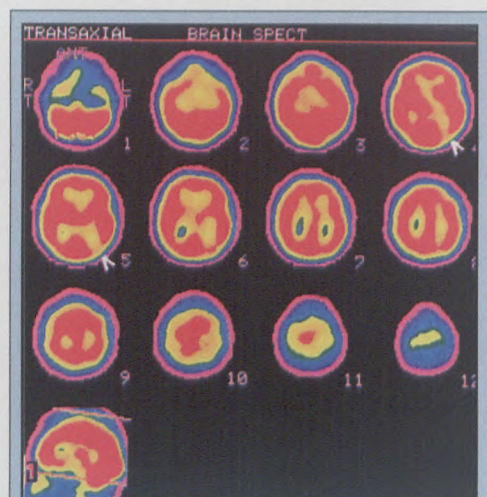


Figure 4: SPECT scan showing significant left parieto-occipital hypoperfusion (arrowheads).

Discussion

A series of case reports has implicated primitive caroticovertebral arteries in stroke and cerebral ischaemia. There is also a known stroke risk associated with cervicocephalic redundancy, with various mechanisms such as thrombosis and embolism postulated and dissection significantly associated.¹⁶ The most likely mechanism of ischaemia in our patient is a transient hypoperfusion or thromboembolism from the redundant coils of the primitive

hypoglossal artery which was the main posterior circulatory supply vessel in this patient. When these primitive arteries persist, aplasia or hypoplasia of the vertebrobasilar system is usual as in the patient under discussion. Positional ischaemia in anatomically normal vessels and atherostenosis with neck turning is well described by Sturzenegger *et al.*¹⁷ In our patient, head turning to either side but more to the left as well as flexion provoked dizziness. Both embolic and haemodynamic mechanisms could be pathomechanisms with the normal dynamic transcranial Doppler study marshalling evidence against the latter. This could be a mechanism of ischaemia especially as the abnormal hypoglossal artery served as the sole posterior circulation supply without the benefit of the normally dual vertebral arterial supply. Although no posterior circulation infarct was imaged with the aid of an MRI brain scan, functional imaging with SPECT scanning revealed left posterior hemisphere hypoperfusion, a finding that would correlate well with the symptomatology. The reported Bell's palsy was more likely to have been a minor pontine infarct as the clinical presentation was compatible with brain stem ischaemia. Although surgical reconstruction of carotid redundancy has been reported to abolish symptoms of cerebrovascular insufficiency, such treatment would theoretically be contraindicated in our patient in view of the anatomically abnormal posterior circulation.

This case demonstrates the utility of MR angiography in the detection of uncommon vascular abnormalities in patients with unexplained transient ischaemic attacks. The hypoglossal artery when present is usually the only functional artery to the brain stem and cerebellum and is associated with aplasia or hypoplasia of the vertebral arteries as demonstrated in our patient.¹⁸

To our knowledge, this is the first

reported instance of a primitive, persistent hypoglossal artery associated with redundancy without other identifiable cause of positional cerebral ischaemia/infarction. The causal relationship of these dual stroke risk factors seems certain. The need to exclude such vasculopathy in patients presenting with posterior circulation insufficiency is emphasised as the mechanism is amenable to treatment. Diagnosis by non-invasive magnetic resonance angiography should suffice.

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