Case Reports

Critical Carotid and Vertebral Arterial Occlusive Disease and Cough Syncope

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Background and Purpose: Cough syncope typically occurs in patients with known chronic lung disease. The mechanism usually involves a combination of decreased venous return, increased cerebrospinal fluid pressure, and secondary hypocapnia, all resulting in cerebral arterial vasoconstriction. Cough syncope has not in the past been associated with occlusive cerebrovascular disease.

Case Description: We describe a 50-year-old man with a 6-month history of episodes of loss of consciousness during paroxysms of coughing. Physical examination showed asymmetrical upper extremity blood pressures and carotid and subclavian artery bruits. Pulmonary function studies were normal. Ultrasound and angiography showed total occlusion of the left common carotid artery, right internal carotid artery, and right vertebral artery; tight stenosis of the right subclavian artery; and a hypoplastic left vertebral artery. The patient had a left subclavian-to-left common carotid artery bypass and has had no syncope since that time.

Conclusions: To our knowledge, this is the first reported case of cough syncope and severe cerebrovascular disease in which surgery led to amelioration of symptoms. Cerebrovascular occlusive disease may contribute to cough syncope. (Stroke 1992;23:1017–1020)

KEY WORDS • cerebrovascular disorders • physiology • syncope

Cough syncope is a well-known cause of episodic loss of consciousness.1,2 Wright and McIntosh3 first described the physiological changes accompanying cough that might result in syncope. Since then, it has been postulated1 that a vasomotor reflex similar to that seen in micturition syncope4 or in the recently described neurally mediated vasovagal syndrome5 may cause syncope in patients with paroxysms of coughing. Typically, cough syncope occurs in patients with long-standing chronic obstructive lung disease.1,3 An association with cerebrovascular disease has not, to our knowledge, been reported. In this report, we describe a man with a 6-month history of cough syncope and normal pulmonary function studies who was found to have critical extracranial carotid and vertebral arterial occlusive disease. Neurovascular surgery resulted in increased transcranial Doppler flow velocity and an amelioration of his symptoms.

Case Report

A 50-year-old white man came to us in March 1990 after 1 year of syncope during coughing. His coughing spells were not precipitated by any recognized activity. The coughing spells were brief and accompanied by tingling pain in the right arm, palpitations, then sudden loss of consciousness, after which he would rapidly awaken and return to normal. Spells averaged one per week and interfered significantly with his ability to work in his profession as a cook. He had used alcohol heavily in the past but had no other past important illnesses. He was a heavy cigarette smoker and had some exertional dyspnea and morning sputum production. There was a family history of coronary artery disease.

On initial examination, the patient’s blood pressure was 204/110 mm Hg. Cardiac and neurological examinations were normal. Diagnoses of hypertension and cough syncope were made, and enalapril was prescribed. The patient had two more spells of loss of consciousness during the next 2 weeks. Record review showed that he was seen 2 years previously for an episode of dizziness. At that time, blood pressure in his arms was asymmetrical, and a diagnosis of subclavian steal syndrome was entertained. A follow-up visit was scheduled but not kept. Physical examination now showed a blood pressure of 160/100 mm Hg in his left arm and 120/90 mm Hg in his right arm. A loud right subclavian bruit and bilateral carotid bruits were noted. The left carotid pulse was diminished. The remainder of the general and neurological examination was normal. His antihypertensive regimen was changed: enalapril was discontinued, and a calcium channel blocker (verapamil) was substituted.

An electrocardiogram was normal. Chest x-ray showed cardiomegaly but was otherwise normal. Pulmonary function testing was normal for age, sex, and height, with a normal forced expiratory volume-to-forced vital capacity ratio (109% predicted) and an essentially normal diffusing capacity when corrected for alveolar volume (83% predicted). Residual volume was also normal (106% predicted). Expiratory reserve volume was markedly diminished (33% predicted), consis-
tent with the patient’s obesity. A 24-hour Holter monitor showed an average of 38 ventricular premature beats per hour with multifocal morphologies but no pairs, bigeminy, or runs of ventricular tachycardia. The patient had no symptoms during Holter monitoring. An echocardiogram showed concentric left ventricular hypertrophy with normal left ventricular and valvular function. Cranial computed tomographic scan showed slight hypodensity in the white matter bilaterally but no areas of infarction.

Extracranial and transcranial Doppler and carotid duplex scanning showed severe carotid, vertebral, and subclavian artery occlusive disease (see Table 1). Provocative tests to study cerebrovascular reserve function were not performed. Cerebral angiography demonstrated total occlusion of the right internal carotid artery (Figure 1), occlusion of the right vertebral artery at its origin (Figure 2), and a preocclusive stenosis near the origin of the right subclavian artery (Figure 2). The left common carotid artery was occluded just beyond its origin from the aortic arch (Figure 3). A hypoplastic left vertebral artery was seen to reconstitute in the neck with slow antegrade flow (Figure 3). On delayed views, the right external carotid artery was found to reconstitute the left carotid bulb by way of collaterals through the left external carotid artery. A left internal carotid artery stenosis was present above the bulb.

The severity of the extracranial occlusive disease and the profound impact of the illness on the patient’s quality of life led to a decision to perform reconstructive vascular surgery. In May 1990, the patient underwent left carotid endarterectomy and placement of a Goretex graft between his left subclavian artery and his left common carotid artery. During surgery, clamping of the left external carotid artery led to profound electroencephalographic changes that were reversed with emergency shunting of the left subclavian to left distal internal carotid artery. The procedure was well tolerated by the patient, and there were no neurological sequelae.

