CASE STUDY

Painful Tic Convulsive as Manifestation of Vertebrobasilar Dolichoectasia and Aneurysm

Tic convulsivo doloroso como manifestación de dolicoestasia y aneurisma vertebrobasilar

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Received 2 January 2014; accepted 1 February 2014

The patient is a 64 year old male, with no clinical history of interest, who was assessed in a different centre 5 years ago after presenting with left hemifacial spasms (HFS, which initially manifested as blepharospasm and later affected the frontal and ipsilateral perioral muscle system). He was treated with Pramipexole and Clonazepam, which led to improvement. Two years ago the patient started having associated with clinical symptoms of trigeminal neuralgia (TN) which affected the left maxillary nerve branch. Nuclear magnetic resonance (NMR) showed a vascular lesion with compression of the anterior margin of the protuberance and left cerebellopontine angle (Figs. 1 and 2). The TN was controlled initially with analgesics and subsequently with Pregabalin 75 mg/8 h (Lyrica) and Amitriptyline 10 mg/24 h. The therapeutic option of an intravascular stent was considered, but was rejected due to its high risk factor. One year ago the patient was referred to our department with hearing loss, presenting a normal audiometric


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Figure 1 3D nuclear magnetic resonance imaging. Anatomy of the Willis polygon. Aneurismatic elongation and dilatation of the vertebrobasilar trunk is appreciated, with a maximum diameter of 18 mm (arrow).
examination. He continued with intermittent bouts of TN (2–3 episodes/year) which were controlled with Pregabalin 75 mg/24 h and continuous HFS which led to the indication of treatment with botulinum toxin. Injections of botulinum type A 100 IU/session were administered, divided into 6 injections at bilateral frontal level and mouth depressor angle and left side face level in the orbital muscle of the eye, nasolabial furrow, upper lip elevator and anterior platysma band. Following moderate improvement of the HFS, the treatment was repeated at 3 months and 5 months after the initial treatment the patient showed greatly improved clinical symptoms; mild sporadic HFS persisted in situations of stress due to motor activity. Over the 5 months of treatment there were no TN symptoms. It was decided to continue with follow-up and further injections every 4–6 months.

Discussion

HFS is defined by involuntary tonic–clonic contractions of the muscles on one side of the face innervated by the facial nerve.1 Facial HFS associated with TN is a rare disorder which was described for the first time by Cushing in 1920 under the name painful tic convulsive (PTC).2,3

Typically, there is no simultaneity of symptoms. HFS presents first and this is later followed by associated NT, with an interval which varies between a few months and several years. It is also common for HFS to begin with ocular symptoms and gradually spread to the ipsilateral perioral musculature.3,4 Both circumstances present in our clinical case.

The majority of PCT cases are caused by aberrant arteries, arteriovenous malformations and cerebellar pontine angle tumours. Other rarer causes exist, such as type 1 Arnold–Chiari malformation or demyelinating diseases such as multiple sclerosis.1–3

With regards to the involvement of aberrant arteries, in the majority of cases the anterior inferior and posterior inferior cerebellar arteries are responsible, whilst involvement of other arteries such as the superior cerebellar artery or the vertebrobasilar artery is rare.1,4

Dolichoectasia is diffuse vascular dilatation with elongation and twisting of the affected vessels. At intracranial level the most affected area is the vertebrobasilar segment. Although vertebrobasilar dolichoectasia (DVB) is frequently asymptomatic, various secondary symptoms such as ischaemia, compression or on rare occasions rupture may present.6

In our case we identified a lesion in the vertebrobasilar artery, which is highly uncommon, and leads to exceptional symptomology such as that of TCD, as a consequence of the compression of the V and VII cranial nerves.

NMR, with the use of angiographic sequences, such as TOF (time of flight) (Fig. 2) or CISS (constructive interference at steady state)7,8 must be considered as the initial procedures for diagnosis in TCD evaluation, which enable the detection or exclusion of other causes such as neoplasias or multiple sclerosis at the same time.1

The treatment of DVB is a controversial issue due to the potentially serious risks in treatment approach. Antiplatelet agents or anticoagulants are used in cases of known previous ischaemic events, but are not indicated in the presence of aneurisms.6 In terms of surgery, an endovascular approach or microvascular decompression via retrosigmoid craniotomy can be used, with the insertion of Teflon or some other material between the vessels and the affected cerebrum tissue.6,9

The injection of botulinum toxin is the procedure of choice for HFS1 treatment, as it is effective and safe and offers moderate improvement for between 85% and 95% of patients. With regards to TN in patients with PCT treated with botulinum toxin, better pain management has been demonstrated in patients even though they do not respond to pharmacological treatment, with the toxin acting on the central nervous system mediated by afferent pathways possibly originating in the muscle spindles.3 The possible adverse effects of treatment are temporary and include facial paralysis (23%), diplopia (17%) and ptosis (15%).1,5,7 We therefore consider this to be the procedure of choice in patients with mild to moderate symptoms or who present a high surgical risk.

Conflict of Interests

The authors have no conflict of interests to declare.
References