

A Painful Syncope: Glossopharyngeal Neuralgia

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



Introduction

Glossopharyngeal neuralgia (GPN) is a rare disorder of the ninth cranial nerve in which paroxysms of severe pain are associated with excessive vagal outflow. This can result in bradycardia, hypotension, syncope and even cardiac arrest. This is likely mediated by the branch off the glossopharyngeal nerve that supplies the carotid body and carotid sinus which conveys chemoreceptor and baroreceptor information. This mechanism is responsible for the arrhythmogenicity and vasoplegia. Causes include neoplasm, infection, vascular malformations, Eagle's syndrome and prior surgical interventions. We present a case of GPN which resolved with treatment of a head and neck cancer.

Case Presentation

A 71-year-old-male presented with left sided headaches and symptomatic bradycardia three months following diagnosis of squamous cell carcinoma (SCC) of unknown primary with bulky left cervical adenopathy. He described a constant dull left sided headache with paroxysms of sharp, stabbing, and shooting pain lasting seconds at a time. The paroxysms were associated with hiccups, anxiety, an impending sense of doom, bradycardia to the 40s, and hypotension to 50s/30s. To stabilize his autonomic symptoms, he required intravenous atropine pushes and a dopamine infusion. A temporary pacemaker was placed. Imaging revealed progression of his left cervical tumor. It measured 3.4cm x4.4cm x 5.1cm with infiltration into the parotid gland and parapharyngeal space. This caused compression of the carotid artery near the carotid sinus branch of the glossopharyngeal nerve. He was also found to have cerebral vein thrombosis.



EKG: HR 35, PR 126, QRS 100, QTc 396. Marked bradycardia.

Glossopharyngeal Neuralgia

Glossopharyngeal neuralgia (GPN) was first described in 1910 by Weisenburg and the term "glossopharyngeal neuralgia" was coined in 1921 by Harris. The first case of cardiac arrest and syncope associated with GPN was published in 1942 by Wortis *et.al*. This is a rare craniofacial pain syndrome. Katusic published a 39-year retrospective study (1945-1984) calculated an incidence of 0.7/100,000 population/year. And syncope is even less common. In 1981 Rushton *et.al* reported 217 patients admitted to the Mayo Clinic with GPN. Only two patients experienced syncopal events.

Syncope is a result of extreme bradycardia and even asystole preceded by intermittent lancinating pain in the oropharynx, retropharyngeal space and occipital-temporal region with occasional radiation to the ear. The mechanism is not fully understood but the close connection of the vagus and glossopharyngeal nerve is presumed to create a vasoglossopharyngeal reflex arc whereby pain triggers arrhythmogenicity and vasoplegia. Thus, pain can activate the reflex and result in syncope.

