Neurosurgical management of carotid sinus hypersensitivity

Report of three cases

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Carotid sinus hypersensitivity (CSH) can cause severe bradycardia, hypotension, asystole, and cardiac arrest. Three patients with the combined (mixed) cardioinhibitory and vasodepressor form of CSH were studied with intensive cardiovascular monitoring. After medical management failed, the patients were successfully treated by transection of the glossopharyngeal nerve and upper rootlets of the vagus nerve at their exit from the brain stem. Methods of diagnosis of CSH, and the benefits and limitations of medical and surgical therapies are discussed.

Key Words • carotid sinus hypersensitivity • glossopharyngeal nerve • vagus nerve • vasodepressor reflex

Carotid sinus hypersensitivity (CSH) has been recognized as a serious and debilitating illness for nearly six decades. Patients with CSH have profound bradycardia and/or hypotension in response to distortion of the carotid sinus. The signs and symptoms of CSH can range in severity from dizziness to cardiac arrest. Medical treatment of the vasodepressor and mixed forms of CSH often does not relieve the symptoms. In such cases, surgical treatment may be required; this may be either adventitial stripping of the carotid sinus or transection of the glossopharyngeal nerve as it exits the brain stem. We report three cases of the mixed cardioinhibitory and vasodepressor form of CSH, with a discussion of methods of investigation and treatment.

Case Reports

Case 1

This 63-year-old man was admitted to the hospital with a 3-month history of pain of the left side of the face, neck, and tongue. The pain initially occurred in brief attacks, each of which lasted only a few minutes, but then became nearly constant, particularly in the left temporal-occipital area. He had a 1-month history of dizziness associated with extending his neck when shaving. Mild hypertension had been diagnosed 5 years prior to admission, and had been treated with hydrochlorothiazide.

Examination. Physical examination revealed a left Horner's syndrome and a mass adjacent to the left temporal-mandibular joint. Laboratory tests, a chest x-ray film, and a cerebral angiogram were normal. A computerized tomography (CT) scan of the head revealed the neck mass as the only abnormality. An electrocardiogram demonstrated nonspecific ST-T segment changes. Echocardiography and treadmill stress test were normal. A biopsy of the mass revealed histiocytic lymphoma. While in the hospital, he had several episodes of severe, profound bradycardia (heart rate < 16 beats/min) and hypotension (systolic pressure < 40 mm Hg) coincident with the onset of paroxysms of pain. Administration of atropine and dopamine was required to maintain a normal heart rate and blood pressure. A transvenous pacemaker was placed but had no influence on these syncopal episodes in spite of maintaining the heart rate. Carotid sinus massage revealed a mixed form of CSH. Asystole occurred for 11 seconds during left carotid sinus massage.

Operation. Because drug therapy and cardiac pacing had failed to control the patient's syncope, surgical treatment was undertaken. A left suboccipital craniectomy was performed with transection of the glossopharyngeal nerve and the superior fibers of the vagus nerve. At the time of transection of the nerves, the patient had severe bradycardia and hypotension, which abated after the nerves were cut. His postoperative course was uneventful. Repeat left carotid sinus massage did not elicit
hypotension or bradycardia. He was discharged on the 10th postoperative day, free of syncope. Until his death 18 months later from infection while receiving chemotherapy, he did not experience a recurrence of syncope.

Case 2
This 59-year-old man was admitted to the emergency room after repeated cardiac arrest. After cardiopulmonary resuscitation was instituted and pressor agents were administered, he recovered consciousness. Two months prior to admission, he had undergone radical neck dissection and colon interposition for treatment of laryngeal carcinoma. He had subsequently received irradiation and chemotherapy.

Examination. Physical examination revealed a functioning tracheostomy and gastrostomy, and extensive radiation-induced soft tissue fibrosis in the patient's neck. Neurological examination was normal. Laboratory tests, a chest x-ray film, and an electrocardiogram were normal. A CT scan of his head and an electroencephalogram were also normal. In the hospital he had multiple episodes of bradycardia (heart rate < 32 beats/min) with no palpable blood pressure. A functioning transvenous pacemaker was placed but did not prevent these syncopal episodes. Left carotid sinus massage revealed a mixed form of CSH.

Operation. Surgical treatment was carried out as described in Case 1. The patient was discharged on the 12th postoperative day, free of syncope. During a 6-month follow-up period, there has been no recurrence of syncope.

Case 3
This 60-year-old man was transferred to the Methodist Hospital after five severe episodes of syncope. Nearly 3 years prior to admission, he had undergone maximum irradiation and chemotherapy for squamous-cell carcinoma discovered in a lymph node on the right side of the neck. Nine months prior to admission the tumor in the left nasopharynx advanced. Radium implants were placed adjacent to the tumor, and during this treatment the first syncopal event occurred. During these 10- to 20-minute syncopal episodes, blood pressure fell to 60/0 mm Hg without asystole or tachycardia.

Examination. Physical examination revealed normotensive blood pressures without orthostatic hypotension. The patient had a left Horner's syndrome, radiation-induced soft tissue changes in the neck, and soft tissue swelling in the palate. The remainder of the examination was normal. Laboratory tests, a chest x-ray film, and bundle electrocardiography were normal. Magnetic resonance imaging of the head and neck revealed tumor in the posterior nasopharynx, with involvement of the left carotid sheath near the skull base. While hospitalized, he experienced periodic presyncopal episodes of 30 minutes' duration. Carotid sinus massage testing revealed left-sided CSH, predominantly of the vasodepressor type.

