BRAIN
ORIGINAL ARTICLE

Prevalence and severity of neurovascular compression in hemifacial spasm patients

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Hemifacial spasm is typically caused by vascular compression of the proximal intracranial facial nerve. Although the prevalence of neurovascular compression has been investigated in a cohort of patients with classical trigeminal neuralgia, the prevalence and severity of neurovascular compression has not been well characterized in patients with hemifacial spasm. We aimed to investigate whether presence and severity of neurovascular compression are correlated to the symptomatic side in patients with hemifacial spasm. All patients in our study were evaluated by a physician who specializes in the management of cranial nerve disorders. Once hemifacial spasm was diagnosed on physical exam, the patient underwent a dedicated cranial nerve protocol magnetic resonance imaging study on a 3 T scanner. Exams were retrospectively reviewed by a neuroradiologist blinded to the symptomatic side. The presence, severity, vessel type, and location of neurovascular compression along the facial nerve was recorded. Neurovascular compression was graded as contact alone (vessel touching the facial nerve) versus deformity (indentation or deviation of the nerve by the culprit vessel). A total of 330 patients with hemifacial spasm were included. The majority (232) were female while the minority (98) were male. The average age was 55.7 years. Neurovascular compression (arterial) was identified on both the symptomatic (97.88%) and asymptomatic sides (38.79%) frequently. Neurovascular compression from an artery along the susceptible/proximal portion of the nerve was much more common on the symptomatic side (96.36%) than on the asymptomatic side (12.73%), odds ratio = 93.00, $P < 0.0001$. When we assessed severity of arterial compression, the more severe form of neurovascular compression, deformity, was noted on the symptomatic side (70.3%) much more frequently than on the asymptomatic side (1.82%) (odds ratio = 114.00 $P < 0.0001$). We conclude that neurovascular compression that results in deformity of the susceptible portion of the facial nerve is highly associated with the symptomatic side in hemifacial spasm.

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Abbreviations: MVD = microvascular decompression; NVC = neurovascular compression; SSFP = steady-state free precession

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Introduction

Hemifacial spasm is a movement disorder characterized by involuntary spasms of the facial muscles. Despite a lack of pain, hemifacial spasm symptoms can be both physically and emotionally debilitating.[1,2] Vascular compression along the centrally myelinated portion of the facial nerve attached to the pons and just beyond is responsible for hemifacial spasm in the majority of patients.[3–6] Anticonvulsant medication is typically ineffective for hemifacial spasm.[7]

The two most widely accepted treatments for hemifacial spasm are botulinum toxin injections[8] and microvascular decompression (MVD) surgery.[9,10] Botulinum toxin injections reduce spasms by blocking signal transmission of acetylcholine at the level of the synaptic cleft. MVD relieves vascular compression of the centrally myelinated portion of the facial nerve, addressing a proposed aetiopathogenesis of hemifacial spasm.

Vascular compression is a known cause of trigeminal neuralgia. Sindou *et al.*,[11] intraoperatively, and later Maarbjerg *et al.*,[12] radiographically, found that the degree of neurovascular compression (NVC) and the point of compression is associated with the symptomatic side of trigeminal neuralgia. The purpose of our study was to determine if this is true for hemifacial spasm. We sought to evaluate the presence, localization, and severity of NVC in hemifacial spasm. We hypothesized that NVC along the susceptible portion of the facial nerve as well as the more severe form of NVC would be observed more frequently on the symptomatic side in patients with hemifacial spasm.

Materials and methods

Study population

We performed a retrospective analysis of hemifacial spasm patients at the University of Pittsburgh Medical Center-Presbyterian University Hospital. Patients were included if they were diagnosed with hemifacial spasm, underwent dedicated cranial nerve MRI (as described later), and were treated with MVD between April 2014 and March 2020. Patients were excluded if they had prior MVD for hemifacial spasm, did not undergo a dedicated cranial nerve protocol MRI, or if there was too much motion artefact on the MRI.

