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Case report

Chiari malformations: An important cause of pediatric aspiration

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ABSTRACT

Chronic aspiration poses a major health risk to the pediatric population. We describe four cases in which work up for chronic aspiration with a brain MRI revealed a Chiari I malformation, a poorly described etiology of pediatric aspiration. All patients had at least one non-specific neurologic symptom but had swallow studies more characteristic of an anatomic than a neurologic etiology. Patients were referred to neurosurgery and underwent posterior fossa decompression with symptom improvement. A high index of suspicion for Chiari malformation should be maintained when the standard work up for aspiration is non-diagnostic, particularly when non-specific neurologic symptoms are present.

1. Introduction

Aspiration is the passage of ingested material, refluxed contents, or oral secretions past the vocal folds into the lower respiratory tract. Recurrent or chronic aspiration poses a major health risk to the pediatric population, resulting in serious infection, chronic lung disease, and even death. Patients with aspiration may present with choking or cyanosis with feeding, recurrent croup-like cough, failure to thrive, recurrent pneumonia, reflux, wheezing, or interstitial lung disease [1–3]. The exact incidence of swallowing dysfunction associated with aspiration is unknown as the clinical signs and symptoms are frequently subtle. In pediatric populations, 70% of patients with aspiration do so without a correlated cough (silent aspiration) and thus may go undiagnosed [4–8]. Aspiration is, however, the most common cause of recurrent pneumonia in children and is the leading cause of death among the neurologically impaired [9,10].

Swallowing is a highly complex process. Imperfect coordination, anatomic abnormality, and lack of airway protective reflexes can all result in aspiration. Given the vast differential, a systems-based approach helps to categorize etiologies into anatomic, neurologic, behavioral, cardiorespiratory, or metabolic pathologies [11,12]. Following a thorough history and physical examination, testing is directed at uncovering the underlying abnormality. In general, patients should receive a chest radiograph to assess pulmonary disease and undergo a videofluoroscopic swallowing study (VFSS) and/or a fiberoptic endoscopic evaluation of swallowing (FEES), depending upon the experience and preference of the health care team and institution, in order to systematically evaluate each phase of swallowing and objectively assess laryngeal penetration and aspiration.

If there is no clear neurologic abnormality on physical exam or VFSS/FEES that could account for the aspiration and/or an anatomic cause is suspected, further evaluation in the operating room with a multidisciplinary team is undertaken. This includes a direct laryngoscopy, rigid bronchoscopy, flexible bronchoscopy with bronchoalveolar lavage, and esophagogastroduodenoscopy (EGD). Occasionally, however, the cause of aspiration remains unknown despite this extensive work up. In this case series, we present four children who fall into this category that were ultimately found to have Chiari I malformations (CMI) on MRI and benefitted from surgical decompression. This is a poorly described etiology of pediatric aspiration in the literature. We aim to describe their presentation and work up, discuss when such a disorder should be considered, and review what interventions may be offered.

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2. Methods

Four patients presenting to the Massachusetts Eye and Ear Infirmary pediatric aerodigestive multidisciplinary clinic between October 2010 and February 2015 for chronic cough or aspiration that were ultimately diagnosed with a CMI were identified by the senior author. During this time period there were 2000 new patients who presented to the aerodigestive multidisciplinary clinic and of those 642 underwent triple endoscopy (laryngoscopy, bronchoscopy, EGD). Institutional Review Board approval from the Human Subjects Research Committee at Massachusetts Eye and Ear Infirmary was obtained. The four identified patient's charts were reviewed for patient presentation, diagnostic work up, treatments, and outcomes. Experts in the fields of otolaryngology, speech and language pathology, neurology, neuroradiology, and neurosurgery were asked to review the cases and provide insight into CMI associated with aspiration. A literature review of the topic was also performed.

The MRI images were evaluated by our institution's pediatric neuroradiologist for tonsillar decent, cerebral spinal fluid (CSF) restriction at the craniocervical junction, and the presence or absence of a syrinx. Tonsillar descent was measured by drawing a line between the basion and the opisthion on the sagittal images and measuring perpendicular to this line down to the tips of the right and left cerebellar tonsils. The average of the two sides was recorded. CSF restriction was evaluated on the sagittal T2 or sagittal MPRAGE images and on the axial images along the anterior, posterior, right lateral and left lateral sides of the distal medulla and cerebellar tonsils at the craniocervical (CC) junction. If the CSF was reduced subjectively circumferentially around the distal medulla and cerebellar tonsils, and no frank point of contact was seen along these four sides, the crowding at the CC junction was termed mild. If there was loss of CSF and contact with the walls of the foramen magnum along one or two of these sides e.g. along the anterior face of the medulla and the right lateral aspect of the right tonsil, the CSF reduction was termed moderate. If there was loss of CSF signal and contact along three or four of the sides, the reduction was termed severe. Deformity of the tonsils or medulla was also noted. The sagittal images and axial T2 weighted images were evaluated for presence of a syrinx.

