**A Painful Syncope: Glossopharyngeal Neuralgia**

Christina Binder MD, PhD - Department of Radiation Oncology
Briana Ketterer MD - Department of Internal Medicine
Oregon Health & Science University, Portland, OR

---

**Introduction/Background**

Glossopharyngeal neuralgia (GPN) is a rare disorder of the ninth cranial nerve (CN IX), in which paroxysms of severe pain are associated with excessive vagal outflow. It represents 0.2-1.3% of facial pain syndromes. About 2% of GPN cases are associated with severe hypotension and bradycardia during pain episodes, leading to debilitating bradycardia, hypotension, syncope, and cardiac arrest. We describe a case of GPN secondary to a head and neck cancer that resolved with concurrent chemotherapy and radiation.

**GPN Diagnosis:**

- clinical diagnosis of exclusion based on number of episodes, triggers, and character of pain

**Table 1. International Classification of Headache Disorder 3rd ed**

<table>
<thead>
<tr>
<th>Criterion</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>At least three attacks of unilateral pain fulfilling criteria B and C</td>
</tr>
<tr>
<td>B</td>
<td>Pain is located in the posterior part of the tongue, tonsillar fossa, pharynx, beneath the angle of the lower jaw and/or in the ear</td>
</tr>
<tr>
<td>C</td>
<td>Pain has at least three of the following four characteristics:</td>
</tr>
<tr>
<td>1</td>
<td>Recurring or paroxysmal attacks lasting from a few seconds to two minutes</td>
</tr>
<tr>
<td>2</td>
<td>Severe intensity</td>
</tr>
<tr>
<td>3</td>
<td>Shooting, stabbing, or sharp in quality</td>
</tr>
<tr>
<td>4</td>
<td>Precipitated by swallowing, coughing, talking, or yawning</td>
</tr>
<tr>
<td>D</td>
<td>No clinically evident neurologic deficit</td>
</tr>
<tr>
<td>E</td>
<td>Not better accounted for by another ICHD-3 diagnosis</td>
</tr>
</tbody>
</table>

**GPN Etiology:**

- idiopathic, vascular, infectious, traumatic, anatomic, or malignant
- pain is due to neurovascular conflict between CN IX and X
- syncope is due to excessive vagal outflow when irritation of CN IX creates a vasoglossopharyngeal reflex arc, stimulating mechanoreceptors which inappropriately activate baroreceptors

**GPN Treatment:**

- First-line treatment is pharmacologic
  - Carbamazepine is most common
  - Failure of medications escalates to a local therapy
  - surgery, SRS, RFA
  - For malignant GPN, treat the underlying tumor

**Case Presentation**

A 71 y/o male presented with headaches and symptomatic bradycardia 3 months after diagnosis of TxN2bM0 p16+ squamous cell carcinoma.

**Symptoms:**

- constant dull left-sided headaches
- paroxysms of sharp, stabbing, and shooting pain lasting seconds
- hiccups, anxiety, impending sense of doom
- bradycardia to the 30s and hypotension to the 50s/30s

**Work-up**

**CTA Neck:**

- tumor invasion of the parotid gland and parapharyngeal space
- compression of the internal carotid artery to the carotid sinus
- compression of the internal jugular vein
- encasement of CN IX

**Figure 2. CTA Neck and Soft Tissues**

A) Axial, sagittal, and coronal views of pre-treatment CTA neck and soft tissue shows tumor (*) invasion of the parapharyngeal space. B) Axial slices show tumor (red) encasement of the internal carotid artery (green), external carotid artery (orange), internal jugular vein (blue), and path of CN IX (magenta).

**Diagnosis:**

- cardiac work-up was negative
- neurologic work-up was negative
- GPN diagnosis made based on ICHD-3 criterion

**Therapy**

- Intravenous atropine pushes and a dopamine infusion to stabilize autonomic symptoms
- Temporary pacemaker
- Pain regimen
- Chemoradiation

- Weekly Cisplatin 40mg/m²
- Radiation (stopped 3 fractions early due to side effects)
- Tumor: 65Gy in 32 fractions (11Gy in 5 and 54Gy in 27)
- High-risk nodal areas: 57.36Gy in 32 fractions
- Low-risk nodal areas: 51.2Gy in 32 fractions

**Figure 3. Patient-reported pain scores**

A) 3-day average pain scores compared with cumulative radiation dose show a decrease in pain after 176Gy. B) 3-day average pain scores compared with tumor volume as contoured on cone beam CT scans show a decrease and steady reduction in tumor volume.

**Figure 4. PET-CT**

A) Pre-treatment PET-CT axial, sagittal, and coronal images show FDG-avid tumor (*). B) Post-treatment PET-CT images show decreased tumor volume and no FDG uptake.

**Conclusions**

- GPN is rare; malignant GPN syncope is even rarer
- GPN syncope can lead to life threatening complications
- Can bridge to definitive therapy with medical management
- Treatment with chemoradiation can lead to quick, durable relief of pain and syncope as well as sustained tumor control