

## Case Report

# A rare case of congenital laryngeal cyst and its surgical management

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### ABSTRACT

Congenital laryngeal cyst is a rare occurrence. It is an important but rare cause of neonatal stridor and should be kept as a provisional diagnosis after ruling out laryngomalacia. The case study tells us the importance of doing a laryngoscopic examination in every neonate presenting with stridor. The patient in our study underwent endoscopic marsupialization of the cyst which happens to be the treatment of choice in case of a laryngeal cyst.

**Keywords:** Laryngeal cyst, Congenital laryngeal cyst, Vallecular cyst, Endoscopic marsupialization

## INTRODUCTION

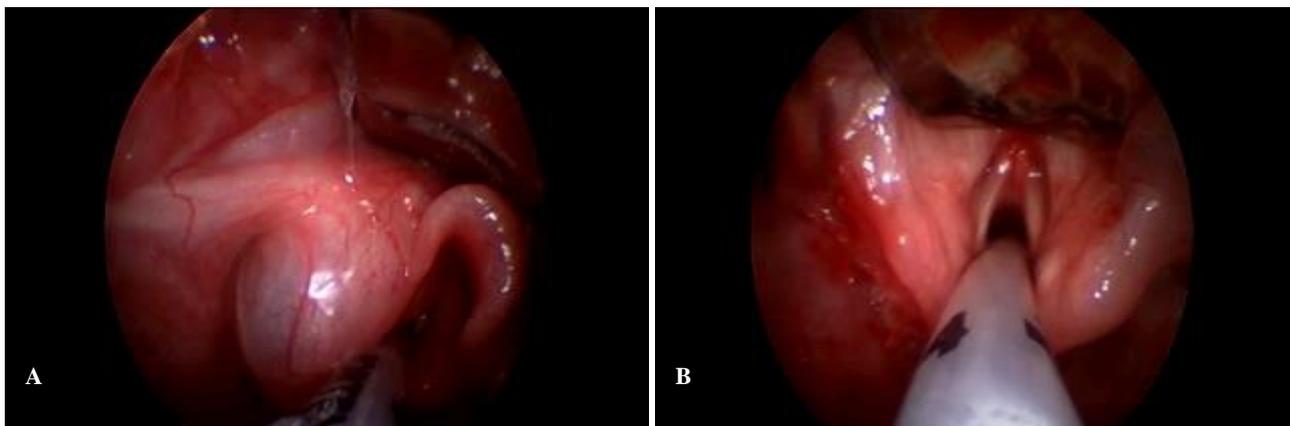
Laryngeal cysts are rare, can be congenital or acquired and may present with respiratory obstruction in infants and young children. Congenital laryngeal cyst is a rare occurrence with an incidence of 1.8/ 100,000 new borns.<sup>1</sup> Laryngeal cyst is a rare, but an important cause of stridor and respiratory distress in an infant. The initial presentation may mimic that of laryngomalacia, which happens to be the most common laryngeal anomaly and cause of stridor in newborns.<sup>2</sup> Laryngeal cysts are categorized as saccular or ductal types as per the classification given in 1970 by DeSanto et al.<sup>3</sup> This classification however did not include congenital cysts as a separate entity. A new classification system was proposed by Forte et al exclusively for congenital laryngeal cysts based on the anatomical extension of cyst who classified it into type I (intra-laryngeal cyst) and type II (extra-laryngeal extension).<sup>4</sup> This new classification was also aimed to help decide on the treatment protocol, with endoscopic excision recommended for type I and open surgical approach preferred for type II cysts. Here we describe our experience with a case of a congenital laryngeal cyst (type I).

