Ectopic Thymic Cyst of the Subglottis: Considerations for Diagnosis and Management

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ABSTRACT

In this report, we review the presentation, management and clinical course of an infant who presented with a subglottic mass that was histologically confirmed as a thymic cyst. This is the third described case of an ectopic thymic cyst presenting as a subglottic mass. The differential diagnosis of subglottic masses in neonates consists primarily of subglottic hemangioma and mucous retention cysts. Otolaryngologists must be prepared for unexpected findings when dealing with critical airways. We compare the presentation and management of our patient with the two previously described cases and propose an embryologic theory for the origin of these rare lesions. An ectopic thymic cyst is a rare and unexpected cause of neonatal stridor. Management of pediatric airway lesions must allow for unexpected findings at the time of diagnostic and therapeutic endoscopy. The appropriate management of subglottic thymic cysts is poorly defined but close surveillance for recurrence is mandatory.

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CASE REPORT

- A 4 month-old male presented with biphasic stridor and retractions since birth.
- Previously admitted to outside institution for croup with improvement after medical management.
- Examination revealed a healthy-appearing Caucasian infant with audible biphasic stridor, increased work of breathing, subcostal and suprasternal retractions. The stridor was most prominent when auscultated over the larynx.
- A flexible fiberoptic laryngoscopy revealed a subglottic mass inferior to the right vocal cord effacing 50% of the airway.
- The patient was admitted and taken urgently to the operating room for direct laryngoscopy and bronchoscopy.

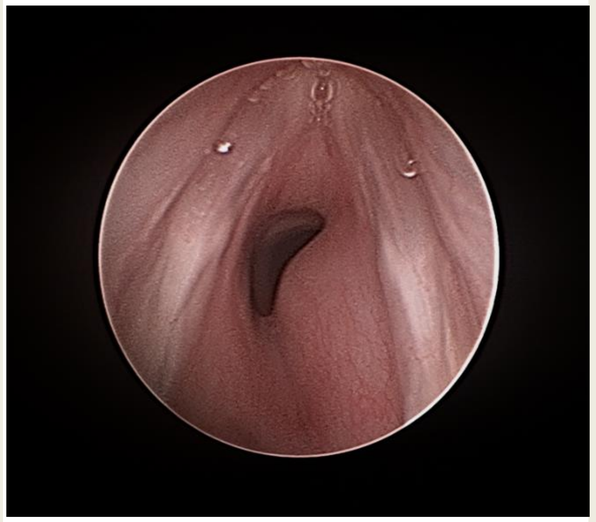

Figure 1: Subglottic Mass

- In the operating room, the mass was visualized with the 4mm Hopkins rod telescope (Fig 1) under spontaneous ventilation.
- The lesion was cystic with viscous fluid noted on entry into the cyst
- Marsupialization was performed with microlaryngeal scissors and a microdebrider (Fig 2) after tissue biopsies

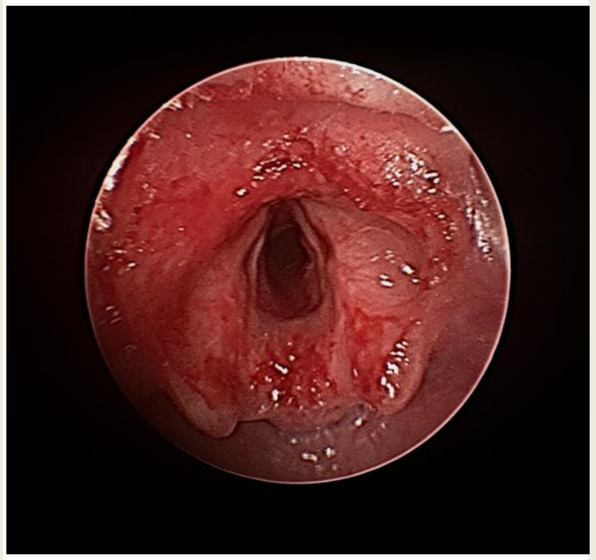

Figure 2: Appearance after marsupialization

RESULTS

- Histopathologic evaluation showed squamous and respiratory epithelium-lined tissue with submucosal nests of lymphocytes in the underlying stroma in which scattered Hassell's corpuscles are found, consistent with ectopic thymic tissue. (Fig 3).

