Carotid sinus syndrome

Report of five cases and review of the literature

FRED L. COHEN, M.D., C. THOMAS FRUEHAN, M.D.,
AND ROBERT B. KING, M.D.

Departments of Neurosurgery and Medicine, State University of New York, Upstate Medical Center, Syracuse, New York

Five cases of carotid sinus syndrome are presented. The syndrome is defined by spontaneous attacks of dizziness and fainting which can be reproduced by graded pressure on one carotid sinus. Three forms of the clinical syndrome, cardioinhibitory, vasodepressor, and cerebral, are discussed. The hyperactive carotid sinus reflex, in which there is ventricular asystole lasting at least 3 seconds or a decrease of more than 50 mm Hg in systolic and diastolic blood pressure, should be differentiated from this syndrome. Treatment modalities include reassurance, drugs, radiotherapy, cardiac pacemakers, and surgical approaches. Carotid sinus syndrome should be considered in the differential diagnosis of unexplained syncope, arteriovenous block, or inappropriate sinus bradycardia.

KEY WORDS • carotid sinus • glossopharyngeal nerve • syncope • cardioinhibitory reflex • cardiac pacemaker • periarterial stripping

Pressure upon the carotid artery was first noted to provoke slowing of the heart and pulse by Czermak in 1866.18 Hering demonstrated that this reflex was initiated by pressure on nerve endings in the carotid sinus rather than direct pressure applied to the vagus nerve.18 Weiss and Baker44 described a clinical syndrome resulting from a hypersensitive carotid sinus reflex which has become known as the carotid sinus syndrome. It consists of spontaneous attacks of dizziness and fainting which may be reproduced by graded pressure on one carotid sinus.

Five cases of carotid sinus syndrome were evaluated and treated at the Upstate Medical Center and Affiliated Hospitals between 1969 and 1975. These cases are presented here with a review of the literature.

Case Reports

Case 1

This 53-year-old man was admitted from the emergency room on October 29, 1974. He had been found lying on the kitchen floor and recalled tilting his head back and feeling dizzy before falling. He had survived an intraoperative cardiac arrest in 1965 during a tympanoplasty for traumatic perforated tympanic membrane.

On admission his general physical and neurological examinations were normal except that, while the neck was being examined...
Carotid sinus syndrome

Fig. 1. Case 1. Rhythm strip, precordial monitor lead. With gentle palpation of the left carotid, the sinus rate transiently slows to 50/min, and there is the sudden onset of complete AV block with ventricular asystole for 6.5 seconds.

without specific carotid massage, the examiner noted transient complete atrioventricular (AV) block on an electrocardiogram (EKG) monitor. During that 8-second period of ventricular asystole the patient felt dizzy and lightheaded, as if he were about to “black out.” Subsequent EKG’s showed first degree atrioventricular (AV) block but were otherwise normal. When the left carotid artery was palpated very gently, he developed third degree heart block (Fig. 1). On October 30, during positioning for skull films with the neck flexed, he had an additional syncopal attack. Brain scan, skull films, and electroencephalogram (EEG) were normal.

A temporary transvenous pacemaker was inserted on the day before surgery and on November 6 he underwent left carotid sinus denervation and periarterial stripping. He did well postoperatively and was discharged on the 18th hospital day. Right carotid sinus massage was without significant effect. He has remained asymptomatic.

Case 2

This 57-year-old man had three syncopal episodes while having a before-dinner drink on Thanksgiving Day. He was admitted on November 28, 1974. He had an 18-month history of palpitations and for the 3 months prior to admission these had been associated with dizziness. General physical and neurological examination on admission were normal. Initial EKG showed atrial fibrillation with rapid ventricular response, which later spontaneously converted to sinus rhythm. Right carotid sinus massage on December 1 produced transient atrial and ventricular asystole and lightheadedness. A temporary transvenous pacemaker was inserted on the day prior to surgery; on December 4 he underwent right carotid sinus denervation and periarterial stripping. Several hours postoperatively he had an episode of hypotension and bradycardia 30 minutes after receiving 50 mg of Demerol intramuscularly for pain. Right and left carotid sinus massage performed on December 6 had no effect. He had no further problems and was discharged 14 days after admission. He has remained well.

