Case in Point

Syncope from an Unusual Cause: Glossopharyngeal Neuralgia

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For this patient, recurrent episodes of severe ear and throat pain progressed to syncopal episodes that interfered with his ability to drive safely. This case highlights the clues that can point clinicians in the direction of this rare syndrome.

Although common, syncope can be life threatening when it occurs while the individual is performing a dangerous task, such as driving a motor vehicle. For this reason, it's essential that clinicians be able to discern the specific cause of syncope from its many possible etiologies.

In this article, we describe the case of a man who was admitted to the hospital after experiencing recurrent syncopal episodes while driving. A thorough evaluation indicated a diagnosis of glossopharyngeal neuralgia—a rare syndrome that causes brief, recurrent episodes of severe pain in the throat and ear region. After presenting this case, we review the evaluation of syncope and provide an overview of glossopharyngeal neuralgia as a possible cause of syncope.

INITIAL EXAM

A 60-year-old man presented to the emergency department (ED) of a VA medical center with multiple episodes of syncope. His symptoms had started one year prior with episodes of stabbing ear pain (which he described as being 10 out of 10 in severity) and sore throat lasting five to 15 seconds. The episodes occurred as often as every three to four minutes and had progressively worsened over the past year. During these episodes, the pain would start in his right ear and radiate down the right side of his face into his throat. There was no loss of hearing, but throat spasms caused him to feel weak and unable to talk. Talking, turning his head to the right, coughing, yawning, chewing, swallowing (especially cold, carbonated drinks), or bumping his head would cause pain that he described as "worse than eating a chunk of red hot barbeque coal." Symptoms would awaken him from sleep.

After multiple visits to VA clinics (including primary care; ear, nose, and throat; and neurology clinics), the patient had been diagnosed with glossopharyngeal neuralgia. Various pharmacological treatments were tried—including carbamazepine, gabapentin, oxycodone, and other pain medications—none of which provided satisfactory pain relief. (The oxycodone provided minimal relief, but the carbamazepine and gabapentin gave no baseline relief.)

In the month prior to presentation, syncope began occurring during the pain episodes. He would recall the typical auricular and facial pain, black out, and wake up seconds later on the floor. After awakening from one syncopal episode, he measured his blood pressure using a home monitor and recorded a reading of 55/20 mm Hg, with a pulse of 20 beats per minute.

After having six syncopal episodes in one day, the patient decided to drive to the nearest ED to seek medical assistance. During the drive, he felt an episode coming on and had just enough time to pull off to the side of the road before he lost consciousness. A law enforcement officer came to his aid after noticing his erratic driving and escorted him to the nearest hospital. The patient was frustrated when the ED of this hospital discharged him several hours later, telling him he was "OK."

The patient, however, remained concerned for his health and drove to our institution. During this drive, he continued to experience episodes, with loss of consciousness, every 20 to 30 minutes. He managed to pull over at the start of each episode before losing consciousness and then resume driving after he regained consciousness.

During his ED evaluation at our institution, results of his physical examination—which included pulmonary, abdominal, and neurologic examinations—were essentially un-

At the time of this case, **Dr. Dunn** was an assistant professor, **Dr. Oehler** was an associate professor, **Dr. Small** was a resident physician in internal medicine, **Dr. White** was a resident physician, and **Dr. Bruton** was a medical student, all at the University of South Florida, Tampa, FL. Dr. Bruton graduated with his MD in 2009 and is currently a resident physician in Michigan. Additionally, Dr. Dunn and Dr. Oehler are both staff physicians at the James A. Haley Veterans' Hospital, Tampa, FL.

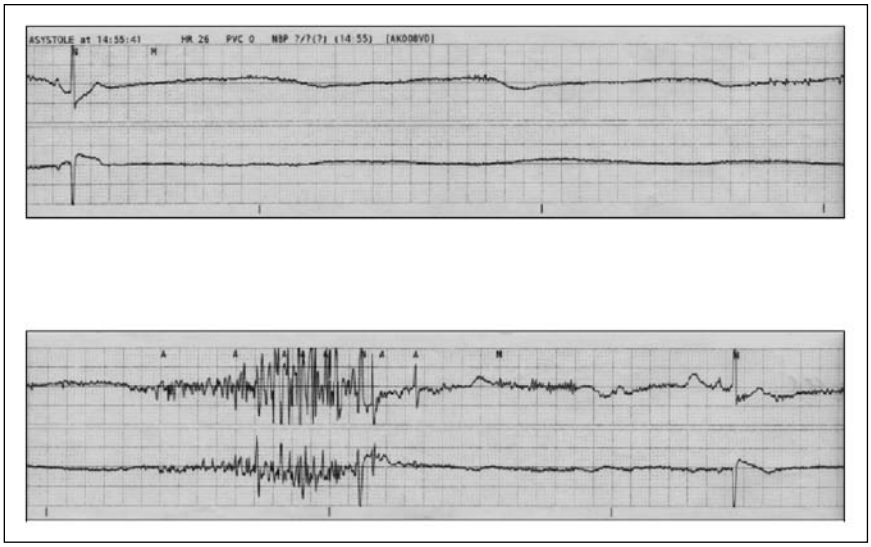

Figure 1. Telemetry rhythm strip showing the patient's 15-second episode of asystole.

