Saccular cyst in an infant: an unusual cause of life-threatening stridor and its surgical treatment

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Saccular cysts in infants are rare lesions of the larynx and may result in respiratory obstruction and severe dyspnea. Herein we present a case of saccular cyst in a three-month– old infant. She was presented with severe stridor and respiratory distress. She had been followed with the diagnosis of tracheomalacia for three months elsewhere. Direct laryngoscopy of the larynx revealed a saccular cyst. Stridor disappeared shortly after surgical excision of the supraglottic saccular cyst under direct laryngoscopy. No complication or recurrence was seen in the 12-month follow-up period. Saccular cysts in infants are rare lesions and should be kept in mind in the differential diagnosis of stridor in infants.

Key words: saccular cyst, stridor, surgery.

Laryngeal saccular cysts in infants are rare entities. They are located in the saccular appendage. They most commonly arise from between the aryepiglottic fold and the arytenoid cartilage with bulging into the rima glottis medially and pyriform sinus laterally. This location and extension commonly result in airway obstruction and severe stridor, a condition obligating prompt medical intervention[1]. Because of the similar symptoms, differential diagnosis should be made with laryngomalacia, laryngocele, laryngeal web, vocal fold paralysis, subglottic stenosis, tracheomalacia and other obstructive lesions of the upper respiratory tract. Direct laryngoscopy is the best method for the diagnosis of these patients, who are presented with stridor. The patients are at great risk after birth because of airway obstruction. An elective surgery for the excision of the saccular cyst has to be planned shortly after initial endotracheal intubation or tracheotomy.

We present an infant who had been followed with the diagnosis of tracheomalacia elsewhere for three months. She was referred to our department with severe stridor and underwent surgery for the excision of the saccular cyst.

Case Report

A three-month-old female infant was referred to our department with severe stridor and respiratory distress. Her past medical history revealed vaginal delivery of a 2200 g female at the end of 36 weeks of gestation. She had no medical problem except a slight perinatal stridor and had been discharged from the hospital without medical intervention. On the eighth day of age she was admitted to the Department of Pediatrics with shortness of breath and cyanosis while crying. Symptoms of the patient had been attributed to tracheomalacia and she had been placed on follow-up for three months. An endotracheal intubation had been applied for severe stridor with intercostal retractions at the end of three months.

The patient was referred to our department to determine the cause of stridor. There was no sign of infection. Partial oxygen saturation was dropping under 90% shortly after cessation of oxygen support. Preoperative computerized tomography revealed a cystic lesion on the left side of the larynx (Fig. 1). The patient was taken to the operating room to perform direct laryngoscopy under general anesthesia. It revealed a fluid filled cyst arising from the

posterolateral part of aryepiglottic plica on the left side, extending to the left ventricle inferiorly and bulging into the rima glottis (Fig. 2). The airway was significantly narrowed in the glottic region. Most of the cyst wall was taken out under microscopic vision through rigid laryngoscope. Endotracheal tube was removed on the postoperative third day. Histopathologic examination was reported as pseudocyst lined with pseudostratified columnar epithelium. Cystic appearance in the left side of the larynx had disappeared on postoperative computerized tomography (Fig. 3). No complication or recurrence was observed in the 12-month follow-up period.

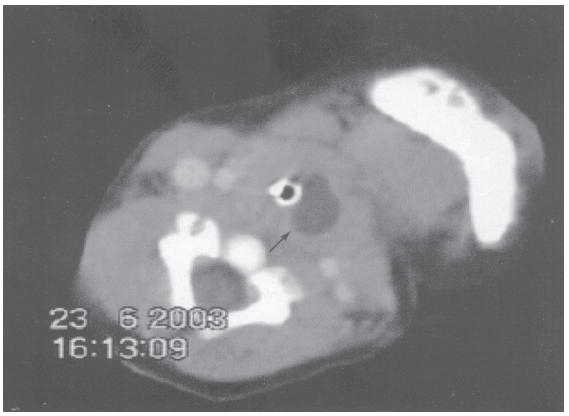

Fig. 1. View of laryngeal cyst on preoperative computerized tomography.

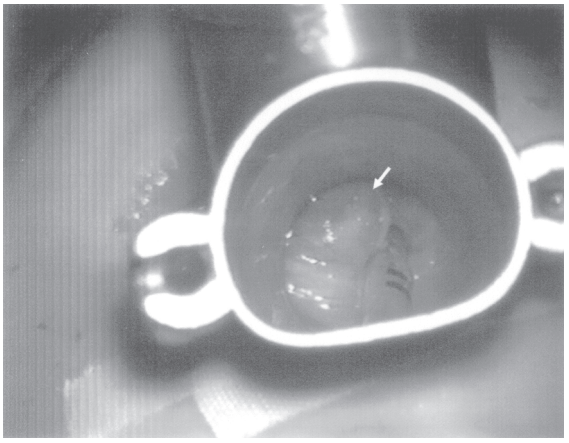

Fig. 2. View of cyst through rigid laryngoscope.

Discussion

Saccular cyst of the larynx is a mucus-filled dilatation of the laryngeal saccule. It may be congenital or acquired. Congenital saccular cyst

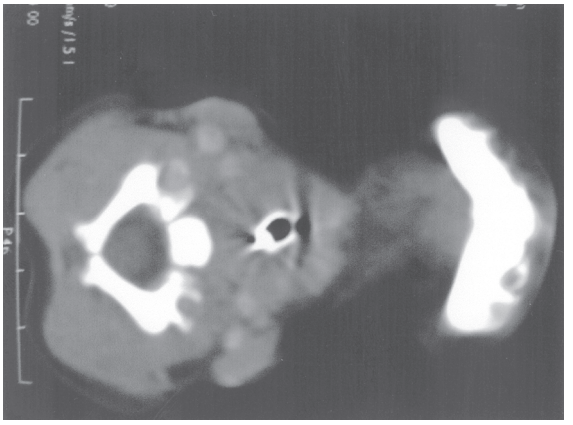

Fig. 3. Cyst appearance had disappeared on postoperative computerized tomography.

may result from atresia of the saccular orifice. Other theories suggest a congenital anomaly associated with embryonic development of the larynx or abnormal migration of the fourth bronchial arch, which may form a sequestrated cyst[1]. Acquired cysts may be due to inflammatory, traumatic or neoplastic obstruction of the saccular orifice. The laryngeal saccule arises vertically from the ventricle and then passes superiorly to the false cords, extends to the base of the epiglottis and inner surface of the thyroid cartilage[1]. As a result of this location, cysts of the saccule may extend laterally or anteriorly. Anterior saccular cysts arise from the anterior ventricle and bulge into the laryngeal lumen between the true and false vocal cords. Lateral saccular cysts extend from false vocal cord to aryepiglottic fold posterosuperiorly and may bulge into the laryngeal lumen and pyriform sinus. The saccular cyst presented in this case was located laterally (Fig. 2).

Age of patient, size of cyst and degree of obstruction of the laryngeal airway delineate the clinical presentation. In infants, the typical clinical signs of saccular cysts are generally noticed shortly after birth[1,2,3]. Stridor during respiration, muffled or weak cry, and respiratory distress during feeding that results in regurgitation and cyanosis are typical findings. Laryngoceles may produce similar symptoms, which are episodic[4]. Adult patients with saccular cysts suffer from hoarseness and airway obstruction.

Direct visualization of the pharynx, hypopharynx, larynx and subglottic area is crucial for the differential diagnosis in infants, who are presented with stridor. Flexible transnasal

endoscopy under local anesthesia may reveal anatomic anomalies and also functional disturbances like vocal fold paralysis and tracheomalacia. However, inadvertent head movements of the infants and abundant secretion in the hypopharynx may not allow sufficient visualization of the field for the exact diagnosis. Direct laryngoscopy under general anesthesia is another alternative for detailed examination of the larynx and subglottic region. Laryngomalacia, laryngocele, laryngeal web, subglottic stenosis and saccular cyst can easily be identified with direct laryngoscopy.

Direct laryngoscopy in infants with a saccular cyst reveals a mass occluding the supraglottic larynx (Fig. 2). Distortion of the aryepiglottic fold, false vocal cord, ventricle and vallecula may also be seen. Aspiration of the cyst yields a thick mucoid material. Lateral radiographs may show a compressed airway but computerized tomography is necessary to determine the detailed location and whole extension.

Endotracheal intubation is generally needed for the emergent treatment of these patients[1,3]. However, if this is unsuccessful, needle aspiration or incision of the cyst may decrease the severity of obstructive symptoms. A tracheotomy may also be needed in some of these cases[1,4,5]. Unnecessary manipulations of the upper airway increase the airway obstruction and should be avoided.

Marsupialization of the lesion may provide permanent relief[6]. Cyst lining can also be vaporized with CO2 laser[1]. Total excision of the cyst is the best choice to prevent recurrences. Smaller anterior saccular cysts can readily be managed endoscopically[4]. View of the cyst can be magnified with operation microscope through the laryngoscope. If it recurs after repeated endoscopic surgeries, a complete removal of the cyst can be achieved through an external approach[7]. However, an external approach may be preferred as the first surgical step depending on size and location of the cyst[2,3]. External approach may be performed through lateral cervical approach, laryngofissure or paramedian thyrotomy[2,3,7].

In conclusion, saccular cysts are rare lesions that should be kept in mind in the differential diagnosis of stridor in infants. Serious airway obstruction may ensue and obligate emergency medical intervention. Direct laryngoscopy is the best choice in the differential diagnosis. Complete excision of the cyst should be considered through an endoscopic or an external approach for permanent relief.

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