

## Title

Bilateral painful tic convulsif

## Authors

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**PATIENT CONSENT – obtained**

## **SUMMARY**

‘Painful tic convulsif’ (PTC) describes the co-existence of hemi-facial spasm and trigeminal neuralgia. In this report, we describe a unique presentation of bilateral PTC in a gentleman with bilateral hemi-facial spasm and trigeminal neuralgia secondary to neurovascular conflict of all four cranial nerves. Following failed medical and radiofrequency therapy, microvascular decompression of three of the four involved nerves was performed, where the offending vessels were mobilised and Teflon used to prevent conflict recurrence. He continues to respond to Botox for right hemi-facial spasm. Since surgery he remains pain-free bilaterally and spasm-free on the left.

## **BACKGROUND**

This report illustrates the rare association of hemi-facial spasm and trigeminal neuralgia in a condition known as ‘Painful tic convulsif’ (PTC), initially described by Cushing in 1920.<sup>1</sup> Since then, there have been relatively few literature reports. Although there have been isolated reports of bilateral hemi-facial spasm and unilateral trigeminal neuralgia, we have failed to find a description of co-existing bilateral hemi-facial spasm and bilateral trigeminal neuralgia. Moreover, this case report also highlights the efficacy of microvascular decompression (MVD) in patients with evidence of neurovascular compression following failed medical therapy. In our patient, neurovascular conflict was present with all four relevant cranial nerves with different offending vessels at each site.

## **Case Presentation**

A 57-year-old previously healthy gentleman presented with classical symptoms of left V1 trigeminal neuralgia. Following initial presentation, he developed left hemi-facial spasm 5-years later. At the age of 64 he then developed right V2 trigeminal neuralgia and subsequently developed right hemi-facial spasms 8 years later at the age of 72.

## **Investigations**

MRI with dedicated FIESTA sequence of the trigeminal nerve revealed neurovascular impingement of the trigeminal and facial nerves on both sides (Figure 1).

## **Treatment**

Initial management with carbamazepine and regular Botulinum neurotoxin (BoNT) injections was successful. However, symptoms of both conditions recurred and additional medications, including baclofen, tramadol and amitriptyline failed to yield good results, even at maximal doses. He went on to receive 3 sessions of radiofrequency ablation of the Gasserian ganglion, twice on the left and once on the right, which only provided temporary relief.

At age 75, right trigeminal MVD was performed without complication. At surgery, a loop of the superior cerebellar artery (SCA) was found to be “grooving the superior and medial aspect of the right trigeminal nerve (Figure A). The offending vessel was dissected away from the surrounding arachnoid and the loop mobilised away from the nerve. Teflon was used to secure the vessel position and prevent recurrent neural conflict. He elected not to have decompression of right facial nerve at the same time since right-sided spasms were not troublesome, being well controlled with BoNT injections.

Seven months later, he underwent MVD of the left trigeminal and facial nerves during the same procedure. The left trigeminal nerve was compressed proximally by a loop of the SCA while the cisternal portion of the left facial nerve was distorted by a medially-placed loop of the anterior inferior cerebellar artery (AICA). Once again, the offending vessels were mobilised from the surrounding arachnoid and interposition of Teflon sponge used to prevent recurrent contact between the offending vessel and nerve.

## **Outcome and follow-up**

Surgery resulted in immediate and complete resolution of bilateral trigeminal neuralgia and left hemi-facial spasm. He remains pain free bilaterally and spasm free on the left, off medical treatment 8 months following the last MVD procedure. He continues to respond to BoNT for right hemi-facial spasm.

## **Discussion**

Hemi-facial spasm and trigeminal neuralgia are hyperactive disorders of the cranial nerves that affect the face and are characterised by paroxysms of involuntary contractions and pain respectively<sup>2</sup>. Independently, these disorders have been reported to have a prevalence of 11<sup>3</sup> and 12.6-28.9<sup>4</sup> per 100,000 per year, respectively.