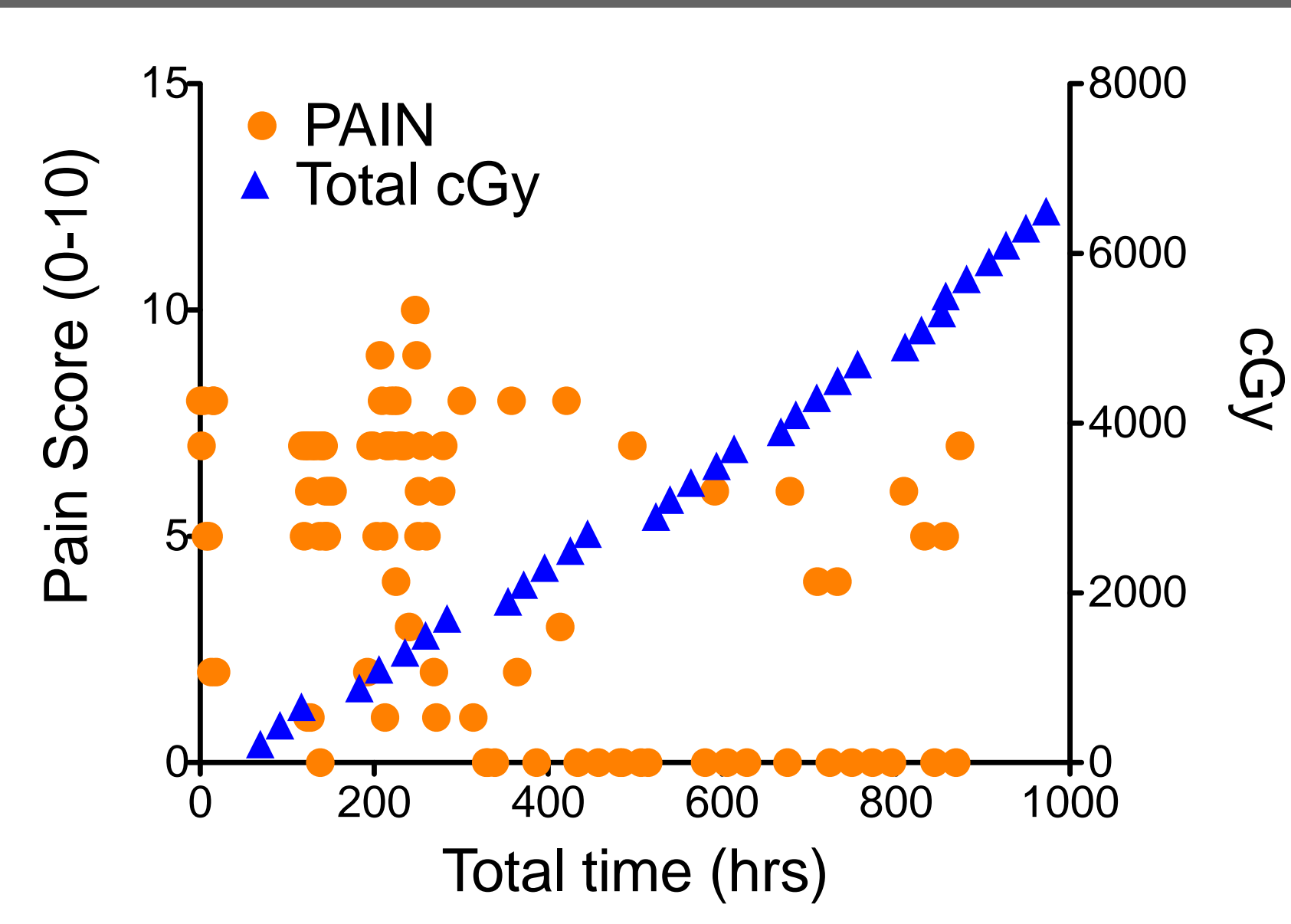


Figure 1 Decrease in reported pain scores over time with increasing radiation represented in cumulative centiGray (cGy).

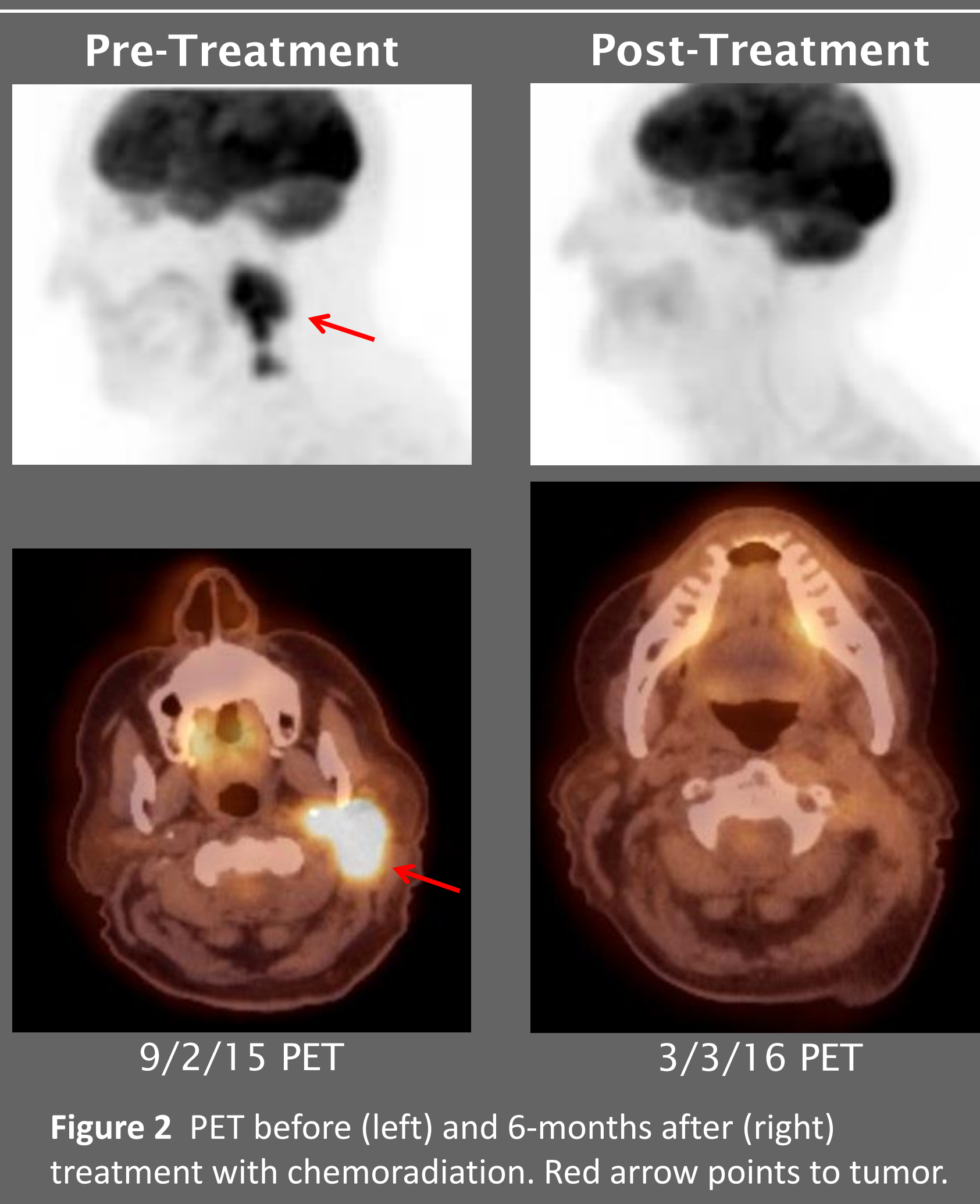
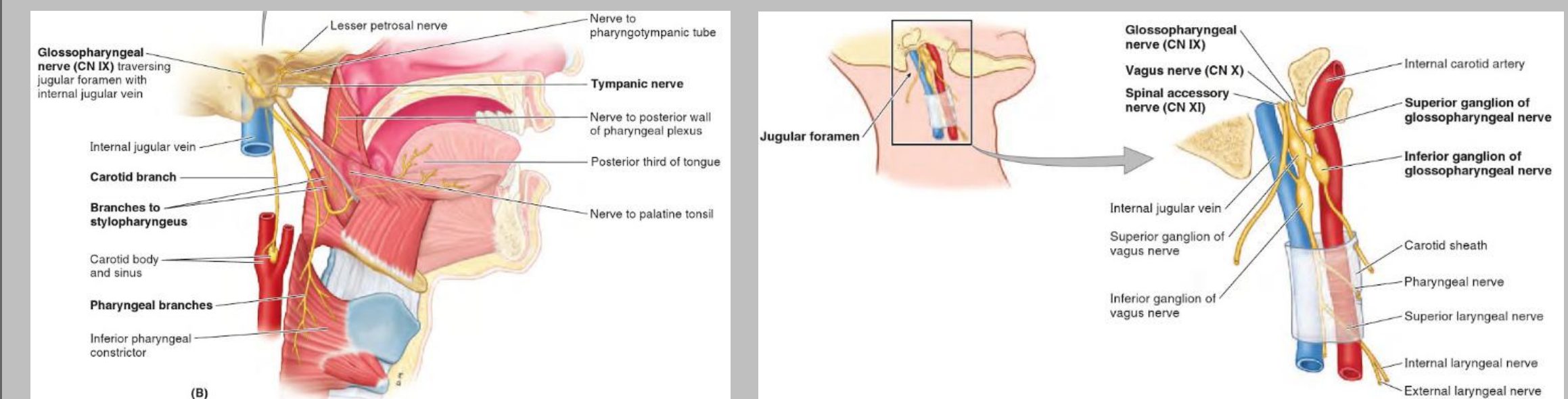


