CASE REPORT

Glossopharyngeal and Limited Vagal Neurectomy for Cancer-Related Carotid Sinus Syncope

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ABSTRACT

Head and neck cancer patients with cervical disease involving the glossopharyngeal or vagus nerves can experience dangerous cardiovascular phenomena, including carotid sinus syncope (CSS). Medical and minimally invasive interventions, including pacemaker placement, incompletely address the etiologies of syncopal episodes and therefore often provide limited benefit. The objectives of this report are to highlight the difficulties of managing cancer-related CSS, to present a highly effective surgical intervention, and to review the literature regarding the pathophysiology and treatment options for patients with cancer-related CSS. Two patients with cancer-related CSS continued to experience syncopal episodes despite medical and pacemaker therapy. Consequently, these patients underwent a suboccipital craniotomy for glossopharyngeal and limited vagal neurectomy. After surgery, both patients had uncomplicated postoperative courses with resolution of syncope.

KEYWORDS: Head and neck cancer, carotid sinus, syncope, glossopharyngeal neuralgia, neurectomy

Cardiac sequelae of inappropriate activation of vasomotor centers in the carotid sinus from cranial nerve dysfunction have the potential for morbid consequences. The associated array of signs and symptoms, such as bradycardia, hypotension,

and syncope, have collectively been described as “carotid sinus syncope” (CSS),¹ “glossopharyngeal neuralgia-asystole syndrome,”² and “glossopharyngeal neuralgia with syncope,”^{3,4} among others. Although most cases of glossopharyngeal neuralgia

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with or without syncope are idiopathic and are believed to be the result of intracranial vascular compression of the glossopharyngeal nerve,^{5,6} there are several pathological processes that can result in secondary CSS, including tumor involvement of cranial nerves. Although syncope resulting from cancerous spread to the glossopharyngeal or vagus nerves is an infrequent occurrence, its estimated incidence in as many as 1 in 250 patients with a head and neck malignancy⁷ suggests that clinicians will likely, at some point, encounter patients with this and other related symptoms; they should therefore be familiar with the available treatment options.

Such syncopal episodes are postulated to stem from the activation of a glossopharyngeal-vagal reflex arc, which is most frequently the result of the intense neuralgic pain that characterizes glossopharyngeal neuralgia.⁴ Accordingly, primary efforts to counter this syndrome have focused on suppressing pain episodes with medical anticonvulsant therapy. However, cancer-related CSS patients often present without any glossopharyngeal neuralgic pain.¹ Other, more invasive techniques to address CSS that have been described include local injection of anesthetic blocks and neurolytic substances, percutaneous rhizotomy, and implantation of pacemakers.⁵ When these options fail to provide adequate relief from dangerous cardiac symptomatology, a surgical intervention may be indicated. Here we describe the case histories of two patients with unresectable head and neck cancers and frequent debilitating episodes of syncope, who underwent successful treatment of their cancer-related CSS with intracranial division of the glossopharyngeal nerve and rostral rootlets of the vagus nerve.

CASE SERIES

Case 1

A 66-year-old man presented to the emergency room with syncope, hypotension, and a heart rate

in the 30s. He was stabilized with fluid resuscitation and atropine, but despite fludrocortisone and pseudoephedrine therapy and dual-chamber pacemaker placement, he continued to exhibit hypotensive and syncopal episodes. On examination, a right neck mass was found, and an otolaryngology consultation revealed a T4N2a right base of tongue squamous cell carcinoma with neck metastasis encasing the carotid artery and involving the skull base. Moreover, the patient demonstrated ipsilateral weakness of cranial nerves IX, X, and XII but did not have any signs or symptoms of glossopharyngeal neuralgia. Neurological and cardiac evaluations were negative, and the patient began treatment with concomitant chemoradiation therapy for palliation. Nevertheless, debilitating syncopal episodes persisted, requiring multiple hospital admissions. Following a neurosurgical consultation, the patient underwent an uncomplicated suboccipital craniotomy and sectioning of the right glossopharyngeal nerve and rostral one third of the vagus nerve rootlets. Syncopal episodes resolved, and he was discharged home on postoperative day four. At last recorded follow-up in 8 months, the patient was still free of syncope and denied any dysphagia or hoarseness.