When existing concurrently they form a rare and debilitating syndrome known as PTC, present in less than 1% of rhizopathies. The most common cause for combined hyperactive dysfunction syndromes of the cranial nerves is neural compression secondary to tumours, cholesteatoma or vascular compression.<sup>5</sup>The latter is the most frequently reported cause for isolated compression, most often due to vertebro-basilar ectasia, AICA, PICA or SCA compression.

We found only 78 reported cases of PTC in the literature<sup>6</sup>, including two reports on bilateral hemi-facial spasm and unilateral trigeminal neuralgia.<sup>7,8</sup> However, to our knowledge, this is the first report of bilateral hemi-facial spasm and bilateral trigeminal neuralgia. Neurovascular conflict was present with all four relevant cranial nerves with different offending vessels at each site. *Both trigeminal nerves were in conflict with the respective superior cerebellar arteries and both facial nerves were in conflict with the respective anterior inferior cerebellar arteries.*

The first line treatment for HFS is BoNT, given periodically (every 3-6 months) via intramuscular injections, while pharmacological treatment is first line for trigeminal neuralgia, initially with carbamazepine. Trigeminal ablation procedures (including radiofrequency, glycerol, balloon compression and gamma-knife therapies) offer alternatives to medication in trigeminal neuralgia. In cases of failed medical therapy, MVD offers the best long term outcome of all the surgical procedures when neurovascular compression of the relevant nerve is present.<sup>9</sup>The literature also suggests a very low long term recurrence rate for both conditions following 6 months of symptom freedom<sup>10</sup>.

## References

- <sup>1</sup>Cushing H. Major trigeminal neuralgia and their surgical treatment. Am J Med Sci. 1920;160:157-184.
- <sup>2</sup>Dou NN, Hua XM, Zhong J, Li ST. A successful treatment of coexistent hemifacial spasm and trigeminal neuralgia caused by a huge cerebral arteriovenous malformation: a case report. J Craniofac Surg. 2014;25:907–910.
- <sup>3</sup>Rosenstengel C, Matthes M, Baldauf J, Fleck S, Schroeder H. Hemifacial spasm: conservative and surgical treatment options. Dtsch Arztebl Int. 2012;109(41):667-73.
- <sup>4</sup>van Hecke O, Austin SK, Khan RA, Smith BH, Torrance N. Neuropathic pain in the general population: A systematic review of epidemiological studies. PAIN 155. 2014;654–662

<sup>5</sup>Fonoff ET, Araújo VP, de Oliverira YSA, Teixeira MJ. Neurovascular compression in painful tic convulsif. *Acta Neurochir.* 2009;151:989-993.

<sup>6</sup>Revuelta-Gutiérrez R, Velasco-Torres HS, Vales Hidalgo LO, Martínez-Anda JJ. Painful tic convulsif: Case series and literature review. *Cirugía y Cirujanos.* 2016;84(6):493-498.

<sup>7</sup>Felicio AC, Godeiro Cde O, Borges V, Silva SM, Ferraz HB. Bilateral hemifacial spasm and trigeminal neuralgia: a unique form of painful tic convulsif. *Mov Disord.* 2007;15;22(2):285-6.

<sup>8</sup>Boscá-Blasco ME, Burguera-Hernández JA, Roig-Morata S, Martínez-Torres I. Tic convulsivo doloroso y toxina botulínica / Painful tic convulsif and botulinum toxin. *Rev Neurol.* 2006;42(12): 729-732

<sup>9</sup>Kalkanis SN, Eskandar EN, Carter BS, Barker FG II. Microvascular decompression surgery in the United States, 1996 to 2000: mortality rates, morbidity rates, and the effects of hospital and surgeon volumes. *Neurosurgery.* 2003;52(6):1251–1262

<sup>10</sup>Zakrzewska JM, Linskey ME. Trigeminal neuralgia. *BMJ.* 2015;350:h1238.

### **Learning points**

- ‘Painful Tic Convulsif’ is a rare condition and refers to the co-existence of hemi-facial spasm and trigeminal neuralgia. This is the first published case report of bilateral tic convulsif.
- Aetiological factors for the condition include tumours, infections, demyelination with neurovascular compression being the most common underlying cause.
- Microvascular decompression is the gold standard treatment in patients with identifiable neurovascular conflict.
- Microvascular decompression is an effective treatment with a favourable outcome and low complication rates in experienced hands.

