A Unique Case of Microvascular Decompression for Combined Trigeminal Neuralgia, Hemifacial Spasm and Glossopharyngeal Neuralgia

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Introduction

Trigeminal neuralgia (TN), hemifacial spasm (HFS), glossopharyngeal neuralgia (GPN) and vestibular paroxysmia (VP) are hyperactive dysfunction syndromes (HDSs) theorized to be due to microvascular compression of the root entry zone (REZ) of the cranial nerves. Cases of combined HDS involving two or more of these disorders is extremely rare. The mean age at presentation for combined HDSs is 52 years old with an initial complain of facial spasm followed by TN and/or GPN several years later. The combined symptoms can occur at the same side but a vast majority develop symptoms on the contralateral side and are related to tortuous vertebrobasilar artery. Most of the cases fail initial medical therapy leading the patient to seek surgical intervention with a microvascular decompression with the use of Telfa between the offending vessel and the cranial nerves affected.

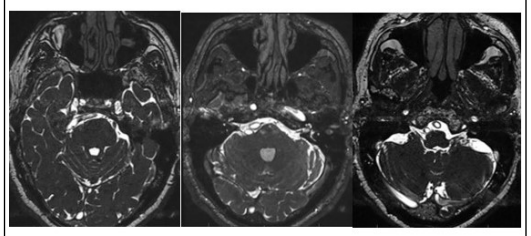
In this case report, we present the rare case of co-existent right synchronous TN-HFS-GPN due to a dolichoectactic vertebro-basilar system. We describe the successful surgical treatment via a retrosigmoid approach for microvascular decompression with a vertebrobasilar artery sling.

Case Presentation

A 66-year-old man with a past medical history of Diabetes Mellitus type 2, hyperlipidemia, hypertension, and coronary artery disease, presented with a 3-year history of severe disabling right sided facial pain that was progressively worsening over the last 6 months prior to presentation. The patient described the pain as located in the right posterior tongue radiating into the jaw and was described as paroxysms of electric shock like pain with parasthesias of the right face and ear. His episodes were precipitated by swallowing, chewing and speaking. The patient also complained of tremors of the constant right eyelid. On examination, speech was muffled due to in an inability to open his mouth. Ongoing tremors were appreciated in the right lower eyelid. The patient was otherwise neurologically intact.

On pre-operative imaging, a brain MRI without gadolinium with CISS sequence (Fig 1) showed radiological evidence of a severely dolichoectatic basilar artery with both vertebral arteries joining on the right side compressing the 9th nerve, the 7th nerve as well as the 5th nerve. There were no other pathologic lesions identified in the area. Patient failed medical therapy and these symptoms where severly affecting his activities of daily living, so we proceeded with a microvascular decompression via a retrosigmoid approach.

FIG 1. A, B, C



Axial T2- weighted image CISS sequence (0.5 mm). Note the tortuous vertebral arteries on the right-side displacing the TN (A), the FN (B) and near the GN (C).

Operation and Post-operative care

Patient was positioned in the lateral decubitus position with the right side up with a 3-pin site head holder. A standard retrosigmoid approach was used with an extended bone flap from the transverse sinus down to the foramen magnum, but not going through it. The dura was then opened with its the flap was put under traction medially. The microscope was brought into the field and CSF drainage from the cerebellomedullary cistern was obtained. The dissection of the arachnoid starting at the 11th nerve in the cerebellomedullary fissure and continued all the way to the tentorium. Right vertebral artery was severely convex towards the right side pushing on the anterolateral medulla, although it was not pushing directly on the 9th or 10th nerve.

The PICA origin was hooked around the 12th nerve A t the exit zone of 7th and 8th nerves, the right vertebral artery was digging into the pontomedullary junction and rostrally it was compressing the trigeminal nerve before it turned away to join the other vertebral artery just below the tentorium at Meckel's cave region to become the basilar artery.

To be able to decompress all 3 points of compression a Gore-Tex sling was placed around the right vertebral artery and used one sutured to attach it to the petrous dura in an anterolateral direction and then another sutured was fixed to another portion petrous dura and used angled permanent Sugita aneurysm clip to join both flaps of the Gore-Tex just behind the vertebral artery. Careful attention was placed to not kink the above-mentioned artery. Several pieces of shredded Teflon implants were between the trigeminal nerve and the basilar artery, the 7th nerve and the PICA, and between the medulla and the right vertebral artery. Closure was followed in its usual fashion.

Patient was seen at our clinics 2 weeks and 6 weeks after surgery with all symptoms resolved with an intact neurological exam. No post operative complications were observed.

Discussion

Combined HDS is very rare. In two large cohorts of HDS from Kobata et al., and Yang et al., combined HDS prevalence was 2.8% (41 of 1,472 patients) and 2.97% (51 of 1,720) respectively. Bilateral compression of the same CN is the most frequent presentation of Combined HDS (15 of 41 and 37 of 51) and the onset is usually metachronous (44 of 51). The combination of TN-GPN is uncommon (3 of 41) and involvement of more than two nerves is not reported in these studies. Association of three cranial nerves or more has only been reported a few times in the literature. Paroxysmal supraventricular tachycardia (PSVT) as a result of vascular compression of the CN IX and X has been found in one case report thus far. Hypertension and atherosclerotic changes are the main risk factors for compression of multiple cranial nerves as they facilitate elongation and increase of redundancy of the vessels,which may lead to dynamic compression of multiple cranial nerves. Individual susceptibility and race are also considered as possible risk factors.

A study in 34 Japanese patients with HFS revealed a small posterior cranial fossa with a narrow cerebellopontine angle cistern resulting in more crowded CN and vascular structures compared to control groups.

In most cases of dual HDS there are multiple offending vessels, similar to those in single HDS. However a torturous dolichoectactic VBA shifted to the affected side can lead to compression of multiple cranial nerves resulting in combined HDS. Microvascular decompression is still the treatment of choice for these cases with highly curative rates. It is recommended to move the VA proximally before the dissection which should start from the caudal cranial nerves. Most of the cases reported are treated succesfuly via a microvascular decompression with Teflon implants but to our knowledge this is the first TN, HM and GPN treated via vertebrobasilar artery sling.

Conclusion

Combined TN-HFS-GPN is rare and may be associated with a dolichoectatic vertebrobasilar system. In such cases, microvascular decompression via a retrosigmoid approach should be considered as a safe and effective option for transposition of the offending artery and decompression of the affected nerve roots via vertebrobasilar artery sling.

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