Case Report

Cough syncope in a 43-year-old woman with glomus jugulare tumor

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A B S T R A C T

We present an unusual case of recurrent cough syncope in a 43-year-old woman, which was initially thought to be seizures. Syncope episodes were triggered by paroxysms of cough and were characterized by unresponsiveness and myoclonic jerks in her extremities. She had a left-sided glomus jugulare tumor that extended into the posterior cranial fossa with evidence of worsening communicating hydrocephalus on brain imaging. We postulate that bouts of cough produced increased intracranial pressure both by raising intrathoracic and intraabdominal pressures as well as by transient obstruction to cerebrospinal fluid flow secondary to intermittent tonsillar herniation during cough. This resulted in diffuse decrease in cerebral blood flow causing syncope. The patient’s syncopal episodes decreased in frequency once an external ventricular drain was placed followed by a ventriculoperitoneal shunt. Search for factors that can increase intracranial pressure seems warranted in patients with recurrent cough syncope.

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1. Introduction

Syncope is caused by transient diffuse cerebral hypoperfusion and is characterized by transient loss of consciousness with a rapid onset followed by spontaneous and complete recovery. Clinical features of syncope may include myoclonic jerks which are often multifocal and asynchronous, convulsions, and urinary incontinence [1], making it difficult to differentiate from epileptic seizure by clinical features alone. Significant fluctuations in cerebral perfusion pressure are prevented by autoregulation of cerebral circulation, but there may be conditions where such mechanism may not compensate adequately. Cough syncope, a rare form of syncope, may be a result of transient failure of the cerebral autoregulatory mechanism to cope with sudden decrease in cerebral blood flow. We present an unusual case of recurrent cough syncope, which was initially diagnosed and treated as seizures, in the context of a left-sided glomus jugulare tumor, a benign paraganglioma.

2. Case report

A 43-year-old right-handed woman with history of glomus jugulare tumor in the left jugular fossa with intracranial extension into the posterior cranial fossa was transferred from another hospital for recurrent seizure-like spells. Her tumor was diagnosed in 2008. She had a 90% surgical resection of the tumor done in 2011 followed by radiation therapy in September 2012. She began to have episodes regarded as for seizures in December 2012.

Her episodes occurred multiple times a day (7 per day on average) during wakeful state. They were triggered by coughing (usually a bout of cough) and were characterized by staring and unresponsiveness as well as stiffening of the body with mild shaking of both upper extremities. The whole episode duration was several minutes according to her husband. She reported urinary incontinence associated with a few of these episodes. She was diagnosed with epileptic seizures but continued to have episodes during treatment with the antiepileptic drugs (AEDs) phenytoin, levetiracetam, and lamotrigine. Escalation of AED therapy made her increasingly drowsy, and she was on all three aforementioned AEDs at the time of presentation.

Her physical examination was remarkable for excessive drowsiness, mild dysarthria, right sixth cranial nerve palsy, mild hypertonia with hyperreflexia in the lower extremities (left more than right), and bilateral ankle clonus. She had a lumbar puncture done at the outside hospital, and the opening cerebrospinal fluid (CSF) pressure was reported to be 25 cm. Cerebrospinal fluid laboratory work was unremarkable. Blood work was also unremarkable except for mild anemia (hemoglobin: 9.4 g/dL), mild hyponatremia (132 mEq/L), and mild hypokalemia (3.1 mEq/L). Antiepileptic drug levels were within therapeutic range (free phenytoin: 1.3 μg/mL, levetiracetam: 5.9 μg/mL, and lamotrigine: 23 μg/mL).