## CASE REPORT

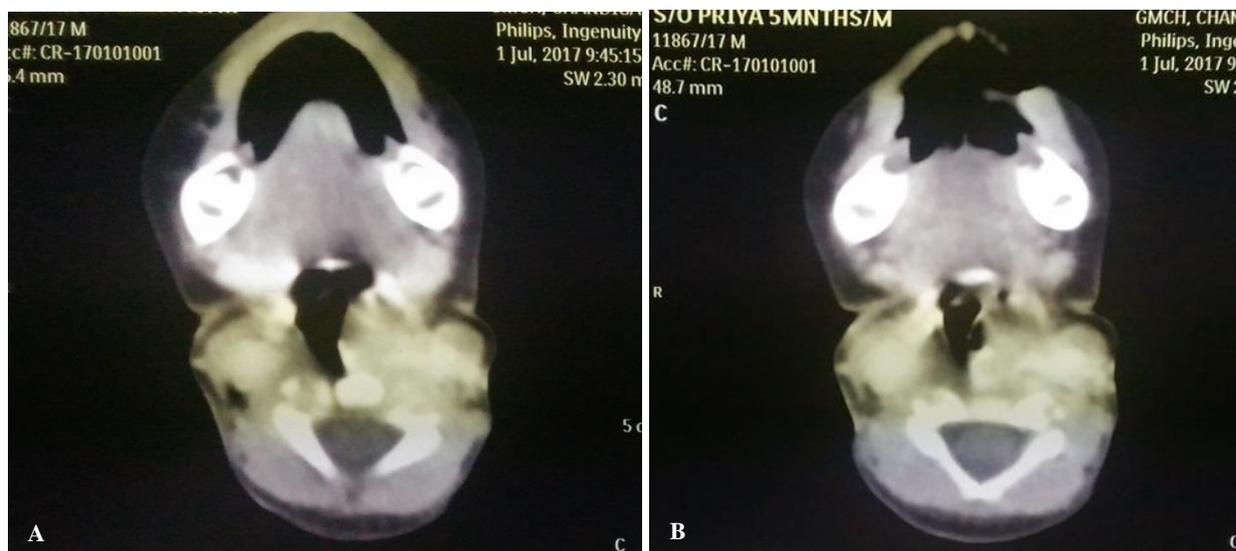
A 5 month old male child presented to OPD with complaint of difficulty in breathing since birth. It was progressing slowly but 2 weeks after birth, the distress increased, especially on crying or agitation. For the past 1 month, the child had respiratory distress even at rest and while sleeping, which increased significantly on crying. The child's birth history was normal and the child was born with a birth weight of 2720 gm with APGAR score 9 and 10 at 1 and 5 minutes respectively. On examination, the child was conscious, afebrile, maintaining an oxygen saturation of 95% on room air with the pulse rate of 166/minute. Inspiratory stridor was present. On respiratory system examination, the child showed use of accessory muscles of respiration with suprasternal retractions and bilateral intercostal retractions present. A paediatric video-laryngoscopy was performed which showed presence of an unidentified mass in the supraglottis region. A contrast enhanced CT of base of skull to forth thoracic vertebra was performed which showed signs of a cystic swelling in supraglottis following which a direct laryngoscopy was performed which confirmed the diagnosis of laryngeal cyst (Figure 1 and 2). Endoscopic marsupialization of the cyst was performed using powered microdebrider under general

anesthesia and the excised sample was sent for histopathological examination which was reported as a laryngeal cyst containing respiratory epithelium with

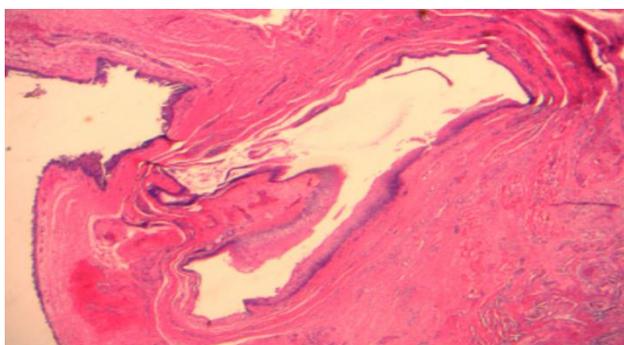
mucous glands and external lining of squamous epithelium (Figure 3).



