Case Report

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Others Section

Concurrent Trigeminal Neuralgia and Hemifacial Spasm due to Anterior Inferior Cerebellar Artery Tortuosity: A Rare Case Report

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ABSTRACT

The Anterior Inferior Cerebellar Artery (AICA) plays an important role in the pathophysiology of both Hemifacial Spasm (HFS) and Trigeminal Neuralgia (TN). The AICA follows a tortuous course, which may affect the facial nerve and trigeminal nerve by compressing these structures. This compression is the common underlying aetiology for both TN and HFS. To date, TN remains an idiopathic condition that is hypothesised to be caused by an underlying irritative lesion involving the trigeminal ganglion. TN is characterised by paroxysmal, lancinating, unilateral pain, often localised along the maxillary (V2) and mandibular (V3) divisions of the trigeminal nerve. In contrast, HFS manifests as involuntary, irregular, or tonic contractions of the facial muscles. It typically begins in the orbicularis oculi and progressively spreads to involve the perioral, platysmal and lower facial muscles. Unlike TN, HFS persists even during sleep and worsens over time. The present case report discusses a 65-year-old male who presented a rare combination of TN characterised by continuous pain and HFS; the concomitant occurrence of both conditions is unique in itself. The present case demonstrates how a dilated, tortuous AICA loop causes a particularly peculiar presentation in a patient, involving both TN and HFS, along with the surgical management and treatment that facilitated a good prognosis for the diagnosis. Additionally, it highlights the rarity of these manifestations stemming from a single aetiological agent, specifically a dilated Anterior Inferior Cerebellar Artery (AICA) loop. Furthermore, it underscores the importance of early diagnosis, neuroimaging and timely surgical intervention, particularly when addressing uncommon neurovascular conflicts that manifest with overlapping cranial neuropathies.

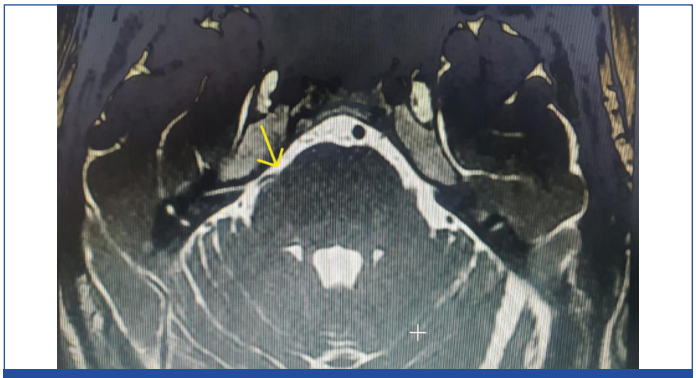
Keywords: Anterior inferior cerebral artery loop, Arteriovenous malformation, Idiopathic, Tonic contractions

CASE REPORT

A 65-year-old male presented to the Outpatient Department (OPD) with complaints of severe, excruciating pain on the right-side of his face, accompanied by Hemifacial Spasms (HFS) for one year. The patient reported the intensity of the pain to be 10 on the pain scale, which was triggered by smiling and other daily activities, such as brushing his teeth and consuming food. Due to the ongoing orofacial pain, the patient was initially managed with dental interventions. He was prescribed carbamazepine, which he continued for the last six months at a dosage of 400 mg once daily, but there was no symptomatic relief. Following the failure of this medication, the patient reported that the pain had become so severe that he attempted to commit suicide after experiencing no remission following the course of conservative management. He was advised to undergo further dental treatments, but again, no symptomatic relief was achieved.

Upon neurological examination, the Temporomandibular Joint (TMJ), muscles of mastication and intraoral findings revealed no abnormalities. Light probing of the left cheek with a cotton swab and lifting of the overlying skin upwards provoked a piercing pain lasting for a few seconds. The pain was localised to the vertex and did not radiate to the postauricular region. Magnetic Resonance Imaging (MRI) imaging with the Fast Imaging Employing Steady-state Acquisition (FIESTA) sequence revealed a prominence of sulcal spaces, sylvian fissure, cisternal spaces and cerebellar folia suggestive of atrophic changes [Table/Fig-1]. Additionally, significant dilation and tortuosity of the right AICA loop were noted, which was found to be compressing the entry zones of the 5th Cranial Nerve (CN).

A diagnosis of Trigeminal Neuralgia (TN) was made. Following the primary diagnosis, conservative management with carbamazepine

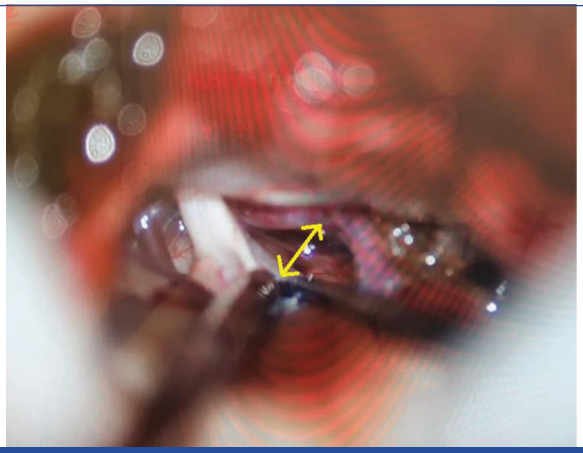


[Table/Fig-1]: FIESTA sequence of MRI Brain s/o arterial loop just at the root entry zone of right 5th CN (Yellow arrow).

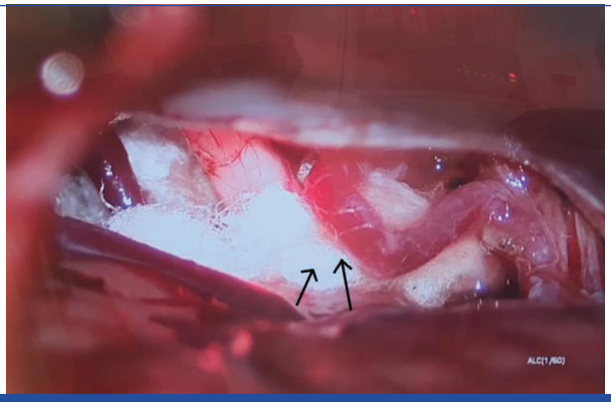
was initiated according to protocol and literature and the patient was started on tablet carbamazepine at a dosage of 100 mg twice daily. However, two weeks later, as the paroxysmal pain had not subsided, the patient's dosage was increased to 200 mg twice daily. Despite this adjustment, pain control remained a challenge, necessitating a referral to the Department of Neurosurgery. The patient was subsequently scheduled for surgical decompression to relieve the compression.

The patient underwent a right Retromastoid Suboccipital Craniotomy (RMSO) and Microvascular Decompression (MVD) of the 5th [Table/Fig-2] and 7th cranial nerves [Table/Fig-3]. The surgery was performed using a retro-sigmoid approach, located inferior to the transverse sinus and medial to the sigmoid sinus. The arachnoid membrane was dissected after retracting the cerebellar hemispheres. The right tortuous AICA loop was visualised compressing the entry zones of the 5th and 6/7 nerves from the lateral aspect. The degree of tortuosity of the right AICA loop restricted the decompression of the nerve via

the conventional approach. Therefore, the loop was approached from the level of the medulla to the trunk of the vertebral artery and a Teflon felt was suspended. The tortuous, hairpin-like loop of the right AICA compressing the root entry zones of the 5th and 7th cranial nerves along its course was mobilised and separated. The Teflon felt was interposed to maintain this separation [Table/Fig-4]. The incision was then closed in the standard fashion.



[Table/Fig-2]: MVD of 5th CN during right RMSO craniotomy.

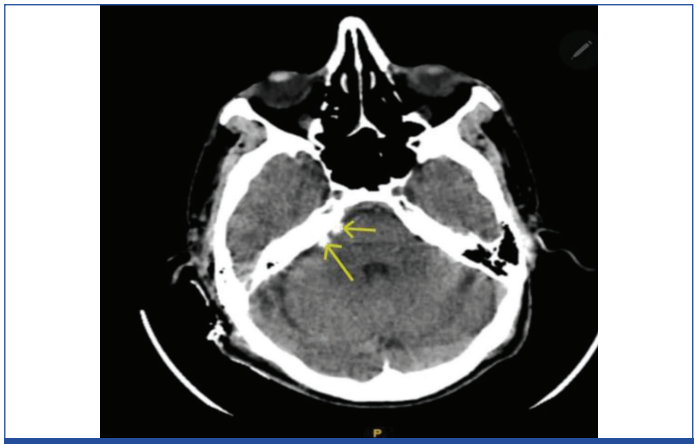


[Table/Fig-3]: MVD of 7th CN during right RMSO craniotomy.

Post-surgery, the patient reported immediate relief from pain and HFS, followed by a 100% successful recovery. The patient was discharged one week later after complete mobilisation, with regular follow-ups every two weeks. After six months of follow-up, the patient reported no further episodes of pain or facial spasms and expressed satisfaction with the medical treatment.