Figure 2 PET before (left) and 6-months after (right) treatment with chemoradiation. Red arrow points to tumor.

Glossopharyngeal Nerve

The glossopharyngeal nerve is the ninth cranial nerve (CN IX). It emerges from the medulla and traverses the cranium through the jugular foramen with the vagus nerve (CN X) and the spinal accessory nerve (CN XI). It has several components and functions:

- Somatic Motor: motor to stylopharyngeus for swallowing
- Visceral Motor: parasympathetic innervation to the parotid gland
- Special Sensory: visceral sensation from the parotid gland, carotid body and sinus, pharynx and middle ear
 - Carotid body and sinus (Nerve of Hering): chemoreceptor and baroreceptor
- Somatic Sensory: taste to the posterior third of the tongue and cutaneous sensation from external ear



Glossopharyngeal nerve (CN IX) anatomy. Adopted from Clinically Oriented Anatomy for Review.

Therapy and Resolution

There is no standard treatment for GPN due to the variety of causes. Case reports describe improvement with medical therapy alone with antiepileptics such as carbamazepine, gabapentin and amitriptyline. Other reports show improvement with microvascular decompression surgically or with stereotactic radiosurgery. Given our patient's bulky, invasive, Stage IVa (TxN2bM0) p16+ SCC, he was treated with chemotherapy and radiation in conjunction with neuromodulating medications. He completed thirty-two radiation treatments to a cumulative dose of 65Gy concurrently with cisplatin. This decreased the size of the mass as seen in Figure 2. His pain and hemodynamic symptoms improved with therapy. He self reported lower pain scores with increasing cumulative Gray as seen in Figure 1. With improvement in symptoms of pain and syncope, the temporary pacemaker was removed, and he was transitioned to maintenance therapy with gabapentin. In this instance, he achieved sustained resolution of GPN and its hemodynamic consequences with chemotherapy and radiation to his left cervical mass.

Conclusion

This case displays how a large squamous cell carcinoma resulted in a painful syncopal phenomenon called glossopharyngeal neuralgia. It also reveals how chemotherapy and radiation produced symptomatic relief. While this is a rare entity, it is worthwhile for both general practitioners and subspecialists to draw a connection between facial pain syndromes and syncope as it may prevent life threatening complications.

References Available on Request