**Figure 1: (A) Direct laryngoscopic view showing cystic lesion in supraglottis; (B) supraglottis after endoscopic marsupialization of the cyst.**



**Figure 2 (A and B): CT showing cyst present in supraglottis.**



**Figure 3: Histopathological examination of the laryngeal cyst shows respiratory epithelium with mucous glands and external lining of squamous epithelium.**

Post operatively, patient’s intercostal retractions were absent and no noisy breathing was noticed. Patient was discharged on post-operative day 2 and is doing well after 1 year post-operative follow-up.

**DISCUSSION**

Stridor is a high pitch noisy sound due to obstruction of larynx and trachea. History and examination are insufficient for a child in stridor for a firm diagnosis. Laryngomalacia is the most common congenital cause of stridor<sup>5</sup>. The stridor caused by laryngomalacia is variable, noticeable mostly when the child is active and disappears when the child is asleep. The severity of the stridor tends to increase in the first few months of life as the child

becomes more active and then gradually diminishes by the age of 2 years.

Congenital laryngeal cysts are rare in occurrence and an even rarer, but important cause of stridor in an infant. They can be congenital or acquired. DeSanto et al divided all laryngeal cysts into saccular, ductal and thyroid cartilage foramina cysts with ductal cysts being the more common variety originating from obstruction of the laryngeal epithelial mucous glands.<sup>3</sup> Forte et al proposed a separate classification for congenital laryngeal cysts based on the extent of the cyst and the embryological tissue of origin.<sup>4</sup> The new classification system also suggested the surgical treatment of choice to be endoscopic marsupialization of the cyst for congenital type 1 intra-laryngeal cyst.

History and examination alone are insufficient for a firm diagnosis to be drawn in case of a child presenting with stridor. Awake flexible endoscopy is useful but gives little or no information beyond the level of glottis. The definitive diagnostic technique is Laryngotracheobronchoscopy. Congenital laryngeal cysts are uncommon lesions that present in the neonatal period with stridor and respiratory distress. The severity and timing of appearance of symptoms depends on the size and location of the cyst.<sup>6</sup> The most constant symptom is stridor which mimics the presentation of the most common cause of stridor in an infant, which is laryngomalacia. There may be other symptoms like muffled cry, feeding difficulty, hoarseness, cyanotic episodes and phonation defects depending on the size of the cyst. An important point of differentiation between laryngomalacia and laryngeal cysts is that in laryngomalacia, the stridor typically improves with prone position, whereas it decreases when the patient lies on the affected side in case of laryngeal cyst.<sup>7</sup>

Initial suspicion of laryngomalacia, which is far more common can be ruled out by performing laryngoscopy in every stridorous child.<sup>8</sup> In our case, the child came with difficulty in breathing since birth which aggravated on crying, was investigated with the help of pediatric bronchoscopy which revealed some unidentified mass just at supraglottis level for which ENT consultation was taken after which CECT-Base of skull to fourth thoracic vertebra was performed which showed signs of cystic swelling at supraglottis following which direct laryngoscopy was performed which confirmed the diagnosis of laryngeal cyst. Recommended treatment depends on the extent of the cystic lesion, with the treatment of choice being endoscopic marsupialization for intra-laryngeal cysts and for a cyst with extra-

laryngeal extension, an attempt should not be made to excise or marsupialize the cyst endoscopically, which will likely lead to incomplete removal and subsequent recurrence of the cyst.<sup>4</sup>

## CONCLUSION

Although rare, congenital laryngeal cyst should be considered as a diagnosis in a child presenting with stridor after laryngomalacia has been ruled out. The two may co-exist as well and the majority of infants with laryngeal cyst have been reported to have concurrent laryngomalacia. The aforementioned case highlights the importance of early diagnosis and management of neonatal laryngeal cyst and endoscopic marsupialization is considered the gold standard procedure for the same.

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