An Unusual Cause of Pediatric Dysphagia: Bronchogenic Cyst

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Bronchogenic cysts (BCs) are bronchopulmonary foregut malformation, mostly located in the middle and superior mediastinum. BCs result from abnormal budding of the primitive tracheobronchial tube. The location of the cyst depends on the embryological stage of abnormal budding. Although periesophageal BCs have been frequently reported in adults, a completely intramural cyst is very rare, with about 22 adult cases reported. Approximately 40% of these adults presented with chest pain, cough, or dyspnea, and 14% with dysphagia.[1] There were no pediatric cases of an intramural esophageal cyst presenting with dysphagia in the English literature on PubMed search from 1960 to 2015.

Case Presentation

A 12-month-old boy presented with failure to thrive since birth, with a decrease in weight from the 75th to the 5th percentile at time of presentation. Beginning at age 9 months, he began having frequent episodes of vomiting with solid foods. He was started on a histamine H2-receptor antagonist without significant improvement. Past medical history was significant for vesicoureteral reflux treated with nitrofurantoin prophylaxis but was negative for recurrent pneumonia and chronic lung disease. Review of systems was unremarkable except for refusing to accept solid foods, and physical exam revealed no unusual findings. Based on this presentation, further imaging was undertaken to rule out anatomical abnormalities as the cause for his presentation.

An upper gastrointestinal series (Figure 1) revealed a fixed defect along the left lateral aspect of the distal esophagus. Computed tomography (Figures 2 and 3) with contrast uncovered an ovoid, fluid attenuated, middle mediastinal structure that was adjacent to the distal esophagus. This was compatible with an esophageal duplication given the proximity to the esophagus, although the differential also included BC. Thoracoscopic resection revealed a cystic mass of the esophagus that was intramural with mucoid material contents. Though the cystic mass appeared intramural, it was completely excised without damaging the native esophagus. Postoperatively the patient did well and tolerated fluids and solids without any vomiting or aversions. Several months later, the family reported that the patient continued to do well with no feeding issues.

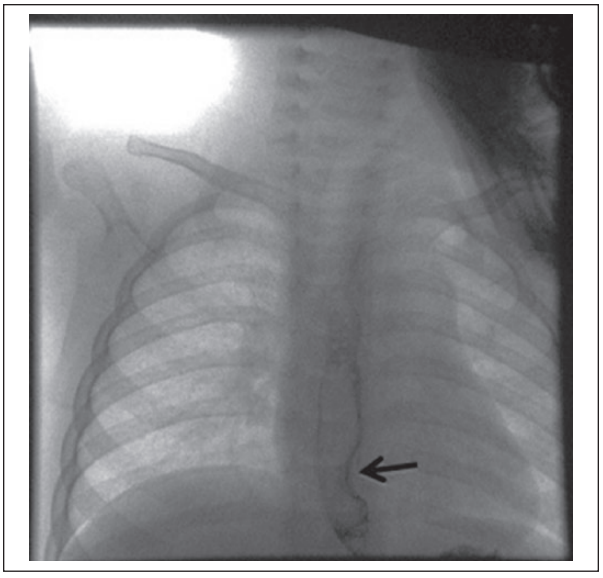

Figure 1. Barium upper gastrointestinal study showing mass effect on left lateral aspect of esophagus (arrow).

On histological exam the cyst was lined with ciliated pseudostratified columnar epithelium overlying a layer of discontinuous smooth muscle and an outer wall composed of fibrovascular connective tissue with islands of hyaline cartilage. Mucous glands were noted in this layer (Figures 4 and 5). These findings were consistent with a BC.