This study was approved by the University of Pittsburgh Institutional Review Board.

MRI technique

Patients underwent dedicated brainstem MRI typically performed 2–3 days prior to MVD to delineate the facial nerve and adjacent vessels. Studies were performed on a 3 T MR scanner (GE Healthcare). Studies were performed using our previously described cranial nerve protocol which includes sagittal T_1, axial FLAIR (fluid-attenuated inversion recovery), and DWI (diffusion weighted imaging) sequences of the whole brain with the addition of dedicated thin-section axial, coronal, and sagittal steady-state free precession (SSFP) images through the brainstem[4] (Table 1).

Anatomical terms

We adapted anatomical terms used by Tomii *et al.*[13] and Campos-Benitez and Kaufmann[3] The facial nerve exits the brainstem at the most concave area of the pons, near the pontomedullary sulcus, termed the 'root exit point'. The attached segment of the facial nerve stretches from the root exit point until the nerve detaches from the pons, termed the 'root detachment point'. The facial nerve between the root exit point and root detachment point is tightly bound to the ventral surface of the pons by both pia and connective tissue for ~8–10 mm, and is termed the 'attached segment'[3,4,13] (Fig. 1). The proximal cisternal segment of the facial nerve extends from the root detachment point to the area of transition from central to peripheral myelin. Several reports have described the proximal cisternal segment occurring between 1 mm and 3 mm lateral to the root detachment point. We define the 'proximal cisternal segment' as the 3 mm segment lateral to the root detachment point.[13] The facial nerve lateral to the proximal cisternal segment then travels anterolaterally to the porus acusticus, through the internal auditory canal, and exits at the level of the internal auditory canal fundus.[4] We define the distal cisternal segment as any portion of the facial nerve lateral to the proximal cisternal segment to the porus acusticus (Fig. 1). The canalicular segment extends from the porus acusticus to the internal auditory canal fundus. The distal cisternal segment of the facial nerve is peripherally myelinated and considered to be resistant to NVC. The susceptible portion (i.e. the centrally-myelinated segment of the nerve) is considered to be any part of the facial nerve from the root exit point, near the pontomedullary sulcus, through the proximal cisternal segment, which extends a few millimetres past the root detachment point of the facial nerve from the pons. These anatomic sites can be approximated with the usage of thin-section SSFP MRI images. NVC is defined as any point where a vessel touches the intracranial facial nerve. We graded the degree of NVC as either contact (Figs 2 and 3) or deformity (Fig. 4). Contact alone is where there was no discernible CSF between the vessel and the facial nerve. If the vessel displaced the facial nerve, this was termed 'deformity'.

Image interpretation

Blinded to the symptomatic side and surgical results, two authors reviewed the first 20 patients' MRIs, and kappa coefficients were calculated to determine the inter-rater reliability. Kappa coefficients were determined for three criteria: presence of any arterial NVC, site of NVC along the facial nerve, and severity of NVC.

In all patients' bilateral facial nerves were interrogated for the presence of any NVC along the susceptible and non-susceptible portions of the facial nerve. It was noted whether the contacting vessel was an artery, a vein, or both. Any arterial NVC includes

Table 1 MVD imaging protocol

Sequence and plane	Flip angle	Field of view, cm	Repetition time/ echo time, ms	Section thickness, mm	Spacing, mm	Matrix
SSFP-axial	65°	18	Default to minimum	1	0.5	384 × 256
SSFP-coronal	65°	18	Default to minimum	1	0.5	384 × 256
SSFP-sagittal	65°	20	Default to minimum	1	0.5	384 × 256

The protocol also includes routine whole brain sagittal T_1-weighted, axial FLAIR and diffusion sequences.

arterial NVC alone or both arterial and venous NVC. The site of NVC was also noted as occurring along the attached segment (extending along the ventral pons from the root exit point to the root detachment point inclusive), proximal cisternal segment or distal segment (distal cisternal and canalicular segments) of the facial nerve. Additionally, the severity of the NVC was categorized as contact versus deformity of the nerve. If there was both contact and deformity, it was categorized as deformity as this is the more severe of the two forms of NVC.