## **Patient's Perspective**

I remember that the attacks of pain started around 1994 and they affected only the left side of my face. At first the attacks did not last long and happened every four, six and even twelve months at a time. Between one attack and another I felt fine and carried on with my life as normal so that I did not take any notice of the pain and brushed it off.

Then the attacks of pain started to become more frequent and the time between one attack and another became shorter. The pain I felt was like a lash from a whip spreading from my forehead to my lower jaw. It was intense and even the simplest things could bring it on and make it worse, for instance washing my face was agony – the pain was so severe it would bring me to my knees. Talking, eating, brushing my teeth and shaving also triggered the pain.

In 1998, I visited a consultant neurologist who assessed me and prescribed medication. Around this time, I also started to experience a spasm on the left side of my face. At first I thought it was related to the pain since the spasm was happening on the same side of my face. I did not think much else of it since controlling the pain was a priority. But the spasm started to affect my eye giving me a tingling sensation and making me wink involuntarily. Although I tried my best to ignore it, I was sometimes embarrassed to speak to people because of the spasm. The consultant neurologist advised me to visit an ophthalmologist who started me on regular botox injections to control the spasm on the left side of my face. On starting the botox injections the spasm improved significantly.

My symptoms were reasonably controlled to the extent that I had stopped taking medication. However, in 2005, the pain came back with a vengeance. This time it was not only affecting the left side of my face but also affected the right side. Sometimes the pain was only on one side of the face and sometimes both sides were affected at the same time. I went back to the consultant neurologist who advised me to start having medication again as before.

In 2008, as the pain got worse, I was referred to the Pain Clinic. I underwent trigeminal pulse radio-frequency ablation initially on the left side and then on both sides of my face. At the beginning, it was very successful so much so that I was invited to speak at a local press conference about my positive experience with the treatment.

However, in 2013 I started to feel the pain again on both sides and it was very severe. Around this time, I also started to notice a spasm affecting the right side of my face and after about a month of trying to cope, I sought advice from a consultant neurologist. He prescribed me medicine including baclofen, carbamazepine and pregabalin. But the pain never went completely away on the medication. I had good days and bad days which required the

neurologist to increase and decrease the dose of my medication according to the level of my pain.

Seeing I had made little progress with the medication, in December 2015 the neurologist decided to speak with a consultant neurosurgeon working in London about my problem. I had never met him before but I was willing to do anything to feel better. I had an MRI of my head done and on the January 2016 I had a meeting with the neurosurgeon. After explaining to me the cause of my problem we decided to go for surgery, first to try to eliminate the pain from the right side of my face and then from the left.

The first operation was carried out in February 2016 on the right side. On waking up from anaesthesia, the pain on the right side had disappeared and I did not feel it again since then. Since I still needed the medication for the pain on the left side, I underwent another operation in August 2016. This time the neurosurgeon decided to tackle the spasm on the left side too. Again on waking up from anaesthesia both pain and the spasm on the left side had disappeared. On both occasions, the surgeons, doctors, nurses and all the staff in the operating theatre and in the ward were very helpful towards me. After all that I have been through over the years, I truly believe that these two doctors have been sent to me from heaven because since the surgery, thank God, I have remained pain free on both sides.

## Figure Legends

### Figure 1:

Axial FIESTA MRI sequence at 1.5T. A & B: Oblique axial section through the trigeminal nerves at the level of the Pons. A loop of the superior cerebellar artery can be seen “grooving” the superior surface of the trigeminal nerve on the right and abutting the medial aspect of the trigeminal nerve on the left. C & D: Oblique axial section through the VII and VIII cranial nerves. The anterior inferior cerebellar arteries can be seen in conflict with the facial nerves, its proximal portion on the right and its cisternal portion on the left. The Vth and VIIth nerves are highlighted in yellow in B & D with conflicting vessels highlighted in red. Ba: Basilar artery; Co: cochlea; IAM: Internal Auditory Meatus; Meck: Meckel’s cave, PMJ: pontomedullary junction.