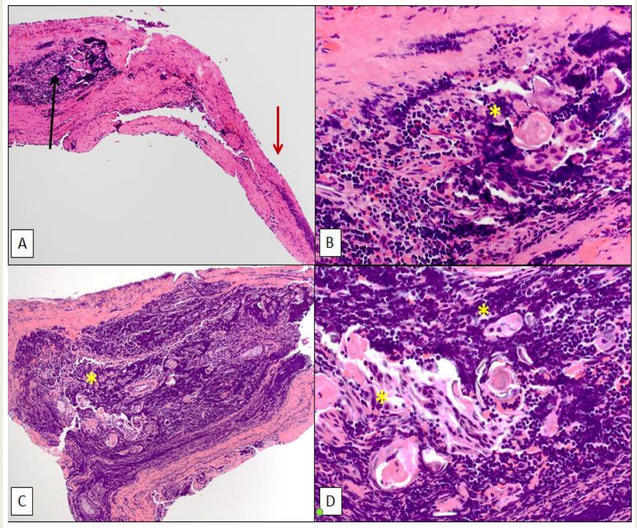

Figure 3: Histopathology: A) Squamous epithelium (red arrow) lined tissue with ectopic thymic tissue (black arrow). H+E x 100. B) Ectopic thymic tissue with Hassel's corpuscles (yellow asterisk) H+E x400. C) Deeper sections reveal a larger focus of ectopic thymic tissue. H+E x 100. D) Multiple Hassel's corpuscles (yellow asterisks B,C) surrounded by lymphoid tissue in deeper section. H+E x 400

- Repeat direct laryngoscopy was performed 1 month later with no recurrent airway lesion noted (Fig 4). Superficial biopsies were negative for thymic tissue.

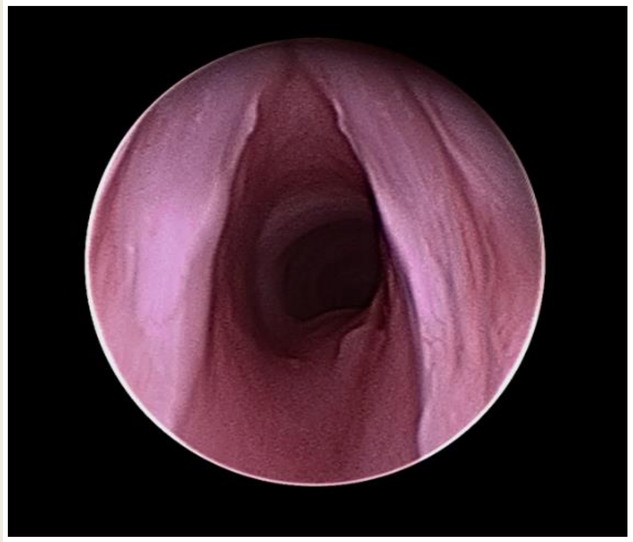

Figure 4: 1 month follow up

- Four weeks later, the child returned to clinic with several days of worsening stridor. Flexible laryngoscopy revealed an irregularity in the subglottis and he was taken to the operating room.
- Intraoperatively, there was a recurrent lesion or edema inferior to the right true vocal fold (Fig 5). The CO_2 laser was used to ablate this area after biopsies were performed. There was no evidence of thymic tissue on pathology.
- At follow up 1 month later the child was doing well
- Repeat laryngoscopy demonstrated well healed, patent airway and no evidence of recurrence (Fig 5). He has continued to be asymptomatic since that time.

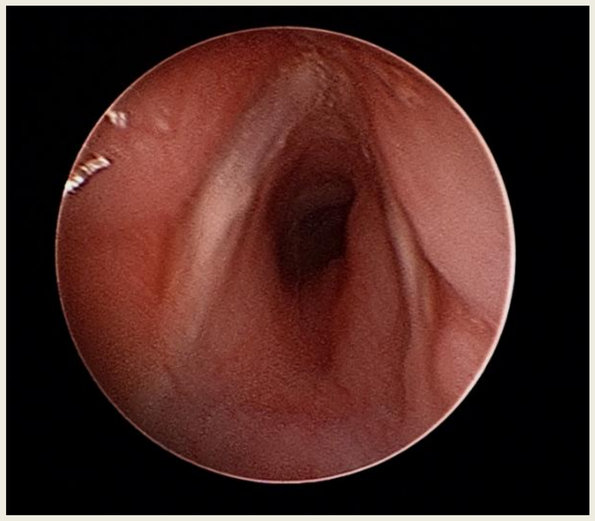

Figure 5: Recurrent lesion vs edema

DISCUSSION

- Ectopic thymic tissue is reported as a rare cervical lesion, although is present in up to 1% of patients in autopsy series.[4] It can present as a cervical thymic cyst, ectopic thymic tissue or a cervical thymoma.[5]
- Cervical thymic cysts can be challenging to diagnose and are difficult to distinguish from other cysts.[5] Histopathologic analysis is necessary for definitive diagnosis.
- Mechanistic theories include an arrest in the descent during the 6th to 8th gestational weeks or as a result of an abnormal sequestration model.[7,8]
- The thymic primordia descends into the mediastinum during the 8th gestational week.
- Unilateral disruption of thymic descent could explain how ectopic thymic tissue is trapped within the endodermal component of the larynx.
- Ectopic thymic tissue has been reported to enlarge after a respiratory tract infection.[7]
- In our case, after initial marsupialization a recurrent mass lesion was noted. Repeat biopsies failed to reveal recurrent thymic tissue.
- Scarring and airway remodeling may be the cause for the morphological abnormality seen on the third laryngoscopy.
- Tissue biopsies should be performed in order to establish a histopathologic diagnosis and guide subsequent management.
- Our management with follow up tissue examination suggests that marsupialization may be adequate for management, as opposed to complete excision.

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