Isolated Cervical Esophageal Duplication Cyst in an Infant with Stridor: A Case Report

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Background

- Cervical esophageal duplication cysts are very rare with fewer than 100 cases reported in the literature
- Arise from the embryonic foregut which gives rise to the bronchopulmonary and alimentary tracts (Figure 1)
- Result of a foregut budding error during 3-6 weeks of gestation
- Can be asymptomatic but often present with compressive symptoms including dysphagia, coughing, dyspnea, respiratory distress, and rarely stridor
- Surgical treatment is recommended for all cases as there are rare instances of malignancy harbored in the cysts. Surgery performed by or in combination with pediatric surgery
- We aim to describe a case of an isolated cervical esophageal duplication cyst presenting with stridor

Methods

- Case report of single patient who presented with stridor and was found to have an isolated esophageal duplication cyst
- This study is presented as a case report and review of the literature

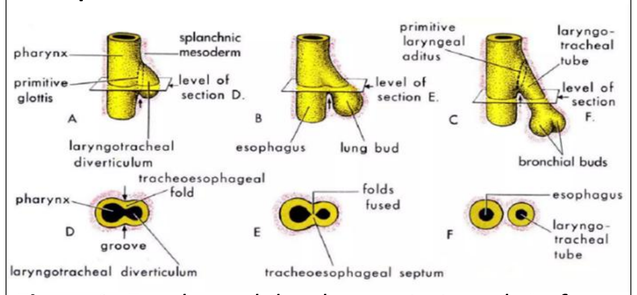

Figure 1. *Esophageal development at week 4 of gestation.* Diagram from The Developing Human: Clinically Oriented Embryology, 5th ed.

Case Report

- 5-month-old female presenting with 3-4 months of stridor and retractions
- Healthy with appropriate weight gain and no difficulties feeding
- Trial of steroids, antibiotics, and albuterol without improvement

Case Report Continued

- At initial presentation, her exam was significant for loud biphasic stridor and flexible laryngoscopy with normal vocal fold mobility, no laryngomalacia, and possible subglottic narrowing
- Initial XR tracheal deviation but no obvious findings so patient was taken to the OR for direct laryngoscopy

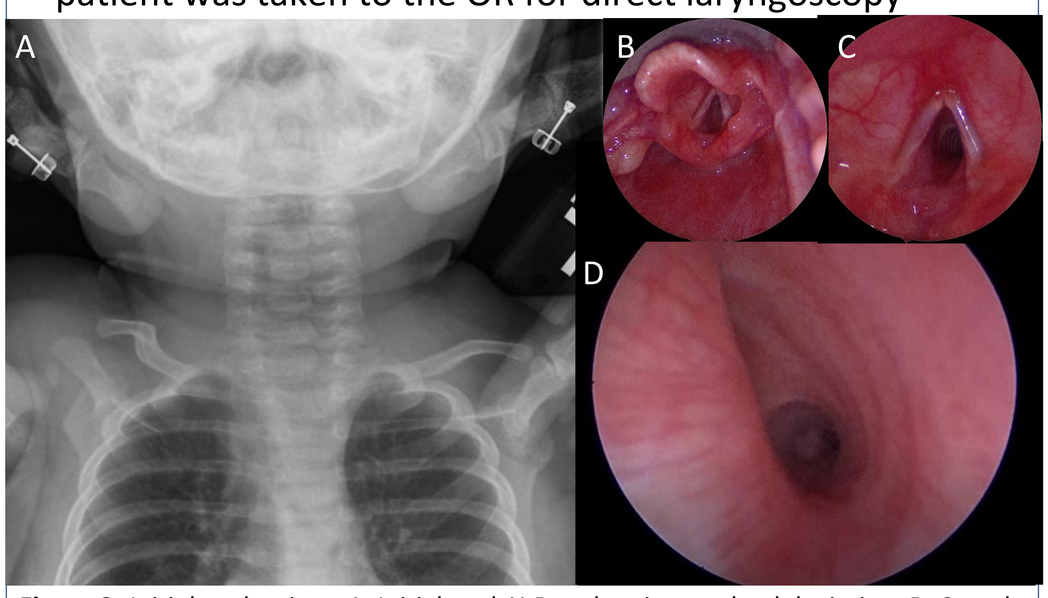

Figure 2. *Initial evaluations.* A. Initial neck X-Ray showing tracheal deviation. B, C, and D. Initial direct laryngoscopy. B. Grade I view with no evident abnormalities. C. No evident subglottic narrowing. D. Carina with left posterior incursion of tracheal wall

