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BRIEF REPORT

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An unusual case of vascular loop syndrome

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Abstract Coexistence of hemifacial spasm (HFS) and trigeminal neuralgia (TN) is a rare entity known as painful tic convulsif (PTC). Here, we present a case of right-sided HFS after which left TN developed, which is an unusual form of PTC. Both disorders were caused by bilateral vascular compression of the cranial nerves and successfully treated with botulinum toxin and carbamazepine. As PTC is benign in nature and can be treated with botulinum toxin, neuroradiological investigations should be performed for an accurate aetiological diagnosis, particularly in young patients with atypical disease manifestations.

Keywords Hemifacial spasm • Trigeminal neuralgia • Vascular loop syndrome

Introduction

Coexistence of hemifacial spasm (HFS) and trigeminal neuralgia (TN), known as painful tic convulsif (PTC), is a rare entity and occurs particularly in women over the age of 50 years. It is usually caused by vascular compression of the ipsilateral fifth and seventh cranial nerves [1]. Vascular compressions in contralateral sides that caused HFS and TN had been previously emphasised in individual case reports [2].

Here, we present a case with HFS and contralateral TN caused by different aberrant vascular loops. The patient was successfully treated with botulinum toxin and carbamazepine

Case report

A 71-year-old male was admitted with a complaint of involuntary contraction and twitching of right eyelid that was spreading to the ipsilateral angle of the mouth and chin. Although he had had this complaint for the preceding 7 years, the intensity of symptoms had increased just before admission. The contractions were occurring spontaneously and sometimes extending into sleep. They were frequently aggravated by stress, anxiety and voluntary facial movements such as eye closure. He had had hypertension for 15 years. The neurological examination was normal with the exception of spasms of orbicularis oculi (O.oc) and orbicularis oris (O.or) muscles.

Complete blood count and serum biochemistry were normal. Needle EMG, which was carried out by means of concentric needle electrodes in O.or and O.oc muscles of right side, demonstrated irregular bursts of high-voltage motor unit potentials. Blink reflex was elicited by percutaneous electrical stimulation and was recorded by surface electrodes placed in O.oc. Magnetic resonance imaging (MRI) demonstrated an ectatic vessel causing impingement of the right facial nerve (Fig. 1A). As there was no response to previous pharmacological treatments, a total dose of 75 U botulinum toxin type A was injected in the central portion of the pretarsal O.oc of the upper eyelid, the reflection of the O.oc lateral to the orbital rim, the procerus and corrugator muscle group, and the mentalis as well as the zygomaticus major muscle; injections were made in lower doses in the contralateral parts to avoid asymmetry and his complaints completely disappeared within one week. As significant improvement occurred after the fifth day of botulinum toxin injection and lasted for 2.5 months, he needed botulinum toxin injections with 3-month intervals. After 3 years without any complaint, he had sudden, brief, severe electric-shock-like pain confined to the territory of the left trigeminal nerve. The pain was triggered by tactile stimuli such as chewing or brushing the teeth. Cranial MRI was repeated and a second aberrant vascular loop compressing the left trigeminal nerve entry zone was noticed (Fig. 1B). In addition to botulinum toxin injections similar to previous injections, he was also treated with carbamazepine 400 mg/day and his complaints diminished.

The development of HFS and TN was considered to be related to vascular loops localised on different sides.

Discussion

The clinical, electrophysiological and radiological findings of a patient with a 10-year history of HFS, which was responsive to botulinum toxin, and new-onset contralateral TN were presented.