Case 3

This 47-year-old woman was admitted from the emergency room on November 29, 1974, after having fallen down a flight of stairs for no apparent reason. She was found unconscious. She had previously been in good health. She was slightly confused but otherwise normal on general physical and neurological examination. Right carotid sinus massage in the sitting position on December 4 produced asystole, faintness, unresponsiveness, and two to three clonic trunk movements. Massage was repeated in the supine position and produced 3.8 seconds of ventricular asystole. Brain scan, skull films, EEG, and EKG were normal. She was asymptomatic during the remainder of her hospital stay and was discharged on December 6. She returned on two separate occasions for 24-hour outpatient cardiac rhythm monitoring (Holter recording), which were normal. She is currently being followed as an outpatient without treatment, pending additional symptoms.

Case 4

This 56-year-old woman was admitted on May 16, 1974, with a 1-week history of lightheadedness and palpitations. She had well-documented, severe, three vessel cor-
FIG. 2. Case 4. Rhythm strip, precordial monitor lead. Gentle carotid sinus pressure causes second degree AV block (Mobitz Type I, with Wenkebach’s phenomenon).

Coronary artery disease with intractable angina. Cardiac catheterization 2 years previously showed left ventricular hypokinesis with end-diastolic pressure of 29 mm Hg. She was taking digoxin, isosorbide dinitrate, nitroglycerin, and quinidine gluconate.

An EKG on admission showed sinus rhythm and was normal. The slightest carotid sinus pressure on the left side produced Mobitz Type I second degree AV block (Fig. 2). The patient was symptomatic during this maneuver. Carotid sinus denervation was rejected as a modality of treatment because of the apparently terminal nature of her coronary artery disease which obviated the need for long-term cure. She required continued digitalization for severe heart failure. A permanent transvenous pacemaker was inserted on May 21. On May 24, she experienced an episode of lightheadedness and palpitations, augmented by head movement. She experienced no further difficulty and was discharged on May 24. She has since suffered a myocardial infarction and is now virtually bedridden.

Case 5

This 68-year-old woman was admitted on January 14, 1969, with a history of repeated syncopal attacks over a 10-year period. She had initially been treated for temporal lobe epilepsy with Mysoline, phenobarbital, and Dilantin, but there was no change in her clinical symptoms. Multiple EEG’s were negative. She sustained bilateral fractured hips during her syncopal attacks. During hospitalization in December, 1968, for an episode of “collapse,” the slightest carotid sinus pressure produced a pronounced second degree AV block (Fig. 3).

On the present admission, gentle left carotid sinus pressure produced 8 seconds of ventricular asystole and syncope. Atropine did not modify this response. A temporary transvenous pacemaker was inserted on February 3, after which bilateral carotid sinus hypersensitivity was again demonstrated and could be abolished by local Xylocaine infiltration. On February 4, right carotid sinus denervation and periarterial stripping were performed without incident. On February 11, she developed an episode of paroxysmal atrial tachycardia which did not respond to left carotid sinus massage. This recurred several times over the ensuing 2 or 3 days. On February 12, the left carotid sinus was denervated; once again, on the next day, she sustained an attack of paroxysmal atrial tachycardia. She was then digitalized, had no further difficulty, and was discharged on February 18. She has been asymptomatic since.

Fig. 3. Case 5. Rhythm strip, standard lead II. Gentle left carotid palpation slows the sinus rate markedly, produces transient third degree AV block, and causes 5 seconds of ventricular asystole.
Carotid sinus syndrome

As early as 1935, Ferris, et al., suggested that cardiac arrest occurring during anesthesia might be due to activation of a hypertensive carotid sinus reflex. Other reports have substantiated this danger. Our Case 1 probably exhibited this phenomenon.

Three separate types of hypersensitive carotid sinus syndromes have been described. The vagal or cardioinhibitory type produces marked sinus bradycardia, AV block, or both. During this period there may be ventricular asystole without an initial drop in blood pressure. Atropine has been used to treat these symptoms by modifying a portion of the efferent arc of the reflex. It has generally proved unsuccessful. This form occurs in about 70% of persons exhibiting carotid sinus hypersensitivity.