She was admitted and was monitored with continuous EEG (cEEG) with video. Baseline blood pressures ranged from 96 to 121 systolic and 58 to 61 diastolic. Three episodes were recorded on cEEG over the
Fig. 1. EEG (longitudinal bipolar montage, LFF at 1 Hz, HFF at 70 Hz, sensitivity: 7 μV/mm, 20 s/page) associated with a typical syncopal episode: A, onset; B and C, during the episode; and D, offset.
next day. All started with a bout of cough when the patient was lying in bed (in supine or in lateral position) which was followed by brief (less than a minute) distal upper extremity tremor and subtle proximal upper extremity myoclonic jerks and prolonged unresponsiveness for up to 10 min. All of these episodes were associated with hypotension (72–78/31–47 mm of Hg as revealed by continuous arterial pressure monitoring) and bradycardia (54–59 bpm). The EEG during the spells was characterized by generalized synchronous and asynchronous high amplitude 1- to 2-Hz delta activity which progressed to generalized attenuation and then transitioned to generalized delta activity again with recovery (Fig. 1). A head CT showed recurrence of the glomus jugulare tumor and communicating hydrocephalus. An external ventricular drain (EVD) was placed in the evening of hospital day 2. After placement of the EVD, her drowsiness gradually started to improve, and episodes decreased in frequency to one per day. Antiepileptic drug taper was started.

An MRI of the brain (Fig. 2) done on hospital day 3 showed an enhancing T2 hyperintense left skull base mass in the region of the left jugular foramen with extension into the posterior cranial fossa and below the base of the skull. Brain imaging showed evidence of hydrocephalus that had increased compared with her previous brain imaging done 2 months back. A ventriculoperitoneal shunt was placed on the seventh hospital day. Antiepileptic drugs were stopped. Her mental status continued to improve, and she had only one mild episode triggered by cough during the next two days before her discharge. Repeat surgical resection of the tumor was recommended by the otolaryngology team, which the patient declined.

3. Discussion

Based on the clinical features and EEG findings, the episodes observed in our patient are most consistent with cough syncope [2]. The mechanism underlying cough syncope is not definitely established, but it is postulated that coughing increases intrathoracic and intraabdominal pressures leading to a transient increase in ICP [3]. Increased ICP, in turn, causes a decrease in cerebral perfusion pressure which, if it drops below a critical level, may result in global cerebral hypoperfusion leading to syncope. Transient cerebral circulatory arrest has been demonstrated by transcranial Doppler measurements during cough syncope [4]. Our patient also had a drop in blood pressure and heart rate but probably not sufficient to cause syncope by itself. Cough syncope has been associated with posterior fossa mass lesions or tonsillar herniation and with hydrocephalus. Our patient had a glomus jugulare tumor with extension into the posterior cranial fossa. It may be speculated that bouts of cough caused transient herniation of cerebellar tonsils obstructing CSF flow that further contributed to the increase in ICP during coughing. Decrease in frequency of events following placement of EVD to relieve ICP lends support to this notion.

Paragangliomas are rare tumors of extramedullary chromaffin cell origin that most commonly occur in the head and neck region [5]. Catecholamine-hypersecreting paragangliomas are uncommon in the head and neck region, and most patients (95%) with hypersecreting paragangliomas have hypertension [5]. Hypotension accompanying syncope observed in our patient was not orthostasis-related (the patient was always supine during spells) and was most likely related to cough. Chao et al. [6] identified a subset of patients with cough syncope who lacked a blood pressure overshoot (expected response) after the relief of straining during Valsalva maneuver. These patients also showed prolonged hypotension after a cough. The authors postulate that cough syncope in these patients might be the result of delayed recovery from hypotension that follows a paroxysm of cough, and this was likely contributing to global cerebral hypoperfusion in our patient.

This case highlights the fact that cough syncope, a rare form of syncope, may be associated with intracranial mass lesions that indirectly exaggerate the increase in ICP in response to cough. Glomus caroticum tumor presenting as recurrent unexplained syncope [7] and posterior fossa meningioma presenting as recurrent cough syncope [8] have been described. Recurrent cough syncope should trigger search for factors, including brain tumors, with the potential to cause transient elevation in ICP. This case also illustrates an important role for cEEG monitoring with video in distinguishing syncope from seizures in cough syncope cases.

Conflict of interest

The authors report no conflict of interest.

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