Intractable Cough as a Presenting Symptom of Chiari Malformation

Ashutosh Kaushal1  Surya Kumar Dube1  Hemanshu Prabhakar1

1Department of Neuroanaesthesiology and Critical Care, All India Institute of Medical Sciences, New Delhi, India

Address for correspondence Surya Kumar Dube, MD, DM, Department of Neuroanaesthesiology and Critical Care, 7th Floor, Neurosciences Center, All India Institute of Medical Sciences, New Delhi 110029, India (e-mail: surya.dube@yahoo.co.in).


In adults, chronic cough is usually defined as cough lasting for 8 weeks or more, and the common causes include allergies, cold, rhinitis, sinusitis, asthma, acid refluxes, respiratory tract infections, medications, and lung diseases. Eliminating the underlying cause is the main stem of treatment for chronic cough. Occasionally, chronic cough persists despite extensive pulmonary workup and treatment. We report an interesting case of persistent intractable cough as the presenting symptom of Chiari malformation type 1.

A 45-year-old woman presented with chronic paroxysmal nonproductive cough for 2 years, difficulty in swallowing for 6 months, and regurgitation of food for 6 months. Cough was associated with on and off suboccipital headache, and it persisted despite initial pulmonary workup and treatment for common causes of cough. After admission, magnetic resonance imaging (MRI) done to evaluate her headache revealed cerebellar tonsillar herniation with cerebellar tonsil reaching to C2 level suggesting of Chiari malformation type 1. The patient underwent foramen magnum decompression, along with suboccipital resection of cerebellar tonsil with lax duraplasty under standard general anesthesia technique. The intraoperative anesthetic, surgical, and postoperative courses were uneventful. The patient demonstrated significant symptom improvement over next 15 days and at subsequent follow-up visits.

Cough-triggering headache is an unusual finding and is classified as primary cough headache and symptomatic cough headache.1 Primary cough headache is a headache that is precipitated by coughing or straining without having any associated intracranial disorders and it can last up to 30 minutes.1,2 Symptomatic cough headache, on the other hand, is associated with some underlying abnormality and is associated with concomitant symptoms, higher pain intensities, and variable duration of headache.1 The most common underlying abnormality in patients with symptomatic cough headache is Chiari type 1 malformation. The other probable abnormalities include posterior fossa lesion, obstructive hydrocephalus, spontaneous low cerebrospinal pressure, cerebral aneurysm, and carotid or vertebra basilar vascular pathologies.1 In our case, the underlying cause of the occasional headache was Chiari malformation type 1.

Chiari malformations are structural defects in the skull base and cerebellum. They consist of a downward displacement of the cerebellar tonsils through the foramen magnum. Though suboccipital headache is the most common symptom of Chiari malformation type 1, patients may present as well with other symptoms such as weakness, numbness of the extremities, gait abnormality, loss of temperature sensation, cranial nerve dysfunction, diplopia, tinnitus, dysphasia, apnea, and dysarthria.3 Brainstem compression is present in approximately 20% of the patients with Chiari malformation leading to laryngeal sensory loss, dysphagia, swallowing dysfunction, and recurrent chronic aspiration manifesting as food regurgitation and chronic cough.4 Chronic cough has previously been described as a secondary symptom in an adult patient having Chiari malformation type 1 presenting with progressive dysphagia.5 In our case, the patient presented to us with symptoms of chronic cough associated with dysphagia, regurgitation of food, and occasional suboccipital headache. Though the patient’s symptomatology was indicative of underlying neurologic abnormality, the primary and most distressing symptom of the patient was persistent cough. Hence the patient underwent exhaustive pulmonary workup before presenting to us. However, chronic cough is rarely reported as a presenting complaint in patients of Chiari malformations, and a definitive explanation for this is missing.

The cough reflex is considered as a brainstem reflex in which sensory input is carried by the vagal or laryngeal nerve as an afferent pathway to the brainstem, which ultimately converts the sensory input into a complex motor output to produce cough.6 Hence, the brainstem compression by cerebellar tonsil can trigger a chronic cough by directly irritating one of the above neural components associated with the cough reflex. Our assumption is further supported by the fact that the surgical resection of herniated cerebellar tonsil led to alleviation of patient’s chronic cough, suggesting that brainstem compression was the probable cause of the symptoms.
Neurosurgical patients and for that matter any patient with active cough are generally not considered appropriate for elective surgery because of the problems related to increased airway reactivity. With this case report, we want to make the anesthesiologist aware of the fact that chronic cough can be a presenting symptom of Chiari malformation type 1 that can be treated by surgery alone. Chiari malformation type 1 may also be considered in the differential diagnosis of any patient presenting with intractable cough without having any primary pulmonary abnormality.

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Erratum: Article has been corrected as per erratum published on June 28, 2018. DOI of the erratum is 10.1055/s-0038-1666890. Author Ashutosh Kaushal’s name was appearing incorrectly. Author’s correct name appears as in the article.