Operation. Surgical treatment was carried out as described in Case 1. The patient was discharged on the 12th postoperative day, free of syncope. During an 8-week follow-up period he has remained free of syncope.

Discussion
Carotid sinus hypersensitivity is an uncommon but dangerous syndrome caused by aberrant autonomic reflex activity. Three types of CSH have been described: 1) cardioinhibitory; 2) vasodepressor; and 3) a mixed form, consisting of both cardioinhibitory and vasodepressor components. Cardioinhibitory activity is preponderant in 50% to 75% of cases. However, both cardioinhibitory and vasodepressor components are generally present. The etiology is thought to be baroceptor hypersensitivity produced by pressure of tumors, adenopathy, scar, aneurysmal dilation, or idiopathic mechanisms.

The afferent and efferent limbs of the reflex arc have been extensively studied. Activity from the carotid sinus baroreceptors is carried to the medulla largely via Hering's nerve, a major branch of the glossopharyngeal nerve. Afferent fibers from the carotid sinus also travel with the hypoglossal nerve and cervical sympathetic nerves before joining the glossopharyngeal nerve. Other afferent fibers are also carried centrally by the vagus nerve. Within the medulla, afferent activity is processed by the solitary nucleus and transmitted to the cardioinhibitory and vasodepressor centers. Efferent signals are communicated to the effector organs primarily via the vagal nerve fibers arising in the dorsal motor nucleus. Efferent sympathetic activity is also integrated into the reflex. Although this description readily applies to the cardioinhibitory form, the neuroeffector pathway involved in the vasodepressor form of CSH is not completely understood.

Most often CSH affects patients aged 40 to 60 years with a 3:1 male predominance. The severity of CSH increases with age and parallels the progression of atherosclerotic disease. Attacks can occur as infrequently as once a year, or on a daily basis, and may have a duration of 30 minutes or more. Symptoms range from mild dizziness to loss of consciousness and seizures. Death from cardiac arrest can occur.

The carotid baroceptor reflex should be evaluated while continuously monitoring intra-arterial blood pressure and the electrocardiogram. Carotid sinus massage is performed for 5 to 20 seconds. Both carotid arteries should be massaged sequentially since CSH is frequently bilateral. A diagnosis of CSH is established by producing asystole for more than 3 seconds and/or a fall in systolic pressure of more than 30 mm Hg with symptoms (or 50 mm Hg without symptoms) during carotid sinus massage. Vasodepressor CSH is identified as a fall in blood pressure that is not accompanied by a reduction in heart rate. Since an acute reduction in heart rate results in a corresponding fall in blood pressure, temporary cardiac pacing to maintain heart rate
constant during carotid sinus massage is needed to distinguish the pure cardioinhibitory form of CSH from the mixed form of CSH. Administration of intravenous atropine in the absence of cardiac pacing does not effectively distinguish cardioinhibitory from vasodepressor mechanisms.8

A number of treatments have been proposed for relief of symptoms of CSH. Irradiation of the carotid sinus was reported to be successful in 58% of cases.4 However, considerable radiation exposure is required and the beneficial effects are generally delayed.18 Paradoxically, radiation has also been implicated as a cause of CSH.3 Drug therapy has been of very limited value, particularly in the vasodepressor form of CSH.1 Ephedrine has been successful in treating the vasodepressor component in some patients, but is hazardous in elderly patients with coronary artery disease.1 Vasolytic drugs have questionable benefit in the cardioinhibitory form of the disease and have significant side effects.17 Ventricular or sequential cardiac pacing primarily controls the cardioinhibitory component of CSH.10,11 Syncopeal symptoms caused by the vasodepressor component are often exacerbated by cardiac pacing due to the secondary loss of atrial augmentation of cardiac output.11 Pacemaker therapy complications may occur in up to 30% of patients.8

Surgical treatment has included sectioning of Hering’s nerve, adventitial stripping of the carotid sinus, and transection of the glossopharyngeal nerve.6 Complete deafferentation of the carotid sinus by Hering’s nerve transection is difficult to achieve, and a significant recurrence rate of up to 50% has been reported.2,4,9

Adventitial stripping of the carotid artery for 1 to 2 cm above and below the sinus has also been used as a treatment.3,18 In patients with atherosclerotic cerebrovascular disease, manipulation of the carotid artery during this procedure can result in stroke.14 In patients with neck masses or with radiation-induced fibrotic changes in the tissues, adventitial stripping may be impossible to perform.13 Recurrence of symptoms following this procedure have been described, presumably the result of incomplete denervation.9 It has also been suggested that CSH may occur after adventitial stripping.13

Suboccipital craniectomy and transection of the glossopharyngeal nerve was first described in 1933 as a method for treating CSH.19 All of the glossopharyngeal afferent fibers can be interrupted using this procedure,2 and the superior rootlets of the vagus can also be divided.15 Complications can include dysphagia, dystonia, and hypegesia and loss of taste sensation at the base of the tongue.13,15 A transient increase in blood pressure can also occur which generally resolves in a few days.18 Other baroreceptors in the vascular system continue to regulate heart rate and blood pressure, even after bilateral glossopharyngeal section.2 The three patients reported here illustrate the combined form of CSH as a sequela of tumors of the neck and angle of the jaw. In fact, CSH was the presenting problem in our first patient, who had histiocytic lymphoma. The combined form of CSH has no effective medical treatment. If symptoms are severe, section of the glossopharyngeal nerve and upper rootlets of the vagus nerve is an effective treatment.

References

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