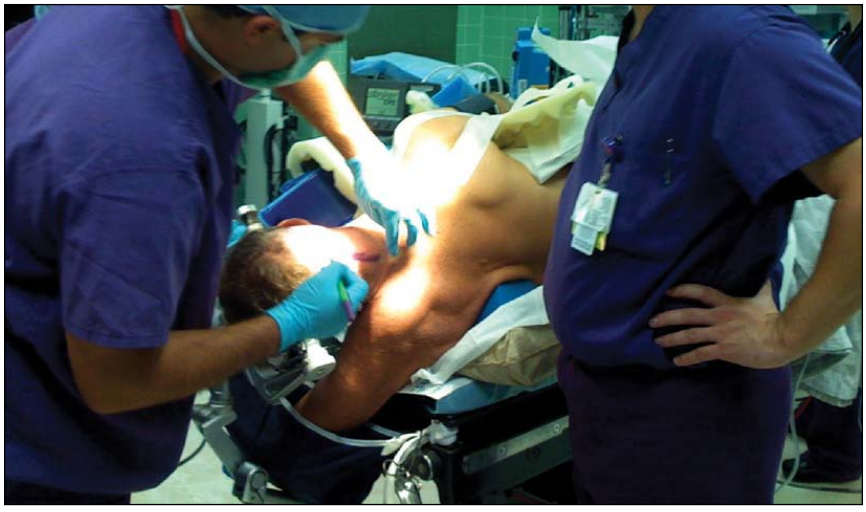

Figure 2. The patient being prepared for microvascular decompression. Image courtesy of Norberto Andaluz, MD and Clinton Burkett, MD.

remarkable except for noting that the patient had upper dentures and a left sided facial palsy from an old injury. Subsequent electrocardiography (ECG) showed normal sinus rhythm. Results of a complete blood count and a comprehensive metabolic profile were unremarkable.

Despite the benign findings of initial testing, the history of the patient's condition prompted his transfer to a cardiac telemetry unit for further monitoring. While on the unit, the patient had multiple episodes of shaking and discomfort due to facial pain that caused him to hold the right side of his head in his hands. During one of these episodes, the telemetry computer detected an episode of asystole (Figure 1), which lasted for 15 seconds. Based on this finding, as well as his history of ear and throat pain, we determined the cause of the patient's pain and syncope to be consistent with his previously diagnosed glossopharyngeal neuralgia.

TREATMENT COURSE

Due to the patient's episodes of asystole, a cardiology consultation was requested for consideration of pacemaker placement. Four days after admission, the cardiologist placed a dual chamber pacemaker, which would function if the patient's intrinsic heart rate fell below 45 beats per minute.

While in the hospital, the patient underwent a nerve block to relieve his pain, but this treatment provided relief for only six hours. The inadequacy of this treatment, along with the previous failure of multiple pharmacologic therapies, suggested that surgery might be necessary to address the glossopharyngeal neuralgia.

The medical team initiated imaging studies to evaluate for any underlying cause of the glossopharyngeal neuralgia that might indicate the need to consult a particular surgical specialty. Magnetic resonance imaging (MRI) of the brain with contrast was conducted to exclude any masses and showed no significant intracranial findings. MRI and magnetic resonance angiography of the neck were conducted to evaluate the vasculature, with unremarkable results. A skull x-ray showed a somewhat prominent styloid process on the right, but a follow-up computerized tomography (CT) scan of the temporal bones showed these styloid processes to be at the upper limits of normal.