DISCUSSION

This article provides a detailed account of a rare case of Trigeminal Neuralgia (TN) with Hemifacial Spasm (HFS) caused by a dilated and tortuous right AICA loop. According to the literature, TN can arise from many possible aetiologies. Neurovascular conflict is by far the most accepted theory for TN, alongside several others, such as root compression or traction by the Superior Cerebellar Artery (SCA) on the 5th cranial nerve and dysfunction of cortical mechanisms of pain modulation, the brainstem and the basal ganglia [1,2].



[Table/Fig-4]: Postoperative Computed Tomography (CT) showing Teflon felt (seen as hyperintensity) around the right 5th CN.

The HFS is characterised by unilateral dysfunction of the facial nerve, which manifests as intermittent, painless spasms of the orbicularis oculi muscle. Other muscles involved in facial expressions may eventually become affected as the frequency and severity of dysfunction increase. There is extensive evidence supporting the hypothesis that dysfunction of the seventh cranial nerve is caused by compression from loops of the Posterior Inferior Cerebellar Artery (PICA), AICA, vertebral artery, or cochlear artery [3,4]. In this patient, it was observed that both TN and HFS are caused by the same aetiology: a tortuous AICA loop compressing the 5th and 7th cranial nerves. To date, no such case has been recorded in the literature, potentially making this the first instance.

Hyperactive Dysfunction Syndrome (HDS) is a neurovascular condition characterised by cranial nerve root entry zone compression due to a vascular aetiology. The concomitant presentation of TN with HFS is a variant of this syndrome [5]. Combined cranial nerve HDS is a rare condition, comprising only 3% of reported HDS patients. Several studies reveal a prevalence of 7.3% (3 of 41 combined HDS cases), 13.5% (5 of 37 combined HDS cases), 27.4% (14 of 51 combined HDS cases) and 31.8% (14 of 44 combined HDS cases) for the co-existence of TN and HFS among cranial nerve cases [1-4].

It was found that less than 3% of the population has concurrent TN and HFS [1]. A total of 10 cases, including the one presented here, have been reported for TN and HFS due to vertebrobasilar dolichoectasia over the decade (2013-2023) [Table/Fig-5] [6-12]. Furthermore, there is only one reported case in the literature where both TN and HFS are associated with a large Arteriovenous Malformation (AVM), with symptoms of both spasm and pain gradually relieved following endovascular embolisation of the nidus [13].

Author name	Place	Age/Gender	Duration	Treatment	Outcome
Lakhan SE [6] 2013	Cleveland, Ohio, United States of America	64/M	2.1 years	Pain relief was done with gamma knife surgery and botulinum injection was given for spasms. Conservative treatment with carbamazepine, oxcarbazepine and levetiracetam had failed	Complete improvement.
Revuelta-Gutiérrez R et al., [7] 2016	Mexico, United States of America	64/M, 75/F	Case 1: 2-4 years, case 2: 2 years	Case 1: MVD. Case 2: MVD techniques were used	After surgery, a complete symptomatic resolution was seen in both patients.
Han J et al., [8] 2018	Hengshui, China	65/F	3 years	Pharmacotherapy, carbamazepine, vitamin B1, methylcobalamin	Symptomatic improvement after a week but a right HFS was noted 5 months later, followed by a loss of follow-up by the patient over the next 6 months. No new treatment was initiated.
Rodriguez CA et al., [9] 2019	Brazil	79/M	4 months	NA	MRI with angiography revealed left-sided compression of 5, 7 and 9 cranial nerves by vertebrobasilar dolichoectasia.
Perez-Roman RJ et al., [10] 2020	Miami, Florida, United States of America	66/F	NA	The MVD method was used and the clip-sling technique was used to achieve MVD	The patient had immediate complete relief from TN, HFS and Glossopharyngeal Neuralgia (GPN) postoperatively.

Liu JJ et al., [11] 2022	Portland, Oregon, United States of America	67/M, 79/M	Case 1: 2.5 years, case 2: 6 months	MVD, a novel technique was described, involving the anterolateral mobilisation of the vertebrobasilar system posteriorly via a sling for brainstem decompression	Both patients had immediate radiographic improvement in the degree of brainstem compression and significant improvement after two months.
Alotaibi RF et al., [12] 2022	Dammam, Saudi Arabia	78/F	2.2 years	Pharmacotherapy, vertebrobasilar artery dilatation improved by medical therapy with carbamazepine	Initiate medical treatment first; it may be effective and may help avoid, especially in comorbid patients, patients with a contraindication to surgical intervention.
Present case (2023)	Maharashtra, India	59/M	5 months	MVD: VA trunk approached from the level of the medulla using techniques such as gradual displacement of the VA trunk via the insertion and suspension of Teflon felt (TachoSil)	Immediate resolution of the paroxysmal pain and HFS in the postsurgical period with no reported complications.

