Congenital laryngeal saccular cyst: Report of a case in an infant

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Abstract
Respiratory obstruction and stridor in infants and children are not uncommon. A rare cause of these life-threatening symptoms is congenital saccular cyst. An accurate diagnosis of saccular cyst can be made by eliciting a good history, by endoscopic visualization of the lesion, and by computed tomography. Endoscopic excision is the preferred treatment for infants, whereas an external approach is reserved for older children. It can be difficult for anesthetists to intubate infants when the anatomy of the larynx is distorted, but the choice of tracheotomy for an infant has many drawbacks as well. We discuss the clinical presentation and management of a 3-month-old boy who was brought to us with a congenital laryngeal saccular cyst.

Introduction
Congenital saccular cysts of the larynx are rare. When they do occur, they usually present as a respiratory obstruction in infants and children. Symptoms of a saccular cyst in a newborn are nonspecific and common to other causes of laryngeal obstruction. Early recognition and treatment of these disorders is important because of the high mortality associated with undiagnosed conditions. In this article, we describe a case of a symptomatic saccular cyst in an infant who was successfully treated with microlaryngeal surgery.

Case report
A 3-month-old boy was brought to us for evaluation of a history of noisy breathing and feeding difficulty that had been manifest since 4 days following his birth. He had been referred to our hospital with a diagnosis of a larynx-compressing mass that had been detected on computed tomography (CT).

Our examination revealed that the child had inspiratory stridor. The accessory muscles of respiration were functioning, and the intercostal, suprasternal, and substernal area was recessed (figure 1). Fiberoptic endoscopy detected a cystic swelling in the supraglottic area that involved the left aryepiglottic fold. The epiglottis was curled, and the true vocal folds were not visible. The larynx was tilted and pushed to the opposite side. A second CT confirmed the cystic mass in the left supraglottic area (figure 2).

At this stage, a diagnosis of a saccular cyst was made. The case was reviewed with a pediatrician, and other than the laryngeal stridor, no other congenital anomaly was noted. The infant was scheduled for microlaryngeal surgery. Surgery via an external approach was reserved as an alternative if deemed necessary. The case was then discussed with an anesthesiologist, who anticipated a difficult endotracheal intubation. Nevertheless, the surgical team's consensus was to attempt an intubation and to be prepared to switch to tracheotomy if necessary. The patient was induced with 2.5% halothane. As anticipated, the distortion of the laryngeal anatomy made the intubation difficult (the laryngeal inlet could not be visualized) but not impossible. A 2.5-mm uncuffed tube was successfully introduced along the epiglottis where air bubbles were seen, indicating the airway passage. The patient was maintained with a mixture of nitrous oxide and oxygen in a ratio of 2.5:1.5 L and with halothane 0.5 to 1%.

An operating microscope with a 400-mm lens was used to visualize the cyst in the left aryepiglottic fold (figure 3, A). The cyst extended superiorly up to the tip of the epiglottis, medially overlapping the glottic chink, and laterally filling the left piriform fossa. The lower extent of the cyst could not be discerned. A cup forceps was used to hold the roof of the cyst, and a sickle knife was used to incise it. When the cyst was punctured, a whitish gelatinous fluid was released and the cyst walls collapsed (figure 3, B). The larynx immediately reverted back to its normal position. Both vocal folds were visualized and found to be normal. The entire sac was excised, and the apex of the piriform fossa was found to be normal. Bleeding from the excision site was minimal. The child was extubated in the operating room.

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and observed in the intensive care unit for 24 hours.

Histopathology revealed that the cystic tissue was lined with ciliated, pseudostratified columnar epithelium. The subepithelial tissues contained a mild chronic inflammatory cell infiltrate and lobules of salivary gland embedded in the fibrocollagenous tissue. These histologic features confirmed the diagnosis of a saccular cyst.