Discussion

Commonly found in children, dysphagia, also known as swallowing dysfunction, is observed in 25% to 30% of

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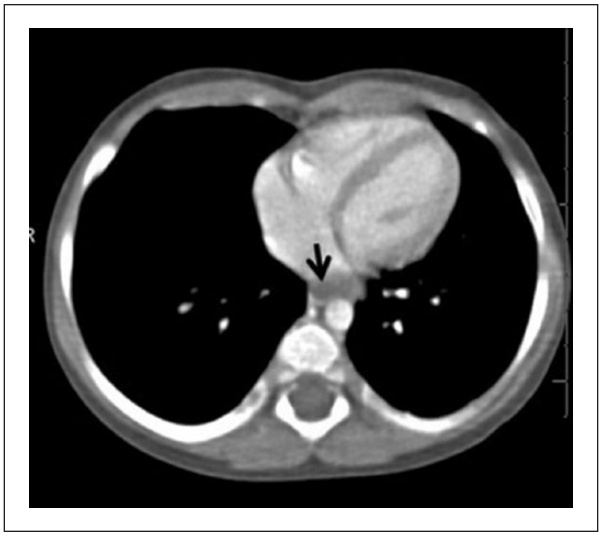

Figure 2. Axial computed tomography scan after intravenous contrast shows low attenuation mass in the posterior mediastinum (arrow), anterior to the aorta, inseparable from the esophagus.

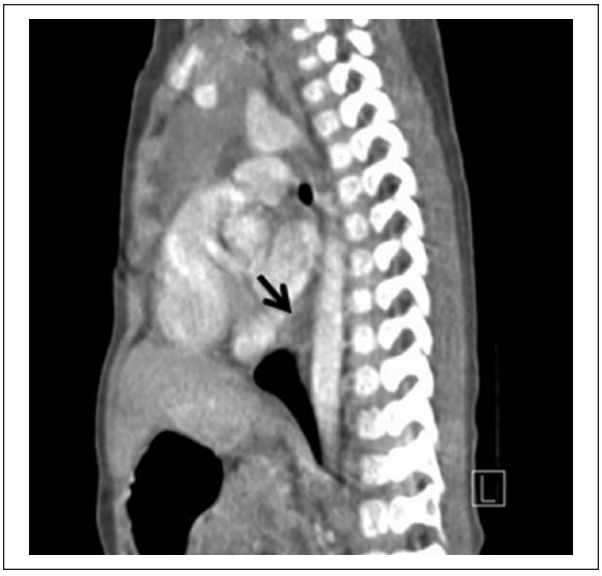

Figure 3. Sagittal reconstructed image from the computed tomography scan showing cystic mass in the posterior mediastinum (arrow).

normal children who present with minor feeding issues. In children with neurologic, respiratory, or anatomic abnormalities, severe feeding difficulties are observed in 40% to 70% of children.[2,3] Gastroesophageal reflux is commonly found in infants, but it is a relatively uncommon cause of long-term feeding problems with resulting failure to thrive.[4] As such, other causes for the

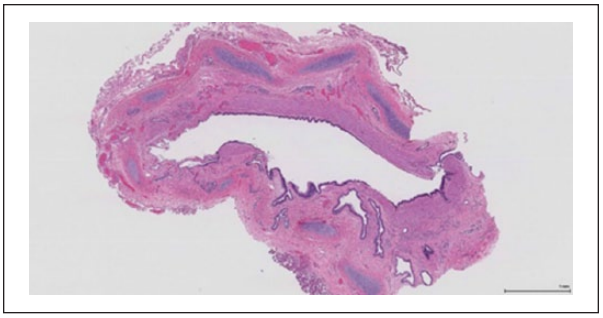

Figure 4. Low-power (2× objective) view of a cross-section of the cyst showing the discontinuous submucosal smooth muscle (star), and cartilage plates (arrowheads) admixed with glands (arrows) in a fibrovascular outer wall.