3. Case presentations

3.1. Case 1

An ex-35 week 5-year-old female with a history of asthma presented with 3.5-years of progressive coughing with oral intake. At 18 months she developed intermittent coughing with feeds and concurrently developed drop attacks characterized falling anteriorly without bracing, intermittent poor balance, and headaches with light sensitivity. Initial neurologic workup, EEG, and Holter monitor were unrevealing. Flexible fiberoptic scope examination revealed a normal supraglottis and symmetric vocal fold motion. VFSS demonstrated penetration into the laryngeal vestibule and silent aspiration during the pharyngeal phase that improved with thickening. Given the patient's history of other neurologic type symptoms, an MRI was performed that demonstrated a CMI with a 10 mm herniation of the cerebellar tonsils through the foramen magnum, moderate restriction of CSF flow at the foramen magnum, and no syrinx (Fig. 1, Table 1). She was referred to neurosurgery and underwent posterior fossa decompression. On follow up, she had good coordination and normal gait. Repeat VFSS 12 weeks post-operatively revealed improved pharyngeal swallow function with mild penetration only of ultra thin liquids without aspiration.

3.2. Case 2

An ex-35 week female with a history of asthma, GERD, and multiple hospitalizations for croup presented at age 4 years with choking on liquids and dry foods. Mom noted that at age 3 years, she began experiencing projectile vomiting, weekly headaches, poor sleep, snoring, and apneic episodes for which she underwent adenotonsillectomy for presumed obstructive sleep apnea. Flexible fiberoptic examination revealed a paretic right vocal fold, direct laryngoscopy and bronchoscopy were normal, and she had normal swallow function on VFSS; however, the study was limited by only capturing single swallows. An MRI was performed for work-up of her vocal fold paralysis, revealing a CMI with 5 mm descent, moderate CSF flow restriction, and no syrinx (Fig. 2, Table 1). The patient was referred to neurosurgery and surgical decompression was performed. One month post-operatively, the patient was noted to have significant improvement in vomiting, snoring, and quality of sleep. At four months post-operatively, coughing, vomiting, and headaches had resolved and repeat fiberoptic examination revealed full vocal fold mobility. The patient's mother declined post-operative VFSS given symptom resolution.

3.3. Case 3

An ex-32 week male presented at 1 year of age with a history of laryngomalacia, severe reflux, intermittent stridor, and coughing/choking with liquid intake. A VFSS demonstrated deep penetration

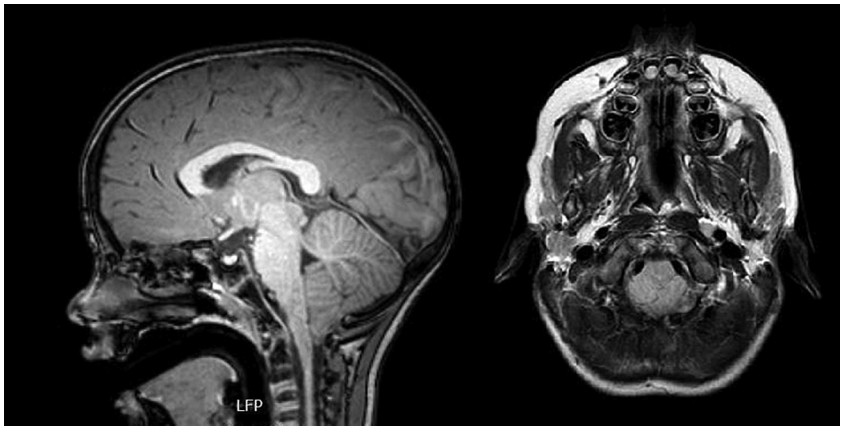

Fig. 1. MRI images of Case 1. Left: Sagittal T1 MRI demonstrating descent of the cerebellar tonsils through the foramen magnum. Right: T2 axial image through the foramen magnum demonstrating restriction of CSF flow.