TN is a relatively common disease of trigeminal nerve and presented with intermittent, shooting pain on the face. HFS is characterised by intermittent or continuous, painless, involuntary twitching of one side of the face. The development of TN following HFS may be associated with the bilateral vascular loops causing irritation of cranial nerves localised on different sides. Although several theories have been proposed to determine the aetiology, most reports concentrate on the anatomic relation between the nerves and the juxtaposed vascular loops. Compression at the root entry or exit zone of the cranial nerves is suggested to be responsible [1, 3]. However, vascular compression was also incidentally noted along the course of cranial nerves intraoperatively or in asymptomatic patients when MRI of the brainstem was

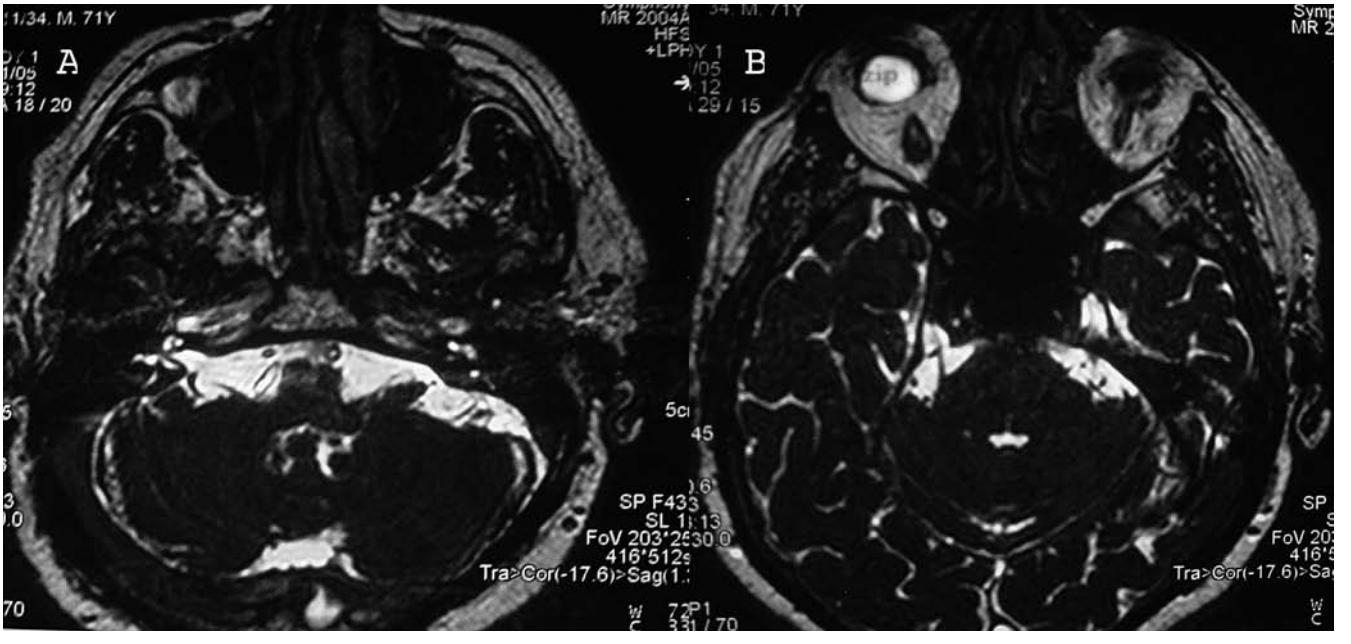

Fig.1 a MRI demonstrating an ectatic vessel causing impingement of the right facial nerve. **b** Aberrant vascular loop compressing the trigeminal nerve

performed for other reasons [4]. Therefore it is hypothesised that hyperactivity and hyperexcitability of the nervous structures as well as vascular impingement play a role in the development of symptoms [5]. Hypertension and ageing accelerate atherosclerotic changes, causing the formation of ectatic vessels and resulting in the development of vascular compression [1]. In this case, hypertension would be the risk factor in the formation of ectatic vessels and impingement of the fifth and seventh cranial nerves. However, the exact cause and effect of hypertension has not been clarified, as an ectatic vessel compressing the brainstem may result in hypertension [6].

Although different medical treatment options including carbamazepine, clonazepam, haloperidol and amitriptyline were previously used, the effect of these treatments was often transient. Definitive diagnosis of involuntary facial movements allows appropriate therapy and improves quality of life. In the present case both involuntary movements and pain in PTC was improved after the injection of botulinum toxin, as previously reported [7, 8].

As PTC is benign in nature and can be treated with botulinum toxin, neuroradiological investigations should be performed, particularly in young patients with atypical disease manifestations.

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