The vasodepressor type produces a marked fall in blood pressure without significant bradycardia or AV block. This form is considerably less common, occurring in about 10% of patients. It was formerly treated with epinephrine or ephedrine in an attempt to modify the efferent portion of the reflex and prevent the drop in blood pressure.

The cerebral type, rarest and least well understood, produces the syndrome without marked changes in either heart rate or blood pressure. Although the early literature describes this as a well-defined entity, more recent reports doubt its existence and suggest it may represent inadvertent, transient, manual carotid artery occlusion. No specific medical treatment has been available other than explanation, reassurance, and caution regarding sudden neck movements and wearing tight collars.

Several authors have described a fourth type, a mixture of the cardioinhibitory and vasodepressor types. Classification into distinct entities may be inappropriate since the symptoms rarely occur in a pure form.

One problem in diagnosis and evaluating reports of treatment in the literature is the lack of standardization in applying the clinical test for hypersensitivity. In a widely accepted method the patient lies in the supine position with the head neutral. The examiner's fingers feel for the fusiform carotid sinus or the area of greatest pulsation in front of the sternomastoid muscle at the upper border of the thyroid cartilage. The sinus is pressed lightly for 20 seconds, first on one side and then on the other, never simul-

J. Neurosurg. / Volume 45 / July, 1976 81
taneously, while an EKG is recorded and blood pressure is monitored. If there is no response, stimulus intensity is increased. If there is still no response, the procedure is repeated with the patient in the sitting position. Most positive responses occur within the first 20 seconds and stimulation beyond 30 seconds adds nothing. Although serious complications, such as cardiac standstill and transient or permanent monoplegia or hemiplegia, have been reported from the test itself, these are extremely rare when the test is performed in appropriately selected patients and with EKG monitoring. The test is clearly hazardous in an elderly hypertensive patient with known cerebrovascular or extracranial occlusive vascular disease.

There is relatively uniform agreement as to what constitutes a hyperactive reflex. A clearly pathological reflex response is one in which ventricular asystole lasts 3 seconds or more, or a decrease in systolic and diastolic blood pressure of greater than 50 mm Hg occurs. It should be emphasized that the carotid sinus reflex is a normal physiological mechanism. Marked sinus node slowing and AV conduction disorders can be produced in normal individuals by properly applied vigorous carotid sinus stimulation. This mechanism is extremely useful to cardiologists for termination of certain supraventricular tachyarrhythmias and for aid in diagnosis of some arrhythmias.

The features which distinguish the hypersensitive carotid sinus syndrome from the normal reflex are that the former may produce symptoms spontaneously and may produce cardiac and/or vasomotor effects with far less vigorous carotid sinus stimulation than is required to demonstrate the normal reflex.

There is no agreement on how best to differentiate a hyperactive carotid sinus reflex induced by physical examination from the spontaneously occurring carotid sinus syndrome. A large percentage of people over the age of 40 years have a hyperactive carotid sinus reflex when examined with carotid massage but otherwise have no spontaneous symptoms. Furthermore, most patients with spontaneous syncopal episodes do not have a hyperactive carotid sinus reflex. Thomas required the reproduction of the spontaneous symptom complex by manual carotid sinus stimulation, but Peretz, et al., were only able to reproduce syncope in three of 10 cases, although all patients did demonstrate true hypersensitive carotid sinus reflexes and spontaneous attacks of syncope.

The literature suggests reassurance and caution against rapid neck movements and tight collars for patients with infrequent symptoms. This has been our approach to the patient in Case 3, who had only one spontaneous episode and is currently asymptomatic.

Irradiation was introduced as a form of therapy by Stevenson and Moreton. Greeley, et al., reported 52 cases treated similarly in 1955. No definite rationale for treatment was given. Although the results seemed acceptable (complete-to-moderate relief of symptoms in about 70% with no serious complications), there has been neither widespread acceptance nor utilization of this form of therapy. Some of these patients may simply have had hyperactive carotid sinus reflex on examination but not the spontaneous symptoms which occur with the syndrome.

