Case in Point

Glossopharyngeal Neuralgia

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Diagnosis of this rare but treatable syndrome is often delayed due to unfamiliarity with the condition.

Glossopharyngeal neuralgia (GPN) is a rare condition, with a frequency of about 1.5 to 3 cases per one million people.[1,2] Incidence of the condition peaks between the ages of 40 and 60 years,[1,2] though pediatric cases have been reported.[3] It is characterized by bursts of severe, typically unilateral, sharp or lancinating pain that is stimulated by swallowing, talking, or coughing. Most likely, it is caused by vascular compression of the glossopharyngeal nerve—the small, third branchial arch nerve lying deep in the neck.[4] Although uncommon, it is a distinctive syndrome that is important to consider when diagnosing throat or ear pain. Here, I present the case of a patient referred to the neurology clinic for constant throat pain and review the differential diagnosis and management options.

INITIAL EXAM

A 47-year-old man with a two-year history of throat pain—which occurred daily, when eating meals and snacks, and never at night—was referred to the neurology department at William Beaumont Army Medical Center, El Paso, TX. He reported that the pain had begun at a low intensity but, as the intensity had increased over the course of several months, his primary care physician had referred him for gastroenterologic evaluation. He subsequently underwent endoscopy and an upper gastrointestinal series, but neither revealed abnormal results. With continuing pain, he was referred to otolaryngology. Nasal fiberoptic evaluation also was normal. A speech pathologist was consulted to evaluate his swallowing and oromotor function; the findings were normal.

On presentation to neurology, the patient reported pain upon drinking fluids or swallowing any consistency of solid or semisolid food. The patient described a nonthrobbing, very sharp pain lasting up to three seconds with each swallow and an additional one second after swallowing. It was not burning, electrical, or sharp as in a needle stick, yet was so intense, he "braced himself" before each swallow. The pain was localized to the middle region of the velum (bilateral rather than unilateral) and did not radiate to the ear, ear canal, jaw, angle of the jaw, throat, or larynx. The pain occasionally occurred with no oral-motor activity and was described as a throbbing, dull pain. He did not experience pain with the normal swallowing of saliva and he did not have dysphagia.

The patient did not have a history of headaches; migraines; nausea; vomiting; facial, eye, or ear pain; infections; tinnitus; or visual changes. He had no history of associated bradycardia or syncope with his pain attacks. He reported that laughing, chewing, speaking, coughing, or brushing his teeth did not trigger pain and he was able to turn his head and extend his tongue without discomfort. A review of systems and his family history were noncontributory.

A detailed neurologic examination of all 12 cranial nerves, along with a general physical examination, were normal. Routine laboratory investigations, including serum protein electrophoresis, rapid plasma reagent, and sedimentation rate were normal. Screening for HIV antibody, antinuclear antibody, angiotensin converting enzyme, and extractable nuclear antibodies were negative. Vitamin B_{12} and folate levels were normal. Radiologic assessments included an evaluation for soft tissue cervical spine injury and a modified barium swallow accompanied by fluoroscopic examination, both of which yielded unremarkable results. Computed tomography was normal, with no pathologic calcification of the styloid hyoid process or tendon. Both magnetic resonance (MR) imaging of the brain and MR angiography were normal.

TREATMENT COURSE

Based on the patient's symptoms and history and an extensive review of the differential diagnosis of throat pain, the etiology of his pain syndrome was

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determined to be glossopharyngeal neuralgia (GPN). The patient initially was treated with a total daily dosage of 3,600 mg of gabapentin, due to the drug's favorable adverse effect profile, but the drug was completely ineffective for this patient's pain relief or prevention. Carbamazepine therapy was then initiated at 100 mg and advanced to 600 mg per day over the course of eight weeks. This resulted in an almost complete relief of pain symptoms.

ABOUT THE CONDITION

The clinical history this patient presented—severe paroxysmal episodes of lancinating pain triggered by swallowing food, pills, or drinking fluids—is very typical of GPN. For example, in GPN, the pain usually is localized to the external ear canal, the base of the tongue, the tonsils, or the region inferior to the angle of the jaw. GPN has been associated with syncopal episodes secondary to reflex autonomic effects on blood pressure and cardiac rhythm. The cardiovascular episodes potentially are life threatening due to cardiac dysrhythmias.[5] One study reports GPN in association with Arnold Chiari type I malformation in an eight-year-old child.[6] Various mass lesions may be associated with GPN but, otherwise, idiopathic GPN is most likely secondary to arterial compression of the glossopharyngeal nerve as it leaves the brain stem.

Conditions that have been reported in association with the clinical presentation of GPN include cancer of the (1) posterior aspect tongue, (2) larynx, and (3) cerebello-pontine angle (CPA). Gupta and colleagues reported the case of a 22-year-old man presenting with CPA schwannoma and symptoms suggestive of neurofibromatosis type 2, but without a family history or cutaneous stigmata.[7] GPN also has been reported in association with the penetration of foreign bodies into the neck.[8]

The differential diagnosis of GPN includes Eagle syndrome, trigeminal neuralgia, and sphenopalatine neuralgia. Knowing and recognizing the clinical features of these similar neuralgias can help clinicians delineate the location of a patient's pain and can guide therapeutic options.

Eagle syndrome affects the glossopharyngeal nerve, similar to GPN, but is not idiopathic—an elongated styloid process irritates the nerve. Typically, the syndrome is seen in older patients who report a chronic throat pain or sensitivity after tonsillectomy. It also has been associated with a granular cell tumor.[9] Eagle syndrome is triggered by head turning, swallowing, or moving or extending the tongue.[10] Treatment is usually surgical.

Trigeminal neuralgia (TN) usually presents with paroxysmal facial pain and headaches in patients aged 40 to 60. It affects the mandibular branch of the trigeminal nerve, but in some cases may affect other divisions of the nerve. The pain is usually unilateral, triggered by stimulation of the affected area of the face, and noted primarily during the day. TN may be the presenting sign of multiple sclerosis. Treatment includes oral medication, injection therapy, and surgery.

Sphenopalatine neuralgia is characterized by persistent, nonparoxysmal pain of the eye, cheek, ear, or neck. It usually is not triggered by head turning, swallowing, or moving the tongue.

Treatment for GPN includes both medical and surgical options. The primary medical treatment involves anticonvulsant medication therapy with carbamazepine. This is typically quite effective for patients with TN, but medication trials have resulted in a lower rate of effectiveness for patients with GPN. Additional medication treatment options include phenytoin, gabapentin, divalproex sodium, lamotrigine, oxcarbazepine, amitriptyline, nortriptyline, and baclofen. If medication trials are unsuccessful, or if a patient is presenting with severe symptoms and manifests clinical symptoms of autonomic instability, he or she should be considered for surgical treatment.[11]

The primary surgical therapy is microvascular decompression of the glossopharyngeal and vagus nerves. Of the 40 patients who were treated for GPN with microvascular decompression from 1970 to 1995, the surgical procedure offered complete (or greater than 95%) long-term pain relief without any medication in 76% and substantial improvement in an additional 16% of patients.[12] Due to the potential for cardiac and hemodynamic instability, great care with the anesthetic and surgical technique must be observed. Vessels involved have included loops of the vertebral and the posterior inferior cerebellar arteries.[13–15]

Familiarity with GPN allows the practitioner to make a rapid diagnosis, complete a thorough radiologic and laboratory evaluation, and provide effective relief of the patient's symptoms. Immediate recognition of the disorder will make a great difference for patients experiencing intense pain.

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adverse effects—before administering pharmacologic therapy to patients.

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