Statistical analysis

Continuous measures were presented as mean with standard deviation (SD) and categorical measures as *n* (%). The kappa statistics were estimated to assess the inter-rater reliability in the initial double-read examinations. The categories of NVC on the symptomatic side and on the asymptomatic side were compared using McNemar's tests and odds ratios for paired data were calculated to quantify the degree of association. The NVC categories are listed in Table 2. All tests were two-tailed with an alpha set of 0.05. Analysis were conducted using SAS v9.4 (SAS Institute, Cary, NC).

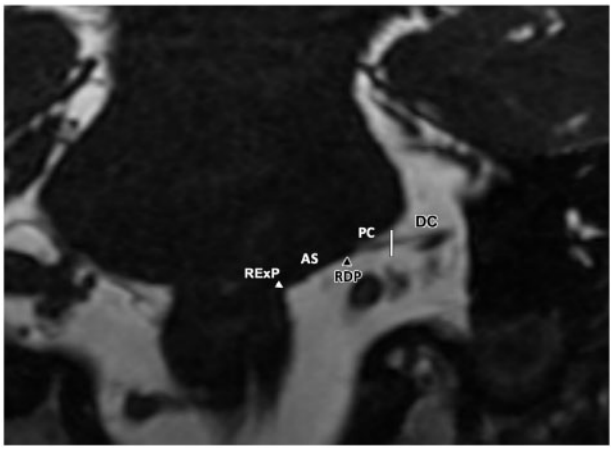

Figure 1 Facial nerve anatomy. Coronal SSFP image shows the expected locations of the root exit point (RExP, white arrowhead), the attached segment (AS) along the ventral surface of the pons, and the root detachment point (RDP, black arrowhead). The proximal cisternal segment (PC) extends ~3 mm from the root detachment point to the lateral margin of the white line. The distal cisternal portion (DC) of the facial nerve extends from the lateral margin of the white line to the porus acusticus, which is not shown.

Data availability

The authors confirm that the data supporting the findings of this study are available within the article and its Supplementary material.

Results

We identified 358 patients who a history of hemifacial spasm and underwent MVD by the same neurosurgeon at our institution. Twenty-four patients were excluded for not having preoperative MRI at our institution and an additional four patients were excluded for excessive MRI artefact or incomplete imaging; therefore, 28 patients were excluded. The remaining 330 patients were evaluated for our study. Of these 330 patients, 232 (70.30%, 232/330) were female and 98 (29.70%, 98/330) were male. At the time of the patient's MVD, the mean (SD) age was 55.7 (±11.34), mode was 65, median age was 56 and the age range was 20–88 years of age. A total of 147 patients (44.55%, 147/330) had right-sided hemifacial spasm, while 183 (55.45%, 183/330) had left-sided hemifacial spasm.

Regarding the initial double-read examinations, kappa coefficients were determined for the three criteria: (i) presence of any arterial contact; (ii) site of contact along the facial nerve; and (iii) severity of NVC. All three criteria had substantial interrater reliability with kappa values ranging from 0.8 to 1. The kappa coefficient for any arterial NVC versus no arterial NVC was 1 [0.95 confidence interval (CI) = 1–1]; a kappa coefficient of 0.78 (0.95 CI = 0.37–1) for site of NVC along the facial nerve; and a kappa coefficient of 0.75 (0.95 CI = 0.42–1) for severity of NVC.

