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Postcricoid vascular abnormalities: Hemangiomas, venous malformations, or anatomic variant☆

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ABSTRACT

Objective: Ten children with pressure-dependent postcricoid masses (PDPCM) previously referred to in the literature as hemangiomas or vascular malformations are presented. We propose these lesions represent an anatomic variant. We review previously reported cases, and report the presentation, diagnosis, and management of the patients in our series, the largest series to date.
Methods: Ten patients, aged five weeks to nine months, were diagnosed, and treated or observed. Of the patients undergoing intervention, one was treated with a gastrostomy tube, fundoplication, and Propranolol therapy; and the other with CHARGE association underwent a tracheotomy.
Results: No PDPCMs demonstrated significant interval change in size or appearance, and eight of ten patients did well with observation.
Conclusion: Based on current information, the majority of PDPCMs likely represent an anatomic variant rather than a hemangioma or vascular malformation. Diagnosis is most readily made with awake flexible fiberoptic laryngoscopy. Because the incidence of synchronous airway pathology is high, direct laryngoscopy and bronchoscopy without routine biopsy is recommended for symptomatic patients. Imaging should be individualized and may be helpful for ambiguous cases. Although numerous treatment modalities have been advocated based on the presumptive diagnosis of a hemangioma, treatment of PDPCMs is not necessary in the majority of cases, as most patients may be safely observed.

1. Introduction

The occurrence of a pressure-dependent vascular-appearing mass in the postcricoid region of a child's hypopharynx is uncommon. To date, 30 cases have been reported in nine publications. The prevailing belief has been that most cases represent hemangiomas, yet the collective evidence of such is weak. More recent reports argue in favor of an alternative diagnosis, termed "postcricoid vascular lesions," specifically venous malformations. Given the unique pressure-dependent characteristics of postcricoid vascular-appearing masses and their increasing frequency of diagnosis, it has long been our conviction that these masses represent an anatomic variant of a regional vascular plexus. We report ten additional patients, the largest series to date, and critically review the literature discussing presentation, diagnosis, evaluation, and management.

2. Report of cases

Over a six year period (2005–2011), ten patients presenting to the Pediatric Otolaryngology Department at Geisinger Medical Center were diagnosed with a vascular-appearing pressure-dependent postcricoid masses (PDPCM), referred to in other publications as a postcricoid hemangioma or "vascular lesion." Patients were between the ages of five weeks and nine months at presentation, with an average age of five months. Four patients were male and six female. Two patients were evaluated by outside otolaryngologists who failed to make the diagnosis. Presenting symptoms included feeding difficulties, stridor, recurrent cyanosis, reflux, snorting, cough, nasal congestion, sleep apnea, and snoring. After evaluation and treatment of concurrent medical conditions, six patients were found to have no symptoms directly related to their postcricoid finding, but instead attributable to co-morbid laryngomalacia and GERD. The PDPCM was felt to be an incidental finding. This is supported by resolution of laryngomalacia and GERD and their associated symptoms over time, despite persistence of the postcricoid abnormality. Of the remaining three patients, symptoms felt to be directly related included feeding difficulties and intermittent stridor. Patients with more severe symptoms had multiple other contributing medical issues, including one with CHARGE association.

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Table 1
Patient summary.

Age at presentation/gender	Symptoms	Comorbidity	Treatment/evaluation	Follow-up
9 mo/M	Obstructive sleep apnea Reflux	Adenotonsillar hypertrophy GERD	Sleep study & pH probe Adenotonsillectomy Observation	10 mo
7 mo/M	Obstructive sleep apnea Cyanotic events	CHARGE syndrome w/ multiple congenital anomalies	Tracheotomy	2 mo
7 mo/F	Feeding difficulty Intermittent inspiratory stridor Recurrent apnea	Laryngomalacia GERD	Sleep study & pH probe Fundoplication Gastrostomy tube Propranolol therapy	1 yr
7 mo/F	Intermittent inspiratory stridor Snoring	Laryngomalacia Chronic adenoiditis	Adenoidectomy Observation	7 mo
6 mo/M	Nasal congestion	Adenoid hypertrophy	Observation	4 yr, 1 mo
5 mo/M	Feeding difficulty Stridor	Laryngomalacia	Observation	6 mo (telephone)
4 mo/F	Intermittent inspiratory stridor	Laryngomalacia	Observation	2 mo
2 mo/F	Stridor Snoring	Laryngomalacia	Pneumogram & pH probe Observation	1 yr
6 wk/F	Stridor	Neglect w/ failure to thrive Laryngomalacia	Pneumogram & pH probe	2 mo
5 wk/F	Stridor Sleep apnea Nasal congestion	Neonatal rhinitis Laryngomalacia	Observation	1 mo

M, male; F, female; GERD, gastroesophageal reflux.