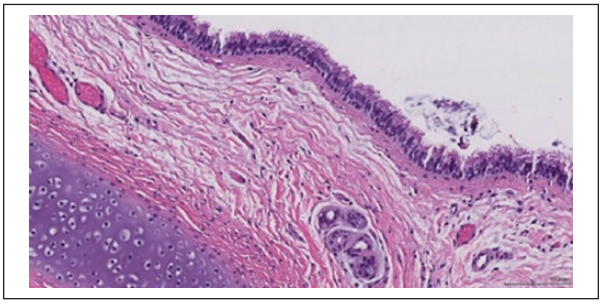

Figure 5. High-power (20× objective) view demonstrating the ciliated pseudostratified columnar epithelium.

presentation of dysphagia should always be considered, including anatomical abnormalities of the oropharynx, larynx, trachea, and esophagus. Anatomical variations that can lead to dysphagia secondary to external compressions include BCs and esophageal duplication cysts.[5]

BCs are included in the spectrum of bronchopulmonary foregut malformations along with enteric cysts and esophageal duplications. The conducting airways of the respiratory system and the esophagus begin to differentiate from the primitive foregut at about 3 weeks of gestation when the laryngotracheal groove forms in the anterior medial foregut. Over the following week the respiratory system develops as a diverticulum caudal to the pharyngeal pouch creating the primitive lung bud, which further divides creating the primitive left and right bronchi. At the same time, septa from the posterior lateral laryngotracheal groove form and unite dividing the foregut into a dorsal and ventral tube, the esophagus, and conducting airways, respectively.[6,7]

BCs are believed to arise from supernumerary lung budding during this early embryonic stage, which subsequently loses communication with the conducting

airways prior to 16 weeks of gestation. The BCs can be mediastinal (66%) or intrapulmonary (33%), with the location dependent on the timing of this separation during gestation.[5,6] Esophageal BCs are rare in comparison but can develop with abnormal budding along the basal aspect of the laryngotracheal groove, somehow becoming embedded in the esophagus.[8]

BCs are classified by location and can be paratracheal, hilar, carineal, pericardic, or paraesophagic. More often than not, they are found on the right side of the mediastinum. As such, a cyst found intra-esophageal could be mistaken for an esophageal duplication cyst, which in contrast always sits adjacent to the esophageal wall.[9] The majority are located in the right thoracic and lower segment of the esophagus. Esophageal duplication cysts are composed of nonkeratinzed squamous or ciliated columnar epithelium as a result of persistence of embryonic epithelium of the foregut[10] with or without muscular layers, which can make distinction from a BC challenging.[11] Therefore, the differentiation of an esophageal duplication cyst from an esophageal BC relies on both surgical and histologic findings.[8]

Most BCs are thought to be asymptomatic and free of complications unless they become infected or are large enough to cause pressure on contiguous vital structures. The most common presenting symptom is chest pain followed by cough and dyspnea. Other symptoms can include heartburn, anorexia, weight loss, respiratory distress, and hemoptysis. For esophageal BCs in the mid or lower esophagus, the most common presentation is dysphagia.[5,8,12]

Diagnosis of BC can be made with chest radiograph showing a well-defined, noncalcified mass, with water density or air space cysts with air-fluid levels. While radiography has a high sensitivity for detecting this lesion (>90%), the specificity is very low (20% to 70%).[8] A barium esophagogram may show external compression of the esophagus.[13] For further evaluation, computed tomography scan is an excellent diagnostic tool because it is noninvasive and demonstrates the exact location of the mass, its cystic nature, and its relationship with adjacent structures. BCs typically appear as well-defined, thin-walled nonenhancing homogeneous cystic masses with water density; if the cyst is infected or high in protein and calcium content, its density may fall into the solid tissue range, which can increase diagnostic uncertainty. In those cases, a magnetic resonance imaging scan is useful because it is sensitive for detecting fluid-filled tissue.[14] Endosonography was reported to be useful in diagnosing a case of intramural esophageal BC, by clearly delineating the submucosal location of the cyst.[15] However, none of these techniques can indicate the nature of the cyst. The most challenging part of arriving at an accurate diagnosis is determining whether the lesions are intramural, within the esophagus, or simply mediastinal, due to the difficulty of precisely defining the outer wall of the esophagus.