Of the 330 patients, 325 (98.48%, 325/330) were found to have NVC (arterial or venous) of the symptomatic nerve, 323 (97.88%, 323/330) were found to have arterial NVC on the symptomatic side, and 318 (96.36%, 318/330) had arterial NVC along the susceptible segment of the symptomatic facial nerve. Of the patients with arterial NVC along the susceptible portion of their symptomatic nerve, there were 86 patients (26.06%, 86/330) with contact only

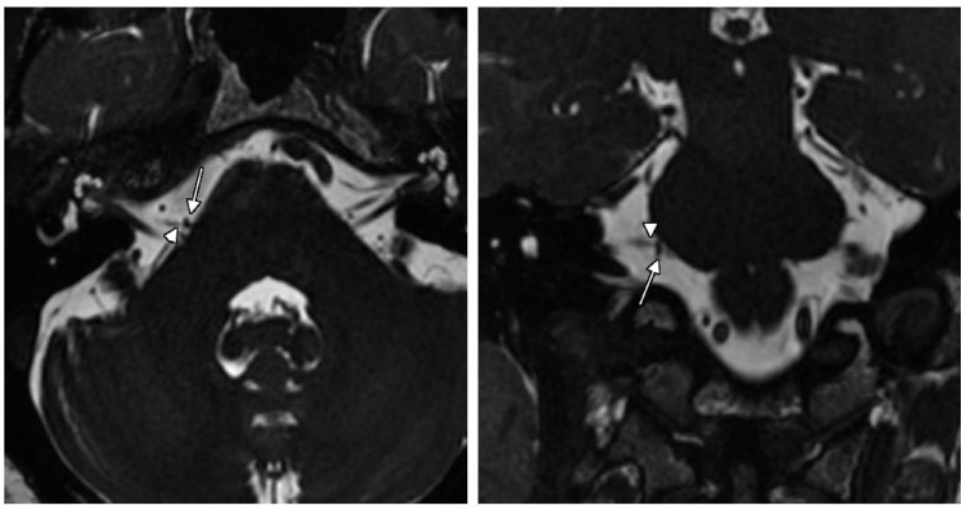

Figure 2 Arterial contact along the proximal cisternal segment of the facial nerve. (**A**) Axial and (**B**) coronal SSFP images demonstrating contact of the proximal cisternal segment (white arrowhead) by the anterior inferior cerebellar artery (white arrow).

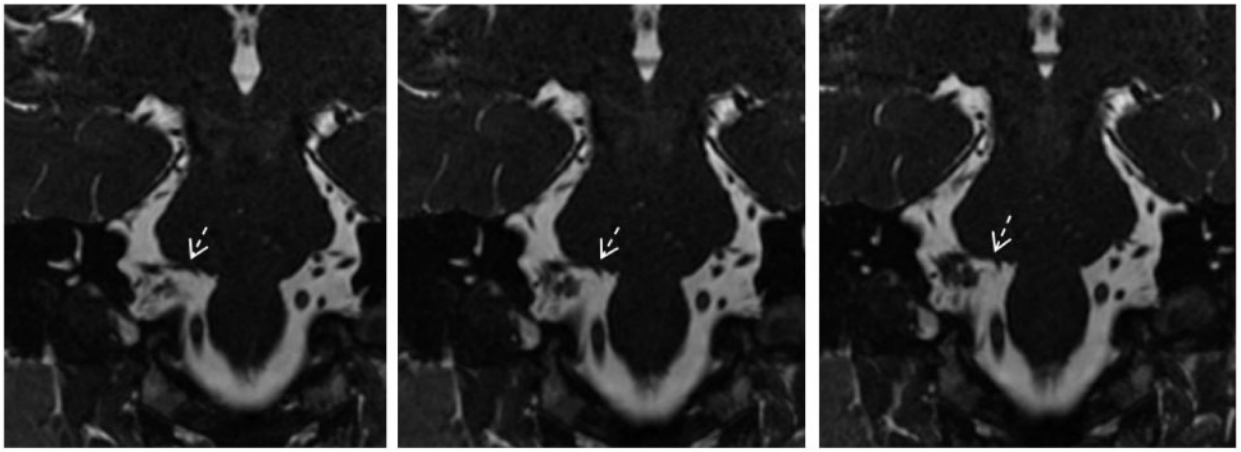

Figure 3 Arterial contact along attached segment of the facial nerve. Subjacent coronal SSFP imaging slices demonstrating the posterior inferior cerebellar artery looping upward and contacting (white dashed arrow) the attached segment of the facial nerve.