Case 2

A 72-year-old man with a complex medical history presented with laryngeal squamous cell carcinoma and was treated with primary radiation therapy. His tumor recurred twice, first requiring a total laryngectomy and then a total glossectomy and pharyngectomy. Two years later the patient experienced his first syncopal episode during which his heart rate was found to be in the 30s. The patient did not complain of any glossopharyngeal neuralgic pain. He was initially stabilized with atropine and fluid resuscitation and was placed on fludrocortisone and midodrine. Both the electroencephalogram and echocardiogram were found to be normal. Despite placement of a dual-chamber pacemaker, the patient continued to experience syncopal episodes. A computed tomography (CT) scan of the neck revealed

Figure 1 Axial computed tomography (CT) image of the neck showing a large left parapharyngeal space mass (arrow).

tumor recurrence in the left parapharyngeal space encasing the internal carotid artery (Fig. 1). The neurosurgical service was consulted and subse-

quently performed an uncomplicated suboccipital craniotomy and intracranial sectioning of the left glossopharyngeal nerve and rostral third of the vagus nerve rootlets (Fig. 2). Syncopal symptoms resolved immediately, and the patient was discharged on postoperative day three. Although the patient died of cancer-related causes 3 weeks later, he was free of debilitating syncopal episodes for this period of time.

DISCUSSION

At least two theories have been proposed to explain the cause of syncope in CSS. The first postulates that a glossopharyngeal-vagal reflex arc is activated by a neuralgic pain attack originating in the sensory afferent fibers of the glossopharyngeal nerve. Transduction of these impulses to the vagus nerve, either through the dorsal motor nucleus of the vagus nerve or through a synaptic connection along the peripheral course of the nerves, stimulates cardioactive mechanisms in the carotid sinus, leading to bradycardia, hypotension, and syncope.⁴ A second theory

Figure 2 Intraoperative video still image demonstrating sectioning of the rostral rootlets (asterisk) of the vagus nerve (X) following the sectioning of the glossopharyngeal nerve (not shown). The spinal accessory nerve (XI) can also be seen.

suggests that the two cranial nerves act independently in response to noxious stimuli. In their study, Barbash and colleagues determined that both vagus nerves can discharge synchronously following afferent neuralgic impulses, resulting in bradycardia or even asystole. Moreover, strong neuralgic stimuli through the glossopharyngeal nerve were found to inhibit vasomotor centers, which lead to peripheral vasodilation and, consequently, to hypotension.⁸ It is believed that peripheral vasodilation explains instances in which cardiac pacemakers, which cannot address hypotension from venous dilation and peripheral pooling, fail to prevent syncopal episodes.^{8,9} These two theories implicate both the glossopharyngeal and vagus nerves in causing syncope, and therefore we elected to divide both the glossopharyngeal nerve and the rostral third (non-motor) rootlets of the vagus nerve, as has been advocated by other authors.^{3,4,10,11}

In addition to the suboccipital intracranial approach to the brainstem, there have been several reports describing alternative extracranial surgical approaches to the glossopharyngeal nerve. However, these methods, which include both cervical¹² and transoral^{13–15} approaches, have been criticized for their difficulty in obtaining adequate exposure and confident nerve identification¹² and failure to address all sensory branches of the glossopharyngeal nerve.⁵ Moreover, in the setting of patients with end-stage head and neck cancer who frequently have already undergone extensive surgical and chemoradiotherapeutic interventions, these approaches may have substantially added risk. Similarly, an extracranial approach may not be achievable secondary to the presence of tumor.

Our review of the literature reveals that the most effective and definitive long-term treatment modality to address CSS is intracranial sectioning of the glossopharyngeal nerve and the upper rootlets of the vagus nerve.^{16–18} The morbidity of this procedure is low, with dysphagia and/or hoarseness seen in as few as 10 to 20% of patients.¹¹ Such side effects were of minimal concern in the patients reported here because of preexisting cranial nerve pareses or prior removal of the larynx. Following

initial stabilization of a patient with new-onset, cancer-related CSS, anticonvulsant therapy should be the first-line therapy. The need for pacemaker placement is controversial, with some authors questioning its efficacy in resolving CSS in the setting of a malignancy.^{1,4,17,19} If debilitating syncope persists, the clinician should consider intracranial glossopharyngeal and limited vagal neurectomy.

CONCLUSIONS

Carotid sinus syncope in head and neck cancer patients is an infrequently encountered syndrome. Nevertheless, when present, the danger of its cardiac sequelae and the nuisance of syncopal episodes can potentially affect both the duration and quality of the life of the patients. Accordingly, we revisit a procedure that safely and effectively addresses CSS and may benefit many head and neck cancer patients with this phenomenon.

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