- Given OR findings, CT and MRI were ordered

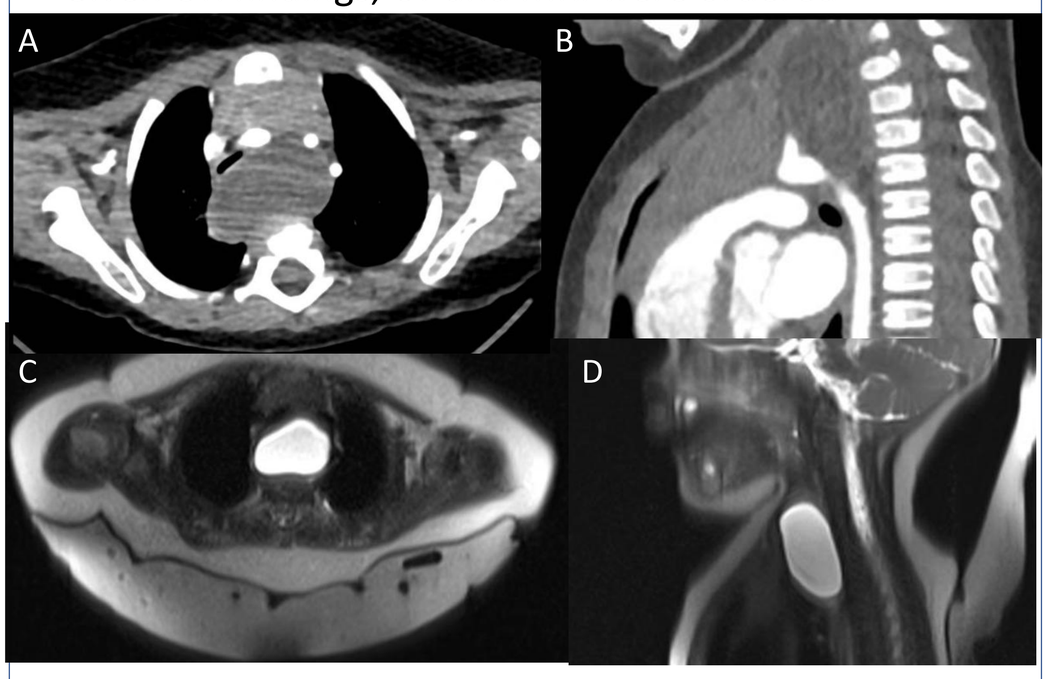

Figure 3. *CT scan and MRI obtained after direct laryngoscopy revealing large mediastinal cystic mass.* A,B. CTA axial and sagittal images showing mediastinal mass with tracheal compression C,D. MRI axial and sagittal images showing mediastinal cystic structure

- Imaging findings were consistent with large foregut duplication cyst so plan was made for trans-cervical resection with pediatric surgery

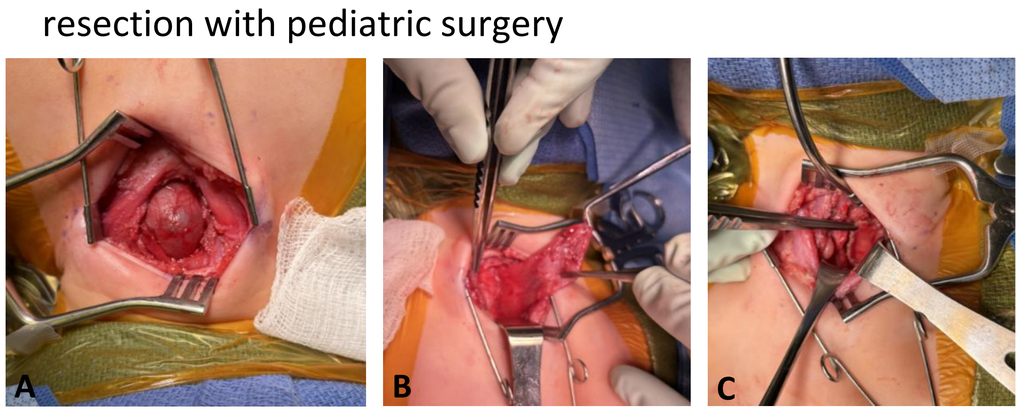

Figure 4. *Intra-operative photos of esophageal duplication cyst.* A. Duplication cyst through trans-cervical incision. B. Retraction of cyst. C. Approximately 2x1 cm cystic wall left in continuity with esophagus

- Operative findings of 8cm cystic mass with thick shared medial wall with the esophagus. Vagus nerve preserved, recurrent laryngeal nerve visualized above and below cyst
- Patient resumed a normal diet the day of surgery, had left true vocal fold paresis. Stridor was resolved and she discharged on postoperative day 2
- At 6-weeks post-operatively, vocal cord mobility had recovered. No signs of recurrence at 6 months

Discussion

- Here we present a case of a rare cervical esophageal duplication cyst presenting with isolated stridor
- In the literature, chest X-ray commonly demonstrates mediastinal mass, tracheal compression, and tracheal deviation. Our case did show tracheal deviation.
- Direct laryngoscopy, like in our case can look similar to tracheomalacia with signs of tracheal compression which warrants more advanced imaging
- Early surgical intervention is recommended but must be wary of recurrent laryngeal nerve or esophageal injuries, in our case cyst wall left to preserve esophageal integrity
- Multidisciplinary collaboration was key to successful treatment of this cyst which extends into chest
- Ultimately, direct laryngoscopy and tracheobronchoscopy should be involved in the workup of infants with noisy breathing of unknown etiology

Conclusion

- This case supports the importance of intra-operative examination of the entire airway for work up of stridor as well as the utility of additional consulting services in managing complex patients.
- Esophageal duplication cysts are a rare cause of isolated stridor that can be successfully surgically treated once identified

References

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