The patient was kept on intravenous antibiotics and steroids. Feeding was resumed 6 hours postoperatively, and the infant did not develop any stridor. He was discharged on postoperative day 3. A follow-up examination 1 month later revealed normal laryngeal structure, vocalization, and vocal fold mobility.

Discussion
Pathogenesis. A laryngeal saccule is a small diverticulum arising out of the laryngeal ventricle. It extends upward between the false vocal fold, the base of the epiglottis, and the thyroid cartilage. It contains mucous glands, and it secretes mucus through an orifice in the anterior part of the roof of the ventricle. The stored mucus probably lubricates the surface of the vocal folds.2 A congenital saccular cyst is believed to form as a result of a developmental failure to maintain patency of the saccular orifice.3

DeSanto et al differentiated saccular cysts from laryngoceles on the basis of communication with the laryngeal lumen.4 In a laryngcele, the orifice of the saccele remains patent, and it is distended and filled with air; a saccular cyst is a mucus-filled dilation of the saccele that does not communicate with the laryngeal lumen.

Clinical features. Most saccular cysts arise from a broad base between the aryepiglottic fold and the arytenoids. They extend inferiorly to the ventricle and encroach into the functional airway and/or laterally into the piriform sinus.5

Children with laryngeal cysts typically present with inspiratory stridor. Stridor may be characteristic of a particular pathology, but it is never diagnostic. Stridor at birth is unusual and generally denotes a fixed congenital narrowing, such as a laryngeal web or subglottic stenosis. Dynamic conditions such as laryngomalacia and congenital vocal fold palsy become evident during the first few weeks of life. A gradual increase in the severity of stridor or airway compromise implies growth of an obstruction, such as in the case of a saccular cyst. The initial presentation of a saccular cyst may mimic laryngomalacia. As the cyst enlarges, the stridor may worsen and there may be voice

Figure 1. At the referral examination, the intercostal, suprasternal, and substernal area is recessed.

Figure 2. CT shows the saccular cyst in the left supraglottic area.

Figure 3. Laryngoscopy shows the affected area before (A) and after (B) puncture and collapse of the saccular cyst.
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changes associated with episodes of cyanosis, retractions, and feeding difficulties.

Diagnosis. A diagnosis of saccular cyst is suggested by a soft-tissue lateral neck radiograph that shows a mucus-filled sac. The presence of the cyst can be confirmed by fiberoptic laryngoscopy. Both CT and magnetic resonance imaging can be quite helpful in determining the exact location and extent of the mass.3

Management. The management of saccular cysts in infants and children has been primarily endoscopic. Holinger et al advocated aspiration and marsupialization as the initial treatment.6 However, this method frequently requires multiple procedures. A lateral cervical approach with concomitant tracheotomy has been recommended by a few authors.2,3 Abramson and Zielinski used a CO₂ laser to effectively incise and vaporize the lining of sacular cysts.5

In our patient, stridor began shortly after birth and it gradually increased, leading to severe respiratory distress. The diagnosis was arrived at on the basis of the clinical presentation and findings on endoscopy and CT.

Prior to our patient’s surgery, the question of tracheotomy was debated in view of the anticipated difficulty in securing the airway by endotracheal intubation. The complications of tracheotomy in the pediatric population can be classified as early and late. Early complications include apneic attacks, surgical emphysema, pneumothorax or pneumomediastinum, accidental decannulation, creation of a false passage, obstruction of the tube, hemorrhage, and chest infections. Late complications include difficult decannulation secondary to many factors, including suprasternal collapse, the formation of granulation tissue, the relative size of the airway, and possibly the development of a psychological attachment to the tracheotomy tube. In view of these complications and the patient’s young age (3 mo), we decided to try endotracheal intubation, which proved to be successful.

Complications aside, removal of a saccular cyst must be carried out with the utmost care in a pediatric population. The microlaryngeal surgery and excision of the cyst in our patient proved to be safe and simple, and the outcome was excellent.

References