An obscure cause of stroke - basilar artery fenestration

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Introduction

Cerebral artery fenestrations or intravascular bridges represent developmental anomalies, that may be incidental findings but sometimes of clinical significance.[1] These embryological abnormalities may also take the form of duplication of an artery. For the basilar artery, the incidence has been cited as 5.3% for the general population,[2] the most frequently involved intracerebral arterial system being the vertebrobasilar system.[3,4] The most important consequence is the tendency to develop arterial aneurysms at the site of fenestrations with a 3% incidence in a retrospective analysis of 5 190 angiograms.[4] Current pathophysiological data has shown that medial defects of the arterial walls predispose to aneurysm formation. Fenestrations have been described in the basilar, vertebral, middle cerebral, anterior cerebral, aortic arch and posterior cerebral arteries.[1,4] In addition to a propensity to form aneurysms, a number of other neurological presentations have been described in association with fenestrations including cerebral ischaemia,[5,6,7] trigeminal neuralgia,[8] cervical myelopathy[9] and symptomatic arteriovenous malformations.[10]

Case report

A 71 year-old white man suffered progressively more debilitating attacks of dizziness, diplopia, imbalance and inco-ordination over a seven year period with several daily attacks occurring at time of presentation.

History

He first reported intermittent diplopia eight years prior to presentation, lasting about 90 minutes. A second attack occurred nine months later. He was seen by his general practitioner and ophthalmologist at the time with no abnormality noted. One year later the attacks increased in frequency to one every few months culminating in a much more severe attack with dizziness lasting two hours but without abnormality seen on a MRI scan. During the next four years the diplopia increased dramatically varying from under one minute up to 30 minutes and could occur up to seven times per day. An episode occurred subsequently with dysphasia, imbalance and dizziness lasting about 30 minutes but with complete return to normality. A second MRI brain scan done at the time was normal. He was given various diagnoses such as transient ischaemic attacks and migraine by different neurologists. An even more disabling attack occurred six years after his first symptoms wherefrom he awoke with speech impairment, inability to walk, loss of co-ordination and difficulty in handling objects. This time a stroke was diagnosed and he improved again only to have a marked exacerbation of his symptoms one month later with intermittent attacks of a similar nature occurring 2-3 times per day lasting about between 30-60 minutes. He was quite disabled by these and described the left sided image (referring to his diplopia) as "coming and going all the time". The most recent presentation was associated with almost continuous diplopia and dizziness. He was otherwise in good health and had no cerebrovascular or cardiovascular risk factors, no deleterious habits and no general or neurological illness.

Examination

Examination revealed a rational man of normal body habitus with a BP of 145/90 and a pulse of 68 per minute regular. The cardiac, chest and abdominal examinations were normal and no stigmata of generalized disease were noted. No cervical or supraclavicular bruits were heard. Neurologically higher functions and cranial nerves were normal. Motor testing was normal save for bilateral upper limb relative hyperreflexia, left more than right. Sensation and limb co-ordination were normal and gait markedly ataxic with tandem gait impossible.

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Clinical assessment

The clinical assessment included a differential of posterior circulation ischaemia and/or infarction due to vertebrobasilar vascular abnormality.

Investigations

Routine blood tests, prothrombotic screen, chest radiograph and electrocardiogram were normal. Doppler sonography of the cervicocephalic and intracranial vessels was normal. Echocardiography and coronary angiography were normal. The third MRI brain scan was also normal.

Cerebral MR angiography proved diagnostic in that a proximal basilar artery fenestration was seen. This was first suspected due to the presence of a dilated mid basilar section with intravascular hypointense signal (Figure 1).

Figure 1: Magnetic resonance angiogram of the vertebrobasilar arteries. A dilatation of the mid basilar artery with an intravascular hypointense signal (arrow).

A transaxial magnetic resonance angiogram revealed a biconcave appearance of the basilar artery typical of a fenestration (Figure 2). Functional brain scanning with SPECT brain, revealed bi-occipito-parietal hypoperfusion.

Management

He was initially treated with Warfarin without relief of symptoms. Subsequent treatment with Aspirin alleviated some of the symptoms which further decreased with the addition of Persantin.

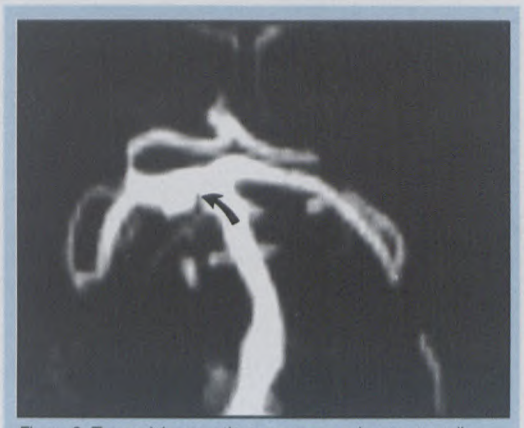

Figure 2: Transaxial magnetic resonance angiogram revealing the biconcave appearance of the basilar artery typical of a fenestration (arrow).

Discussion

Recognition of cerebral artery fenestrations in the context of cerebral ischaemia or stroke is important for at least three reasons:

1. It may represent the mechanism for the ischaemia or stroke.
2. Various treatment options are available which include medical, interventional radiological (Guglielmi coils)[11] and surgical options such as aneurysm clipping. The realisation that aneurysms may be part of this developmental abnormality should demand a comprehensive appraisal of the cerebral circulation by angiography.
3. A precise diagnosis as early as possible will also save unnecessary costly investigations (in this patient three MRI scans) and guide appropriate therapy.

In the case under discussion, presumably turbulent blood flow at the site of the fenestration lead to *in situ* thrombosis with distal embolisation and/or intermittent haemodynamic disturbances. This scenario is especially likely in that all other causes of posterior circulation ischaemia were excluded by a comprehensive stroke work up. In this patient Warfarin failed to alleviate symptoms whereas antiaggregant therapy led to a marked improvement. With the advent of newer antiplatelet agents such as Ticlopidine, Clopidrogel and combination formulae such as Asasantin, such high flow white thrombus type lesions are amenable to mechanism specific treatment.

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