Cerebrovasc Dis 2009;27:413–414

DOI: 10.1159/000209243

**Hemifacial Spasm and Pontine Compression
Caused by a Giant Vertebrobasilar Dolichoectasia***Spyridon Papapetropoulos^a, Andreas A. Argyriou^c,
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Hemifacial spasm (HFS), characterized by irregular, involuntary and recurrent tonic and clonic contractions of muscles innervated by the ipsilateral facial nerve [1], is usually attributed to compression of the facial nerve at the root entry zone by an aber-

rant artery or cerebellopontine angle masses [2, 3]. The offending vessels are usually arterial structures, such as the anterior or posterior inferior cerebellar arteries [4]. We present the case of a patient with HFS secondary to compression by a giant vertebrobasilar dolichoectasia (VBD) and highlight the possible association of HFS with VBD and type 1 neurofibromatosis (NF-1).

A 70-year-old man complained of a 3-year history of intermittent, involuntary muscle spasms involving the left lower and mid portions of his face. Approximately 18 months before presentation, he gradually developed mild speech difficulties that progressed to moderate dysarthria with nasality without diurnal variation or ocular symptoms. His past medical history was significant for NF-1 and renal cell carcinoma (postnephrectomy) and was negative for hypertension or hyperlipidemia.

Physical examination revealed multiple cutaneous neurofibromas and café au lait spots, with axillary freckling, confirming the diagnosis of NF-1 (fig. 1D). Neurological examination revealed intermittent spasms of the mid- and lower-left facial region consistent with left HFS. Mild dysarthria with nasality were also noted. The rest of the examination was otherwise normal. Plantar reflexes were flexors.

Fig. 1. A, B Magnetic resonance angiography images showing the giant vertebrobasilar ectasia (arrows). **C** T2-weighted axial magnetic resonance imaging confirming the pressure effects (arrows). **D** Multiple café au lait spots and neurofibromas (arrows).

Both brain magnetic resonance imaging and magnetic resonance angiography revealed a marked VBD. The tortuous and elongated vertebrobasilar vessels were seen to impinge on the left side of the caudal pons in the root exit zone for the facial nerve and brainstem (fig. 1A–C). Diffuse white matter lesions, especially in the posterior circulation, were also depicted on T2-weighted images. Muscle electrophysiology and muscle-specific tyrosine kinase antibody titer were normal.

The patient received botulinum toxin type A (2.5 U) in the left mid-facial and lower facial region, with alleviation of symptoms and no recurrence at a follow-up scheduled at 4 months.

The association between VBD and HFS has been previously reported [3]. Usually, there are no apparent causes for VBD. In elderly patients with diffuse T2W white matter lesions, it is reasonable to think that atherosclerosis is the main facilitating factor in the pathological interrelation between the constitutionally abnormal vessel and the 7th nerve. However, there was no evidence of hypertension or hyperlipidemia in our patient.

Our patient had been diagnosed with NF-1. NF-1 may cause neurological manifestations by several mechanisms, including compression effects from abnormal arteries and malignant nerve sheath growth and has also been described as a possible cause of HFS [5]. To our knowledge, there is one report assuming a casual relationship between NF-1 and HFS [6], while the association between NF-1 and intracranial vascular disease has also been previously described [7]. Moreover, the potential development of VBD in NF-1 has previously been highlighted [8]. In our case, we assume a causal rather than a casual relationship between NF-1 and VBD-induced HFS. However, a purely casual relationship cannot be excluded.

References

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