Surgical resection of the cyst is the treatment of choice. Thoracotomy and/or laparotomy is usually required. Endoscopic resection of an esophageal BC has been described.[16] Independent of the surgical method offered, the outcomes are extremely satisfactory with no recurrences reported.[8]

Conclusion

Esophageal BCs are rare congenital malformations that can be a cause of food aversions and failure to gain weight in pediatric patients. Although the diagnosis can be suspected noninvasively, definite diagnosis and treatment is done surgically. As our case illustrates, these children have a good prognosis. Although very rare, esophageal BC should be in the differential diagnosis of a child with dysphagia.

Author Contributions

AK: Contributed to conception and design; contributed to acquisition, analysis, and interpretation; drafted manuscript; critically revised manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.
JL: Critically revised manuscript; gave final approval.

Declaration of Conflicting Interests

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References

1. Turkyilmaz A, Eroglu A, Subasi M, Findi G. Intramural esophageal bronchogenic cyst: a review of the literature. *Dis Esophagus*. 2007;20:461-465.
2. Hyman SL, Porter CA, Page TJ, Iwata BA, Kissel R, Batshaw ML. Behavior management of feeding disturbances in urea cycle and organic acid disorders. *J Pediatr*. 1987;111:558-562.
3. Rudolph CD, Link DT. Feeding disorders in children. *J Pediatr Gastroenterol Nutr*. 2002;49:97-112.
4. Rudolph CD, Mazur LJ, Werlin SL, et al. Guidelines for evaluation and treatment of gastroesophageal reflux in infants and children. *J Pediatr Gastroenterol Nutr*. 2001;32(suppl 2):S1-S31.

5. St-Georges R, Deslauriers J, Duranceau A, et al. Clinical spectrum of bronchogenic cysts of the mediastinum and lung in the adult. *Ann Thorac Surg*. 1991;52:6-13.
6. Kravitz RM. Congenital malformations of the lung. *Pediatr Clin North Am*. 1994;41:453-472.
7. Maier HC. Bronchogenic cysts of the mediastinum. *Ann Surg*. 1948;127:476-502.
8. Ko SF, Hsieh MJ, Lin JW, et al. Bronchogenic cyst of the esophagus: clinical and imaging features of seven cases. *Clin Imaging*. 2006;30:309-314.
9. Nazem M, Amouee AB, Eidy M, Khan IA, Javed HA. Duplication of cervical esophagus: a case report and review of literature. *Afr J Paediatr Surg*. 2010;7:203-205.
10. Langston C. New concepts in the pathology of congenital lung malformations. *Semin Pediatr Surg*. 2003;12:17-37.
11. Briganti V, Molle P, Miele V, Vallasciani S, Calisti A. Intramural esophageal bronchogenic cyst: an unusual cause of dysphagia in pediatric patients. Case report [in Spanish]. *Cir Pediatr*. 2003;16:99-101.
12. Eraklis AJ, Griscom NT, McGovern JB. Bronchogenic cysts of the mediastinum in infancy. *N Engl J Med*. 1969;281:1150-1155.
13. Gupta B, Meher R, Raj A, Agarwal S. Duplication cyst of the esophagus: a case report. *J Paediatr Child Health*. 2010;46:134-135.
14. Chafik A, Benjelloun A, Qassif H, El Fikri A, El Barni R, Zrara I. Intramural esophageal bronchogenic cysts. *Asian Cardiovas Thorac Ann*. 2011;19:69-71.
15. Lim LL, Ho KY, Goh PM. Preoperative diagnosis of a paraesophageal bronchogenic cyst using endosonography. *Ann Thorac Surg*. 2002;73:633-635.
16. Sashiyama H, Miyazaki S, Okazaki Y, et al. Esophageal bronchogenic cyst successfully excised by endoscopic mucosal resection. *Gastrointest Endosc*. 2002;56:141-145.