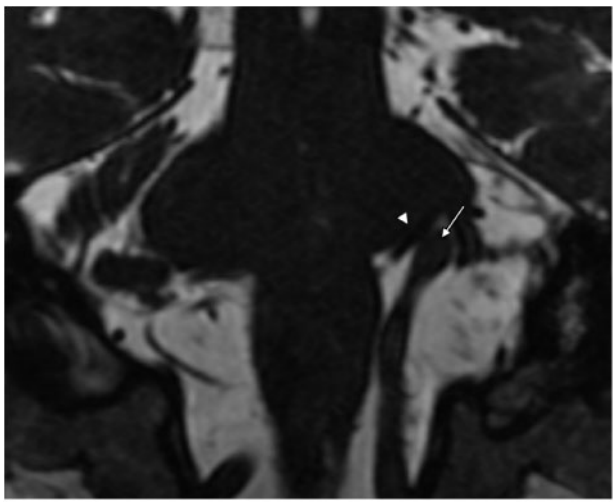

Figure 4 Arterial deformity along the attached segment of the facial nerve. The left anterior inferior cerebellar artery deforms the ventral pons along the attached segment of the facial nerve (white arrowhead). The left vertebral artery lies just inferior to the pons (white arrow).

and 232 patients (70.3%, 232/330) with deformity along the susceptible segment of the nerve. Two patients (0.61%, 2/330) were found to have only venous NVC on the symptomatic side and only one (0.3%, 1/330) had the site of venous contact along the susceptible segment of the nerve. In this patient the vein contacted but did not deform the facial nerve. Additionally, five patients (1.52%, 5/330) were found to have no NVC of their symptomatic facial nerve.

Of the 330 patients, 150 (150/330, 45.45%) were found to have NVC (arterial or venous) of the asymptomatic facial nerve; 128 (38.79%, 128/330) were found to have arterial NVC of the asymptomatic nerve; and 42 (12.73%, 42/330) had arterial NVC along the susceptible segment of the asymptomatic facial nerve. Of the patients with arterial NVC along the susceptible portion of their asymptomatic nerve, there were 36 (10.91%, 36/330) with contact only and six (1.82%, 6/330) with deformity along the susceptible segment of the nerve. Twenty-two patients were found to have only venous NVC of the asymptomatic facial nerve (6.67%, 22/330). Of these, 12 (3.64%, 12/330) had venous NVC along the susceptible segment of the nerve, 10 (3.03%, 10/330) with contact only and two (0.61%, 2/330) with deformity. Additionally, 180 patients (54.45%, 180/330) were found to have no NVC of their asymptomatic facial nerves.

When comparing the symptomatic versus the asymptomatic nerves the odd ratios of having any arterial NVC along the susceptible portion of the nerve was 93.00 (0.95 CI = 29.81–290.11, $P < 0.0001$), of observing arterial deformity along the susceptible portion was 114.00 (0.95 CI = 28.24–458.60, $P < 0.0001$), of observing arterial contact only along the susceptible portion as 2.61 (0.95 CI = 1.73–3.95, $P < 0.0001$), of observing venous NVC only along the susceptible portion of the nerve was 0.08 (0.01,0.64, $P = 0.002$). See Table 2 for a full synopsis of the results.