During the following 2 months, the patient remained free of any cough-related symptoms. He discontinued cigarette smoking but continued to have paroxysms of coughing. Over the next 12 months, he developed some cough-related dizziness, never progressing to loss of consciousness. Transcranial Doppler studies performed postoperatively showed increased velocities bilaterally in the carotid siphon and the cerebral arteries (anterior, middle, and posterior cerebral arteries) (Table 1). The intracranial posterior circulation velocities did not change significantly after the operation. Cessation of syncope and normalization of blood flow velocities in

![Figure 1. Selective right carotid arteriogram demonstrating abrupt occlusion (white arrow) of the right internal carotid artery above bifurcation.](http://stroke.ahajournals.org/)

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ICAS, internal carotid artery siphon; MCA, middle cerebral artery; ACA, anterior cerebral artery; PCA, posterior cerebral artery.

**TABLE 1. Comparison of Normal, Preoperative, and 6-Month Postoperative Transcranial Doppler Studies**
FIGURE 2. Selective innominate artery injection demonstrating occlusion of right vertebral artery at origin (black arrowhead) and preocclusive stenosis of right subclavian artery just beyond origin (white arrow).

FIGURE 3. Aortic arch arteriogram demonstrating occlusion of left common carotid artery just beyond origin (white arrow) and hypoplastic left vertebral artery with slow antegrade flow (black arrowheads).

the carotid circulations bilaterally provide some evidence for normalization of blood flow.

Discussion

Cough syncope is a relatively uncommon but not rare type of syncope.1-3 Because the physiological changes associated with cough that cause syncope have been described so clearly, there is often no diagnostic workup pursued in patients with cough-induced syncope who have no other obvious cause for loss of consciousness. The important features of the present case include the dramatic abnormalities in cerebral vasculature found in this patient with cough-induced syncope, rapid amelioration of symptoms after reconstructive neurovascular surgery, and persistence of minor cough-related light-headedness without syncope postoperatively. Cerebrovascular disease should be included in the differential diagnosis of patients with cough-induced syncope.

An early text on cough syncope6 describes a large number of typical cases of cough syncope that occur in the presence of chronic lung disease. However, one patient (case 26) had cough syncope in the presence of chronic lung disease only after “cerebral damage from vascular disease” had occurred. Cough syncope usually results from decreased venous return and a decrease in cardiac output resulting from the Valsalva maneuver, an increase in cerebrospinal fluid pressure, and hypocapnia due to coughing. The latter two phenomena result in cerebral arterial vasoconstriction, whereas the drop in venous return due to the Valsalva maneuver results in a drop in cerebral blood flow. In our patient with critical cerebrovascular disease and profound changes in the electroencephalogram during vascular surgery, any reduction in cerebral blood flow appeared to be sufficient to cause loss of consciousness.

Loss of consciousness occurs when an insufficient quantity of blood reaches the brain stem or both cerebral hemispheres. Supplying blood to the brain can be likened to the task of supplying adequate water under pressure to allow a satisfactory shower on the third floor of a home. Necessary are adequate volume of water in the water tank and system (normovolemia); effective water pump (good cardiac pump function); adequate pressure in the system (normotension); and open, adequate-sized pipes to the region of supply (open extracranial and cranial vascular bed). The system works in tandem. If pressure, volume, or pump function is compromised, adequate pipes become even more important. When the pipes are nearly completely blocked, any decrease in general perfusion is magnified.

In our patient with severely compromised extracranial arteries, the transient drop in perfusion accompanying cough was sufficient to cause syncope. After reconstructive vascular surgery, the transient fall in
perfusion caused minor dizziness but was inadequate to cause loss of consciousness or tone. Similar tandem problems were also described in series of cases reported by Dobkin and Yanigahara and colleagues. Dobkin reported seven cases of severe occlusive cerebrovascular disease and transient neurological symptoms provoked by orthostatic hypotension. Five of the patients had bilateral severe carotid artery or basilar artery occlusive disease. None of these patients fainted. Yanigahara described three patients with recurrent spells of brief loss of consciousness. Each had severe bilateral occlusive carotid artery disease, and spells occurred while the patients stood, walked, or sat, often after a change in position. Some patients also had transient visual or cerebral symptoms.

In patients with syncope, extracranial vascular disease and systemic perfusion and vascular reflex disorders should not be viewed as vying differential diagnostic considerations. Perfusion and vascular status work in tandem and sequentially, and each contributes to adequate brain blood supply. Each should be considered even when the other is present. It is important to consider cardiac function, blood pressure, blood volume, reflex-mediated changes, and extracranial vascular patency in all patients with recurrent syncope.

A recent discussion of cough syncope focused on the impact of cough in patients with pulmonary arterial hypertension. In these patients, critical drops in cardiac output accompany the Valsalva maneuver of coughing. However, our patient had essentially normal pulmonary function studies and no physical signs of pulmonary hypertension. Thus, the mechanism in our patient is likely to be different from that described in this review.

Subclavian-to-external carotid artery bypass has been previously described for symptomatic severe cerebral ischemia from common and internal carotid artery occlusive disease. In a series of nine patients, local neurological symptoms were present in all patients before surgery and were markedly diminished by surgery. Only two of these patients had had syncope, and both also had experienced episodes of amaurosis and other transient ischemic attacks. The uniqueness of our case lies in the relief of isolated syncope by reconstructive neurovascular surgery.

The prevalence of neurovascular disease in patients with cough syncope is speculative at present. Neurovascular disease is a rare cause of isolated syncope (that is, syncope without accompanying focal neurological signs). New, noninvasive neurovascular imaging studies, such as the transcranial Doppler studies used in the current report, may improve our knowledge of the association between neurovascular disease and syncope.

Acknowledgments
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References
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