Chiari Type I Malformation Presenting as Cough in Older Children

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ABSTRACT

OBJECTIVE: To highlight an unusual central cause of cough in older children and underscore the clinical manifestations that might point towards such a diagnosis.

RESULTS: We present two case reports of older children, a case report (Case 1) of a 17-year-old with chronic cough and mild dysphonia, and a second case report (Case 2) of an 8-year-old with persistent cough.

RESULTS: Case 1: Physical examination revealed right cranial nerve XII palsy that was most prominent during cough. Video fiberoptic examination and operative bronchoscopy were normal. MRI revealed Chiari type I malformation. In each case, surgical decompression provided symptom improvement. Pre- and post-operative video fiberoptic examinations, voice recordings, laryngeal EMG and imaging are presented and pertinent literature is reviewed. Case 2: Fiberoptic examination revealed paresis of the left true vocal cord. Laryngeal electromyography (EMG) demonstrated prolonged fibrillation of the left thyroarytenoid and cricothyroid muscle and delay of the right thyroarytenoid muscle. Given finding consistent with bilateral cranial neuropathy, magnetic resonance imaging (MRI) was obtained and revealed Chiari type I malformation.

CONCLUSION: Chronic cough is a rare presenting symptom in children with Chiari Type I malformation. We emphasize the significance of awareness for unusual cases of cough to aid in the correct identification of Chiari type I malformation in children.

INTRODUCTION

Approximately 30 million child visits to the doctor occur each year for cough in the United States1. The majority of these visits are to primary care physicians, however, children are often referred to otolaryngology for chronic or persistent cough that is defined as lasting greater than 4 weeks without resolution2. The most common causes of chronic cough identified in children include asthma, gastroesophageal reflux disease (GERD), postnasal drip syndrome, environmental irritants, postnasal drip syndrome, and intrathoracic hypertension3,4, although common causes of cough have been shown to vary with age5,6.

In older children, defined as age greater than 6 years, the most common causes of chronic cough include cough-variant asthma, psychogenic cough, and sinusitis, in the setting of a normal chest radiograph7. In evaluating and treating children with chronic cough, an etiologically-based approach to treatment is considered the standard of care, which highlights the importance of uncovering a specific cause8. Following thorough history and physical exam, testing is directed at uncovering the most likely diagnosis. Generally, all patients should receive a chest radiograph and spirometry testing (if older than 6 years of age). Empirical-based treatment approaches with use of antibiotics, nasal steroids, inhaled steroids, or anti-reflux medications are generally not advocated for chronic cough in children unless rhinitis, asthma, or GERD are suspected, respectively, based on randomized controlled trials and systematic review9,10.

An empiric trial of bronchodilators, however, has been advocated for in order to identify cough-variant asthma in some patients without additional symptoms11.

On initial evaluation, it is often useful to classify chronic cough as specific or non-specific based on patient history and physical exam, where specific findings may point toward an underlying pulmonary or systemic disorder. In cases of specific cough, work-up may then continue to uncover a primary pulmonary or systemic disorder. In cases of non-specific cough, a review, counseling, watch, and wait approach is suggested.

With the above approach to evaluating chronic cough, the majority of causes are uncovered and symptoms improved with appropriate treatment. Occasionally, however, chronic cough may persist despite extensive work-up and treatment. We present two cases of chronic cough in older children that persisted despite work-up and treatment for common causes of cough and were ultimately diagnosed and treated for Chiari type I malformation.

RESULTS

CASE 1

A 17-year-old male presented with ten years of chronic cough, progressive over the past year. In addition to cough, he occasionally developed breathy dysphonia. He reported exacerbation of his symptoms with exercise to the point where he could no longer run without severe coughing spells. He did not report cough at night. Prior evaluation had included chest radiograph, chest computed tomography, spirometry, allergy testing, modified barium swallow, and operative laryngoscopy and laryngoscopy, all of which were within normal limits. Bronchovascular larynx from his bronchoscopy reportedly grew out multiple bacterial organisms for which he was treated with a prolonged course of antibiotics without resolution of symptoms. He underwent cough/aphthophophsy monitoring that was positive for reflux and has been treated with high dose omeprazole for several years with minimal symptom relief. Several years earlier, he was also started on epinephrine inhaler fluticasone and inhaled albuterol as needed while exercising, both of which have had limited impact on his symptoms.

RESULTS

CASE 2

An 8-year-old presented with two months of chronic “barking” cough that initially began following an upper respiratory infection. In addition to cough, she reported global loss of sensation and intermittent sore throat. She presented to emergency department twice for uncontrollable cough and subjective difficulty breathing, which reportedly improved after nebulized treatment. She was initially treated for diagnosis of cough with oral steroids without improvement in her baseline cough. She was also treated with empiric antibiotics for sore throat without improvement. She had been on amoxicillin for 18 months for a diagnosis of GERD. Chest radiograph and spirometry were within normal limits.

Physical examination revealed right cranial nerve XII palsy that was most prominent during cough. Video fiberoptic examination and operative bronchoscopy were normal. MRI revealed Chiari type I malformation with herniation of the cerebellar tonsils 7-8 mm into the cervical canal. The patient was referred for surgical decompression. Following posterior fossa decompression with duroplasty, she demonstrated complete resolution of symptoms at six week follow-up. She did develop return of mild, intermittent coughing spells following an upper respiratory infection after this time, but has remained improved from her baseline six months following her surgery.

RESULTS

CASE 2

Chronic cough is a rare presenting symptom in children with Chiari Type I malformation. We emphasize the significance of awareness for unusual cases of cough to aid in the correct identification of Chiari type I malformation in children.

REFERENCES