Discussion

Campbell and Keedy,[14] and later Gardner,[15] were the first to associate vascular compression of the facial nerve with hemifacial spasm. Since then, extensive research has suggested that vascular compression is implicated in the pathophysiology of hemifacial spasm. For this reason, MVD has become an accepted, evidence-based treatment for hemifacial spasm when non-operative therapies do not provide adequate symptomatic relief. MVD has been shown to have a greater than 90% success rate for patients with hemifacial spasm.[16] This study is able to offer further granularity on the variable of 'vascular contact', differentiating between simple arterial contact and deformity of the facial nerve.

Our results demonstrate that although there was a high prevalence of arterial NVC along the bilateral facial nerves, arterial NVC is observed on the symptomatic side in almost all patients (97.88%) and on the asymptomatic side in less than half of patients (38.79%). Arterial NVC (contact or deformity) along the susceptible portion of the nerve is observed on the symptomatic side in most patients (96.36%) and on the asymptomatic side in minority of patients (12.73%). The most severe form of NVC, arterial deformity, was noted along the susceptible portion of the nerve frequently on the symptomatic side (70.03%) and infrequently on the asymptomatic side (1.82%).

There have been variable results of studies looking at NVC along the susceptible portion of the facial nerve in patients with hemifacial spasm. Campos-Benitez and Kaufmann[3] showed that NVC along the susceptible nerve was present in 96% of cases, which is consistent with the findings of our study. NVC causing

Table 2 Summary of results

Type of compression	Symptomatic nerve (%)	Asymptomatic nerve (%)	Odds ratio (CI)	P-value
Any arterial NVC	323 (97.88)	128 (38.79)	98.50 (24.46–396.62)	<0.0001
Along the susceptible portion of facial nerve	318 (96.36)	42 (12.73)	93.00 (29.81–290.11)	<0.0001
Contact alone	86 (26.06)	36 (10.91)	2.61 (1.73–3.95)	<0.0001
Deformity	232 (70.3)	6 (1.82)	114.00 (28.24–458.60)	<0.0001
Venous only NVC	2 (0.61)	22 (6.67)	0.09 (0.02–039)	<0.0001
Along the susceptible portion of facial nerve	1 (0.3)	12 (3.64)	0.08 (0.01–0.64)	0.002
Contact alone	1 (0.3)	10 (3.03)	0.10 (0.01–0.78)	0.007
Deformity	0 (0)	2 (0.61)	0	<0.0001
No NVC	5 (1.51)	180 (54.54)	0.006 (0.001–0.04)	<0.0001

Odds ratio was estimated for symptomatic versus asymptomatic based on paired analysis.

deformity or atrophy along the susceptible portion of the trigeminal nerve has been associated with symptomatic disease in trigeminal neuralgia.[11] The results of the present analysis suggest the same is true for hemifacial spasm.

There are several limitations to this study. This is a single-centre retrospective evaluation at an institution with a dedicated cranial neuralgia program; results may not be generalizable to all institutions. A limitation of our study includes a lack of healthy control subjects. The asymptomatic side in our patients is a reasonable control as previous studies have shown that bilateral hemifacial spasm is extremely rare, on the order of 0.6% in patients with hemifacial spasm, and none of our patients had bilateral hemifacial spasm.[17] Finally, the sample size of this study was determined by the number of patients in the inclusion time period rather than by using a sample-based calculation.

Further research may consider assessing the extent to which the severity of NVC predicts surgical outcome. Hypothetically, a more severe grade of NVC of the facial nerve may be associated with improved surgical outcomes once the NVC is relieved by MVD. This could inform the risk benefit discussions between the surgeons and patients before MVD.

Conclusion

While NVC of the facial nerve is commonly observed in both symptomatic and asymptomatic nerves, arterial NVC along the susceptible portion of the facial nerve and severe arterial NVC of the susceptible portion are seen in the vast majority of the symptomatic and in the small minority of the asymptomatic nerves.

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Competing interests

The authors report no competing interests.

Supplementary material

Supplementary material is available at *Brain* online.

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