[Table/Fig-5]: Studies reported TN and hemifacial spasms due to vertebrobasilar dolichoectasia over the decade (2013-2023) [6-12].

CONCLUSION(S)

Clinicians need to recognise a dilated AICA as a rare and singular cause of the concomitant presentation of Trigeminal Neuralgia (TN) and Hemifacial Spasm (HFS). With the present case, the authors aimed to emphasise the importance of early clinical diagnosis and awareness regarding the appropriate course of treatment, as conservative management provides no symptomatic relief. An early diagnostic MRI as a first-line investigation for this condition may facilitate prompt diagnosis before the onset of other symptoms, offering immediate relief and a better prognosis for the patient.

REFERENCES

[1] Cao J, Jiao J, Du Z, Xu W, Sun B, Li F, et al. Combined hyperactive dysfunction syndrome of the cranial nerves: A retrospective systematic study of clinical characteristics in 44 patients. World Neurosurg. 2017;104:390-97. Doi: 10.1016/j.wneu.2017.05.020.

[2] Kobata H, Kondo A, Iwasaki K, Nishioka T. Combined hyperactive dysfunction syndrome of the cranial nerves: Trigeminal neuralgia, hemifacial spasm, and glossopharyngeal neuralgia: 11-year experience and review. Neurosurg. 1998;43(6):1351-61.

[3] Yang KH, Na JH, Kong DS, Park K. Combined hyperactive dysfunction syndrome of the cranial nerves. J Korean Neurosurg Soc. 2009;46(4):351-54. Doi: 10.3340/jkns.2009.46.4.351.

[4] Zhang YQ, Yu F, Zhao ZY, Men XZ. Combined hyperactive dysfunction syndrome of the cranial nerves: Analysis of 37 cases and literature review. World Neurosurg. 2019;129:e650-e656. Doi: 10.1016/j.wneu.2019.05.237.

[5] Hokazono M, Shimogawa T, Nakamizo A, Yoshimoto K. Hemifacial spasm caused by vascular compression of the anterior inferior cerebellar artery-posterior inferior cerebellar artery common trunk anomaly at the cisternal portion of the facial nerve: A case report. NMC Case Rep J. 2023;10:253-57. Doi: 10.2176/jns-nmc.2023-0125.

[6] Lakhan SE. Teaching neuro images: Concurrent hemifacial spasm and trigeminal neuralgia due to vertebrobasilar dolichoectasia. Neurol. 2013;81(8):e52. Doi: 10.1212/WNL.0b013e3182a1aaeb.

[7] Revuelta-Gutiérrez R, Velasco-Torres HS, Hidalgo LO, Martínez-Anda JJ. Painful tic convulsif: Case series and literature review. Cir Cir (English Edition). 2016;84(6):493-98. Doi: 10.1016/j.circir.2015.08.012.

[8] Han J, Wang T, Xie Y, Cao D, Kang Z, Song X. Successive occurrence of vertebrobasilar dolichectasia induced trigeminal neuralgia, vestibular paroxysmia and hemifacial spasm: A case report. Medicine. 2018;97(25):e11192. Doi: 10.1097/MD.0000000000011192.

[9] Rodriguez CA, Mello HD, Fustes OJ, Teive HA. Painful tic convulsive as manifestation of vertebrobasilar dolichoectasia. Arq Neuropsiquiatr. 2019;77(6):445-46. Doi: 10.1590/0004-282X20190042.

[10] Perez-Roman RJ, Chen SH, Sur S, Leon-Correa R, Morcos JJ. A unique case of microvascular triple decompression for combined simultaneous trigeminal neuralgia, hemifacial spasm, and glossopharyngeal neuralgia because of the dolichoectatic vertebrobasilar system. Oper Neurosurg. 2020;18(6):692-97. Doi: 10.1093/ons/opz205.

[11] Liu JJ, O'neill BE, Mazur-Hart D, Uluc K, Dogan A, Cetas JS. A technical note on anterolateral mobilization in vertebrobasilar dolichoectasia for relief of brainstem compression. J Cerebrovasc Endovasc Neurosurg. 2022;24(3):297-302. Doi: 10.7461/jcen.2022.E2020.11.004.

[12] Alotaibi RF, Al Mudhi AH, Almousa AM, Albaqshi MA, Alturki M. Trigeminal neuralgia and hemifacial spasms due to vertebrobasilar dolichoectasia: A case report. Cureus. 2022;14(12):32148. Doi: 10.7759/cureus.32148.

[13] Inoue T, Shima A, Hirai H, Suzuki F, Matsuda M. Trigeminal neuralgia due to red vein draining a supratentorial arteriovenous malformation: Case report. J Neurol Surg Rep. 2016;77(03):e109-12. Doi: 10.1055/s-0036-1584817.

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