After diagnosis by awake fiberoptic laryngoscopy, nine patients in our series also underwent videostroboscopy. Six of the ten patients underwent examination of their airway under anesthesia. In half of these patients, the fullness could not be visualized under general anesthesia, while in the other half the fullness was visualized but was significantly decompressed. Three of these six patients also underwent esophagoscopy with normal findings.

Follow-up ranged from one month to four years, with all but one patient undergoing serial flexible laryngoscopies to assess for interval changes. One patient's family did not keep their appointments, and follow-up in this case was done via telephone interview.

Of the patients in our series, one patient with mild symptoms required only dietary modification. Two patients with more severe symptoms and multiple other medical problems, required in one case, a gastrostomy tube with fundoplication; and in the other child with CHARGE association, a gastrostomy tube and a tracheotomy. The first of these two patients was additionally treated with Propranolol. One of the patients with severe symptoms (the child without CHARGE association), who presented early in the series, was treated with Propranolol. The remaining patients were all successfully observed. A summary of the age at presentation, symptoms, treatment, and follow-up is presented in Table 1.

3. Literature review

Vascular-appearing postcricoid masses are a unique and only recently-reported entity. Review of the literature yielded nine reports in which a total of 30 patients were presented; 40 patients, including our series [1–9]. In six reports, the finding was felt to represent a hemangioma, and three felt they were a vascular lesion, more specifically a venous malformation.

Examination characteristics are distinctive but easily missed, and this may in part explain why these anomalies have only recently been recognized. PDPCMs demonstrate pressure-dependent behavior (Fig. 1). At rest, these masses are decompressed and generally not visible, but with crying, or a Valsalva maneuver, they distend or "balloon" and are more readily identified. Characteristically, under general anesthesia, PDPCMs will be less conspicuous or inapparent. Trendelenburg positioning can at times be helpful. They are more easily identified on fiberoptic laryngoscopy with the child agitated and crying. Awareness of this unique pressure-dependent behavior is critical to making the diagnosis.

In the literature, of the 30 reported cases, 20% of patients were examined by an otolaryngologist who failed to identify the postcricoid fullness before diagnosis by a second otolaryngologist

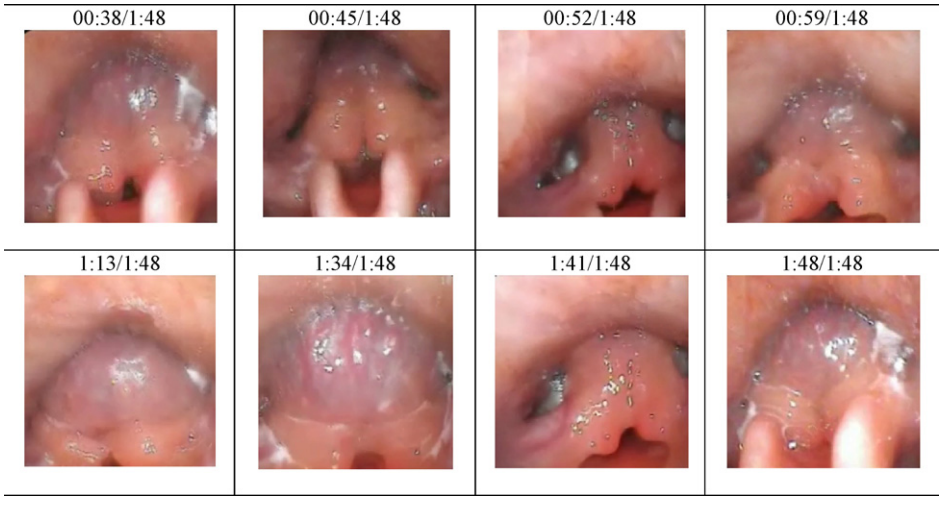

Fig. 1. One minute, 48 second video stroboscopy snapshots of PDPCM displaying resting state and "ballooning" from effects of the Valsalva maneuver induced by crying.