Surgical approaches were first reported by Weiss and Baker in 1933. In one case, unilateral sinus nerve section was performed and a small, localized, sharply circumscribed tumor pressing on the carotid sinus was found and removed. In the other case, unilateral glossopharyngeal nerve section at its entrance into the medulla was accomplished. By 1935, seven additional cases were treated with stripping of the internal, external, and common carotid sheaths above and below the carotid bifurcation. These three methods remain the currently available surgical approaches. Sectioning the carotid sinus nerve alone may be technically difficult since, at operation, it is frequently poorly defined. Recurrence of symptoms is common when this approach alone is chosen. Glossopharyngeal nerve section is a more serious operation and is associated with nasopharyngeal anesthesia and loss of taste over the posterior third of the tongue. In addition, Ray and Stewart showed that the glossopharyngeal nerve in man is not the only nerve through which afferent impulses of the carotid sinus reflex are transmitted. Periarterial stripping remains the surgical procedure employed most commonly today.

The rationale in all forms of surgical treatment is to interrupt the afferent portion of the
Carotid sinus syndrome
reflex arc. Nathanson\textsuperscript{13} and Sigler\textsuperscript{14} criticized this form of treatment, believing the abnormality to be central or at least not always in the afferent limb of the reflex. It is difficult to discount this criticism since there is no clear understanding of the pathophysiology of the syndrome or the site of abnormality in the reflex arc. Surgical failures are probably due to technically incomplete denervation or incorrect diagnosis. The major surgical risk is the transient postoperative hypertension following denervation, first reported by Weiss and Baker\textsuperscript{2} as being without serious sequelae. There have since been two reports in the literature of serious complications (including one fatality) following unilateral and bilateral denervation.\textsuperscript{8,a,10} This remains a serious but rare complication of surgical denervation.

The form of therapy most recently introduced for the carotid sinus syndrome is cardiac pacing. From a mechanistic viewpoint it should be considered only in the cardioinhibitory or vagal form of the disease, since it can correct only rhythm disturbances. The first reports of its use in carotid sinus syndrome were by Voss and Magnin\textsuperscript{4} and Maur, et al.\textsuperscript{11} Peretz, et al.,\textsuperscript{14} reported an additional 10 cases with excellent results. The relative disadvantages of pacing include the need for life-long surveillance of the pulse generator and periodic battery replacements.

Syncope is not an uncommon complaint in patients referred to a neurosurgeon for evaluation. Differential diagnosis includes simple vasodepressor fainting, orthostatic hypotension, blood loss, cardiac arrhythmias, seizures, hypoglycemia, and vertebrobasilar ischemia. The carotid sinus syndrome should be considered in such cases, and in cases of unexplained AV block or inappropriate sinus bradycardia. Although the hypersensitive carotid syndrome is not a common disorder, the treatment is different than for other causes of AV block or unexplained syncope and offers lasting relief from the distressing and potentially fatal symptoms.

References


hypertensive crisis after bilateral carotid sinus

20. Stevenson CA: The use of roentgen therapy in
the carotid sinus syndrome. Radiology 32:
209–214, 1939

21. Stevenson CA, Moreton RD: A subsequent
report on roentgen therapy in the carotid sinus
syndrome. Radiology 50:207–210, 1948

22. Thomas JE: Hyperactive carotid sinus reflex
and carotid sinus syncope. Mayo Clin Proc
44:127–139, 1969

23. Voss DM, Magnin GE: Demand pacing and
carotid sinus syncope. Am Heart J 79:
544–547, 1970

24. Weiss H, Baker JP: The carotid sinus reflex in
health and disease. Its role in the causation of
fainting and convulsions. Medicine 12:
297–354, 1933

This study was supported in part by NINCDS
Training Grant N05 T01 NS 05605.

Address reprint requests to: Fred L. Cohen,
M.D., Department of Neurosurgery, State University
of New York, Upstate Medical Center, 750 E.
Adams Street, Syracuse, New York 13210.