Once these studies established no secondary cause of glossopharyngeal neuralgia, the neurosurgery team was consulted. They offered the patient microvascular decompression of cranial nerves IX and X, a procedure

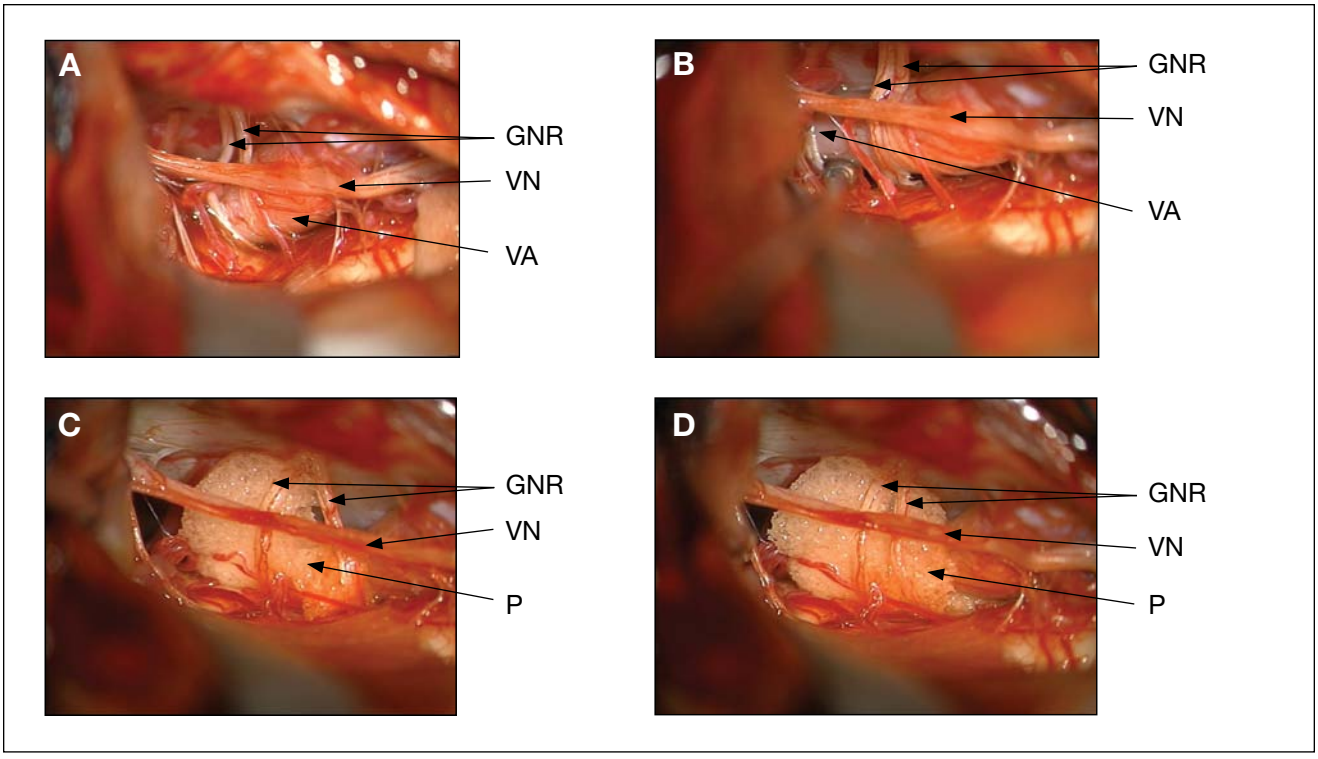

Figure 3. Images from the patient's microvascular decompression procedure. Image A shows the glossopharyngeal nerve rootlets (GNR) being stretched by the vertebral artery (VA). The vagus nerve (VN) is also pictured. In image B, the surgeon dissects the tissue and pushes the VA away from the GNR. In image C, the pledget (P) acts as a cushion between the GNR and the VA (not visible). Image D shows complete decompression of both the GNR and the VN from the VA (not visible). Images courtesy of Norberto Andaluz, MD and Clinton Burkett, MD.

known to relieve pain from glossopharyngeal neuralgia.[1–3] During his surgery (Figure 2), performed two weeks after hospital discharge, the glossopharyngeal nerve appeared to be in intimate contact with the right vertebral artery and was repositioned to relieve the nerve compression (Figure 3).

Despite a technically successful operation, the patient experienced several complications. These included: postoperative meningitis, which required antibiotic therapy; hand myoclonus, which required antiepileptic medication; hydrocephalus, which required a ventriculoperitoneal shunt; and mild ataxia, which required a long rehabilitation.

Despite these setbacks, the patient's condition improved over the course of six months. The patient stated that, before the procedure, he had experienced pain every two to three minutes, that the pain was the worst he had ever had, and that he would have killed himself if the pain had continued. After his procedure, he said that the pain was "100 times better." He reported being glad to have had the operation, despite the complications and long recovery time. He has had no further syncope to date—and as a result, the roads are a little bit safer.

