Hemifacial spasm leading to diagnosis of Moyamoya disease

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Abstract

We present a case of Moyamoya disease presenting as hemifacial spasm due to compression of the facial nerve by a vascular loop related to compensatory enlargement of the posterior circulation vessels.

Case report

A 37-year-old man was assessed for right hemifacial spasm of 7 years duration. Clinical examination showed intermittent co-contractions of the right orbicularis oculi and zygomaticus muscles. His neurological and systemic examination were otherwise unremarkable.

His medical history was significant for a diagnosis of rheumatic fever at age 11 after developing involuntary movement of his right arm. He received penicillin prophylaxis until age 20.

Magnetic resonance imaging with angiography demonstrated several abnormalities:

- Compression of the right facial nerve by a vascular loop originating from the right anterior inferior cerebellar artery (Figure 1a).
- Bilaterally narrow internal carotid arteries consistent with Moyamoya disease (Figure 2a and 2b).
- Compensatory enlargement of the posterior circulation and external carotid arteries (Figure 2a and 2b). The right vertebral artery is hypoplastic (Figure 2a and 2b).
- T2 hyperintensity in the left centrum semiovale consistent with previous infarction (Figure 1b).
Figure 1. Axial magnetic resonance imaging showing compression of the right seventh nerve by a vascular loop from the anterior inferior cerebellar artery (1a, arrow). Axial T2 weighted imaging showing an old infarction in the left centrum semiovale (1b)
Figure 2. Significantly abnormal magnetic resonance angiogram demonstrating Moyamoya disease and enlarged posterior circulation vessels. Narrowed calibre of internal carotid arteries from the origin with marked tapering at the terminations (2a, 2b). Enlarged calibre of external carotid arteries (2b) with prominent superficial temporal arteries (2a). The calibre of the posterior circulation vessels is enlarged (2a, 2b). Note is made of a hypoplastic right vertebral artery and small left posterior cerebral artery.

Abbreviations: R, right; L, left; ICA, internal carotid artery; CCA, common carotid artery; ECA, external carotid artery; VA, vertebral artery; BA, basilar artery; PCA, posterior cerebral artery; STA, superficial temporal artery.
Discussion

Hemifacial spasm usually results from direct vertebrobasilar vascular compression of the facial nerve causing secondary hyperexcitability of the seventh nucleus.\(^1\) Uncommonly compression results from tumours and cerebral imaging is recommended to verify the cause.\(^2\)

Treatment is conservative with anticonvulsants, botulinum toxin injection and microvascular decompression if conservative therapy fails.\(^1\) We postulate the vascular loop had developed from the compensatory flow through the posterior circulation resultant from the Moyamoya disease as evident by the relatively enlarged calibre of left vertebral and basilar arteries.

Moyamoya disease is characterised by progressive non-atherosclerotic vasculopathy of the intracranial carotid arteries.\(^3\) Approximately half of the adult patients present with haemorrhage while children usually present with transient ischaemic attack or ischaemic infarction.\(^3\)

To our knowledge there are no prior reports of Moyamoya disease presenting as hemifacial spasm although in retrospect his childhood right "hemichorea" may have been a manifestation of the left middle cerebral artery infarct seen on imaging. There are no randomised data to guide management of Moyamoya disease. Treatment with anti-platelets has a high rate of recurrence.\(^3\)

Both direct (superior temporal artery to middle cerebral artery) and indirect (encephalo-duro-arterio-synangiosis, encephalo-duro-myosynangiosis or multiple burr hole surgery) bypass surgery are acceptable alternatives to medical therapy and results from non-randomised series suggest long term efficacy in secondary stroke prevention.\(^3-5\)

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References: