Subglottic cysts in a patient with recurrent stridor and respiratory distress

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Subglottic cysts are rare causes of stridor in infancy and should be suspected in the case of intubation. A 15-month-old male presented with recurrent stridor and respiratory distress. Prematurity and intubation were present in his history. Two subglottic cysts below the glottis were diagnosed. The cysts were ruptured by flexible bronchoscopy. The child's stridor and other symptoms disappeared thereafter. Early diagnosis of subglottic cysts is important since the obstruction can be relieved by rupturing the cysts with bronchoscope, whereas development of a fibrotic stenosis may require a tracheotomy, with its attending morbidity.

Key words: subglottic cyst, stridor, intubation.

Stridor following prolonged endotracheal intubation in infants may occur immediately after extubation, when it is usually attributed to mucosal edema of the larynx and trachea, or it may appear weeks or months after extubation, when fibrotic narrowing of the subglottic lumen is the usual finding, and subglottic stenosis is well recognized[1,2]. However, subglottic cysts have also been reported as a relatively rare, non-malignant cause of airway obstruction in infants[1-3]. Since the first case reported by Wigger and Tang[4] in 1968, the finding of subglottic cysts is becoming increasingly common; this may reflect a true increase in incidence, improved recognition, or both[3]. Herein, we report a prematurely born male infant who was intubated for a long period who presented with recurrent stridor and wheezing attacks and was diagnosed as having subglottic cysts. The distinction between subglottic cysts and stricture is important, since severe obstruction due to cysts can be relieved by rupturing the friable cysts by bronchoscope, whereas fibrotic stenosis may necessitate long-term tracheotomy, with its attendant morbidity[2].

Case Report

A 15-month-old male child was born at 28 weeks' gestation with a birth weight of 1250 g. During the follow-up, the child had been intubated for five days for respiratory distress syndrome, and had been extubated on the sixth day and was well at discharge. He was readmitted at the age of four months with respiratory distress characterized by stridor, cough and wheezing. The clinical status of the child improved rapidly with the use of cold vapor, salbutamol and a single dose of steroid, and he was discharged a few days later. The stridor and respiratory distress recurred, and readmissions to emergency medicine were noted. He was first admitted to our clinic when he was 15 months old with inspiratory stridor, cough and respiratory distress. He did not respond to the treatment of inhaled salbutamol or cold vapor. A double-probe pH monitoring of the gastrointestinal system revealed gastroesophageal reflux, and antireflux treatment was started. The wheezing disappeared after the antireflux treatment but the stridor persisted, especially during the daytime. To visualize the upper airways, a flexible bronchoscopy was performed, and two subglottic cysts were revealed 3 cm below the glottic region (Fig. 1); they were ruptured via the endoscopic procedure (Fig. 2). The child was well after the procedure and his stridor disappeared, with the relief of other symptoms.

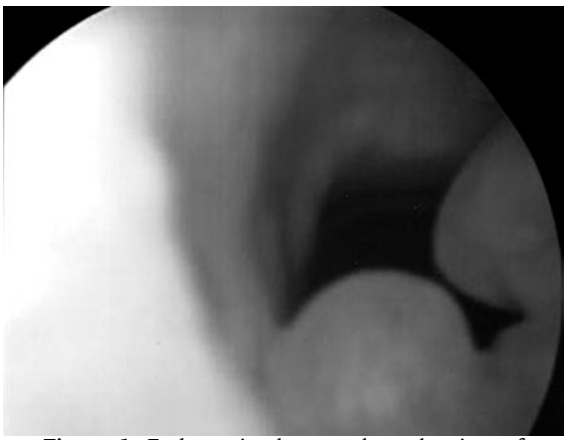

Figure 1. Endoscopic photograph at the time of the flexible bronchoscopy. Two cysts are seen to compromise the subglottic airway.

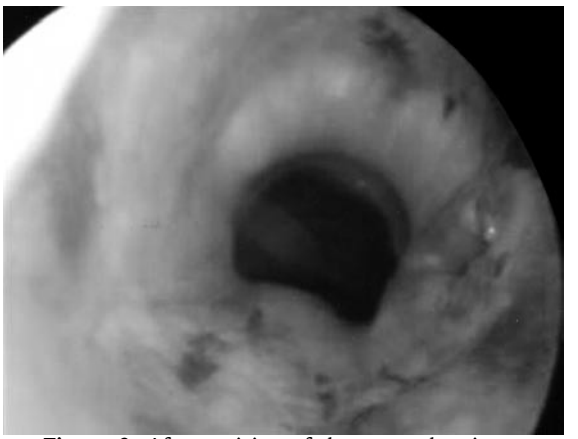

Figure 2. After excision of the cysts, the airway passage opened completely.

One month later, a follow-up laryngoscopy was performed, and a normal mucosal surface was visualized at the former site of the cysts. The child has been under follow-up for three months without any symptoms.

Discussion

Subglottic cysts are rare but increasingly recognized lesions of the infant larynx. Since they are attributed to endotracheal intubation and the survival of pre-term infants has improved, the condition could well be expected to increase[2,3,5]. It is difficult to distinguish between congenital and acquired subglottic cysts since neonates with respiratory distress at birth are usually intubated prior to any endoscopic examination of the larynx. From previous reports in the literature, it seems that subglottic cysts are usually acquired lesions[1-6]. Acquired subglottic cysts develop in response to intubation-induced mucosal damage. Subglottic ulceration and mucosal necrosis lead to the production of granulation tissue and healing with epithelial re-growth, subepithelial fibrosis and squamous metaplasia. Subepithelial fibrosis and squamous metaplasia can obstruct the ducts of the mucous glands in the subglottis, leading to the formation of retention cysts[4,7,8]. In our case, five days' intubation followed by four months of general well-being suggests that the subglottic cysts of the case were acquired rather than congenital and were associated with the intubation.

The median duration of intubation is also important for the development of subglottic cyst formation, but as little as 20 hours is sufficient for mucosal damage and necrosis to occur[9]. In our patient, the intubation period was five days.

Tierney et al. [5] suggested that the true prevalence of subglottic cysts has been underreported. Over a six-month period, they diagnosed five cases of compressible subglottic cysts in low birth weight, preterm infants who were intubated from 3 to 14 days in the neonatal period and presented with stridor and severe respiratory distress at 7-18 months of age. In our case, the first symptoms appeared at four months of age. This is because the retention cyst development takes time and the patients present an asymptomatic time interval.

The largest series of patients with subglottic cysts in the literature was reported by Lim et al.[3]. In their study, 55 patients were followed for a minimum of six months after diagnosis of subglottic cysts. All patients underwent intubation in the neonatal period. Fifty-one patients underwent intervention for cysts during laryngoscopy or bronchoscopy. The treatment of subglottic cysts also decreased the need for treatment of the patients with associated subglottic stenosis by 81%. There was a recurrence rate of 22/51. The authors emphasized the importance of long-term follow-up because of the potential recurrence of the subglottic cysts. In our case, the first follow-up laryngoscopy revealed a normal mucosal line, but repeated follow-up visits are needed to rule out recurrence.

The key to recognition and early diagnosis of subglottic cysts is a high index of suspicion in the differential diagnosis of airway obstruction in an infant with a history of intubation during the neonatal period. For the diagnosis, direct laryngoscopy and bronchoscopy are critical. Early conservative endoscopic treatment of subglottic cysts may preclude the development of subglottic stenosis and obviate the need for tracheotomy[6].

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