ABOUT THE CONDITION

Getting to the bottom of syncope

Causes of syncope are numerous, including cardiac, orthostatic, and neurologic etiologies.[4] Syncope also may be drug-induced. In many cases, the basic evaluation of patients presenting with syncope will provide clues to the underlying cause.

The absence of a prodrome[5] or the presence of such associated symptoms as chest pain or palpitations point to a cardiac origin of syncope.[6] Other clues to cardiac etiologies include cardiac abnormalities on physical examination and abnormal ECG findings. Orthostatic causes are easily excluded by measuring orthostatic pulse and blood pressure at time of presentation.[6,7]

Neurologic disorders are suspected in patients who report seeing an "aura" preceding or experience confusion following the syncopal episode; those who report experiencing

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visual changes, speech impediments, limb weakness, sensory changes, or gait abnormalities during an episode[5]; and those who have neurologic abnormalities on physical examination.[5] A low oxygen saturation may point to a pulmonary cause of syncope. In the patient presented here, the associated symptoms of facial and auricular pain were a clue to the neurologic origin of his syncope.

Although laboratory tests are not routinely recommended to diagnose the cause of syncope,[4] such testing may be useful to screen for low hematocrit levels,[8] if the history warrants it,[4] or if the clinician is unsure whether the patient had a loss of consciousness from a disorder other than cerebral hypoperfusion.[6]

Glossopharyngeal neuralgia

The glossopharyngeal nerve, which refers to either of the ninth pair of cranial nerves, supplies motor and sensory fibers to the pharynx, posterior tongue, and parotid gland. Glossopharyngeal neuralgia is a rare pain syndrome that is characterized by recurrent episodes of severe pain in the back of the throat, tongue, tonsils, and middle ear. These episodes may last anywhere from a few seconds to a few minutes.[9]

In addition to somatic sensory inputs, the glossopharyngeal nerve also receives chemoreceptor and baroreceptor afferent inputs from the carotid body and the carotid sinuses, respectively.[9] The afferent fibers for chemoreceptors and baroreceptors pass on to the dorsal nucleus of the vagus nerve, and from there, to respiratory and vasomotor centers.[9] When the glossopharyngeal nerve induces a vasovagal reflex, this mechanism can result in bradycardia, hypotension, and even asystole and subsequent syncope.[10,11] In this way, glossopharyngeal neuralgia can be life threatening.[1,9]

Most cases of idiopathic glossopharyngeal neuralgia are caused by vascular compression of the glossopharyngeal nerve by nearby blood vessels, but any compression to the glossopharyngeal nerve can cause neuralgia.[9] A common secondary cause is Eagle's syndrome, which involves compression of the nerve by an elongated styloid process or a calcified stylohyoid ligament.[12] Other causes of secondary glossopharyngeal neuralgia include cerebellopontine angle tumors, parapharyngeal space lesions, multiple sclerosis, temporal bone metastasis, localized infections, pharyngeal carcinomas, posterior fossa arteriovenous malformation, or metastatic head and neck tumors.[9,13,14]

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Although pacemakers can control the episodes of asystole and syncope, they cannot alleviate the intense facial, throat, and ear pain that accompany glossopharyngeal neuralgia.[11,13] The debilitating pain can cause depression and has led to several cases of suicide.[11] Anticonvulsants, such as carbamazepine and gabapentin, are widely used to treat glossopharyngeal neuralgia.[11] Other medications—including lamotrigine,[15] pregabalin,[16,17] amitryptiline, baclofen, phenobarbital, and diazepam[11]—also have been prescribed and reported to help in isolated cases.

Surgical options for glossopharyngeal neuralgia range from nerve sectioning to microvascular decompression (the procedure undergone by the patient presented here). During microvascular decompression, the blood vessel is repositioned to relieve the nerve compression. There is significant risk of morbidity with microvascular decompression. Risks include cerebellar or brainstem infarction, IX and X cranial nerve palsies, postoperative paresis, intracranial hematoma, cerebrospinal fluid leak, facial palsy, dysphagia, hoarseness, or operative death. Despite the risks, microvascular decompression is an effective surgical procedure for producing prolonged pain relief in patients with medically intractable glossopharyngeal neuralgia.[1]

IN SUMMARY

Glossopharyngeal neuralgia should be considered as a cause of syncope in patients who have episodes of paroxysmal, lancinating pain in the regions supplied by this nerve, such as the tongue, ear, or throat. The disorder has a number of causes, including intrinsic nerve irritability and external compression. Imaging studies should be performed to exclude secondary causes. Clinicians have a wide range of options when treating this disorder, including medications, pacemaker placement, and surgery.

Author disclosures

The authors report no actual or potential conflicts of interest with regard to this article.

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