Episodic Hemi-laryngopharyngeal Spasm: A Novel Cranial Neuropathy Treatable Surgically

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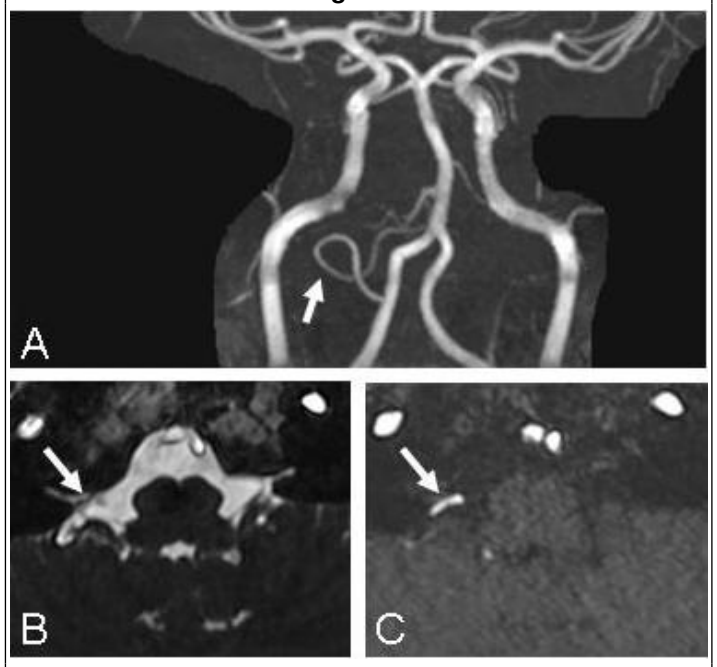
Introduction: We describe a novel cranial neuropathy manifesting with life-threatening episodic hemi-laryngopharyngeal spasm (HELPS).

Methods: We studied a case of a 50-year-old woman presenting with a four-year history of episodic "throat contractions" triggered by speaking, associated with an accompanying distressing feeling of "choking" and a sensation of "tongue swelling". She had no associated pain in her face, ears or throat. The frequency, length and severity of the episodes slowly increased and they began to occur spontaneously even while sleeping. She presented to the emergency department on multiple occasions with severe respiratory distress. Treatments with anti-histamines, epinephrine and bronchodilators were ineffective and typically the episodes stopped spontaneously. On two occasions she required intubation and ICU admission. The otolaryngology service diagnosed her with 'right sided vocal cord dysfunction', and botulinum toxin treatments were initiated.

Results: Botulinum toxin injections into her right pharyngeal muscles and vocal cord reduced the severity of her spasms, but the episodes continued to occur. MRI demonstrated a possible neurovascular conflict involving the cranial nerve IX -X complex and the posterior inferior cerebellar artery (Figure 1). Retrosygmoid approach to cerebello-pontine angle identified the tonsilomedullary segment of PICA which looped between the upper two rootlets of CN X, with tenacious arachnoid adhesions between the artery and the two rootlets (Figure 2). Microvascular decompression (MVD) of the upper rootlets of the vagus nerve was performed by sharply dissecting the arachnoid adhesions and mobilizing the artery to eliminated the distortion of the vagus nerve rootlets (Figure 3). Post-operatively, she had one episode of spasm the day after the surgery and then no further spasms. Her episodic sensation of a "swollen tongue" also resolved. She remains free of the spasmodic episodes at 1.5 year follow up.

Figure 1



A preoperative MRI images demonstrating the neurovascular conflict between the CNIX-X complex and PICA. A) MRI Angiography demonstrating the PICA loop. B) MRI CISS sequence image with the arrows identifying CNIX-X complex and PICA. C) MRI T2 sequence enlarged image with the arrows identifying the neurovascular structures.

Discussion: HELPS consists of i) a sensation of a "swollen tongue" despite its normal appearance, ii) "choking" initiallly triggered by speaking then occurring spontaneously, and iii) laryngopharyngeal spasms prompting emergency consultations. The selective compression of CN X motor rootlets can lead to HELPS (analogues to Hemifacial Spasm). Absence of reporting on HELPS may be due to a combination of its rare occurrence and under-recognition. HELPS should be differentiated from Episodic Laryngospasm reported in otolaryngology literature, which is ascribed to a conversion disorder or occult gastro-esophageal reflux. HELPS can be cured with MVD.

Figure 2

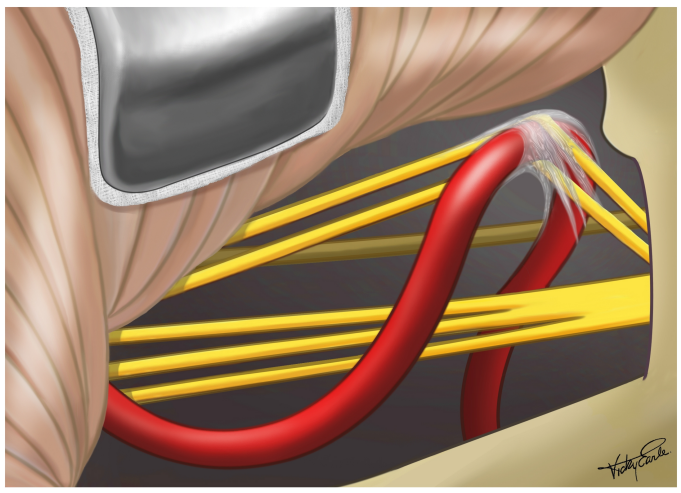


Diagram of the operative view: neurovascular distortion of the upper two rootlets of CN X but no distortion of its lower three rootlets or CN IX.

Figure 3

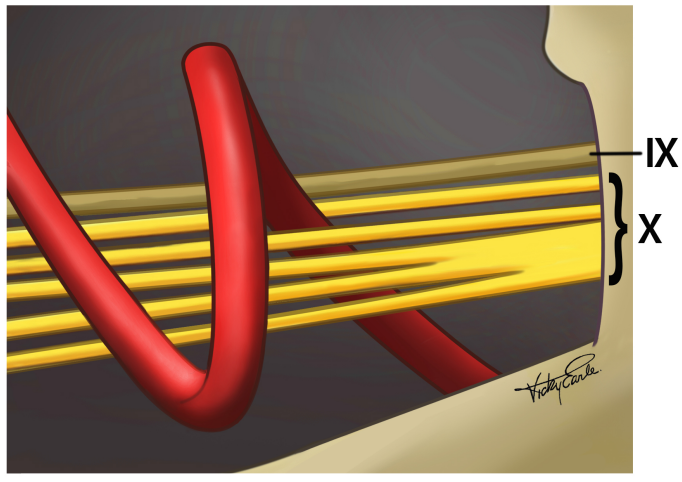


Diagram of the operative view: resolution of the distortion after freeing the arachnoid adhesions.

References:

Will be provided on request.