

## Case Report

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# Congenital laryngeal cyst causing respiratory distress in a neonate: a rare case report

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## ABSTRACT

Congenital laryngeal cyst is a rare but responsible of upper airway obstruction which can be potentially life-threatening. The most common symptoms are inspiratory stridor, dyspnea, cyanosis, abnormal voice and difficulty with feeding. It should be differentiated from laryngomalacia. It is diagnosed by flexible fibro-optic laryngoscopy. Surgical options are endoscopic excision, needle aspiration, de-roofing, external laryngo-fissure and lateral pharyngotomy. The best treatment consists in the entire removal of the cyst. This case report described the case of 28 days old neonate with a severe airway distress and stridor caused by a congenital laryngeal cyst which has been managed by de-roofing of the cyst entirely with flexible laryngoscope.

**Keywords:** Congenital laryngeal cyst, Inspiratory stridor, Flexible fibro-optic laryngoscopy

## INTRODUCTION

Congenital laryngeal cyst is a rare occurrence with an incidence of 1.8/100,000 new borns.<sup>1</sup> But it is 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> Delay in diagnosis may lead to serious and fatal consequences.

Clinical presentation consists of inspiratory stridor, and varying degrees of upper airway obstruction that usually present soon after birth or during the first weeks or months of life. They are usually diagnosed by laryngoscopy. Several surgical procedures are proposed: endoscopic excision, needle aspiration, de-roofing, external laryngo-fissure, and lateral pharyngotomy.

The following report describes the case of 28 days old neonate with a severe airway distress and stridor caused by a congenital laryngeal cyst.

## CASE REPORT

A 28 days old neonate (Figure 1) was initially admitted into pediatric intensive care unit with respiratory distress. Suspecting infectious etiology, the patient had received antibiotics. However, symptoms gradually worsened, and the patient developed laryngeal stridor, for which, he was referred to pediatric surgeon for further management. On examination, the baby was toxic, cyanosed, with intercostal, subcostal and suprasternal retractions and features of failure to thrive. The patient was on high flow oxygen support initially along with injectable antibiotics and intravenous fluids. No significant improvement was noted even after 3 days of the therapy. X-ray soft tissue neck (lateral view) was done which showed narrowing of the laryngeal inlet. The baby was planned for examination under anesthesia next day. Trans oral flexible laryngoscopy was done which revealed a supraglottic mass (Figure 2) reducing the laryngeal lumen. Under general anesthesia, the patient underwent direct laryngoscopy, which confirmed a supraglottic cystic mass of the left

aryepiglottic fold. De-roofing of the cyst entirely with flexible laryngoscope done, and the cyst wall was sent for biopsy which revealed stratified squamous epithelium lined cyst wall with mild lymphocytic infiltrates (Figure 5). Post-operative period was uneventful. The patient's breathing was quiet and satisfying (Figure 4). X-ray soft tissue neck (lateral view) was repeated on post-operative day 3 which showed significant improvement (Figure 3). Oral feeds were started from post-operative day 4 and the baby was discharged on post-operative day 5 with the advice of monthly follow-up. For the last 3 months, the patient had no significant complaints with adequate weight gain.



**Figure 1:** Pre-operative picture of the baby showing intercostal, and suprasternal recession.



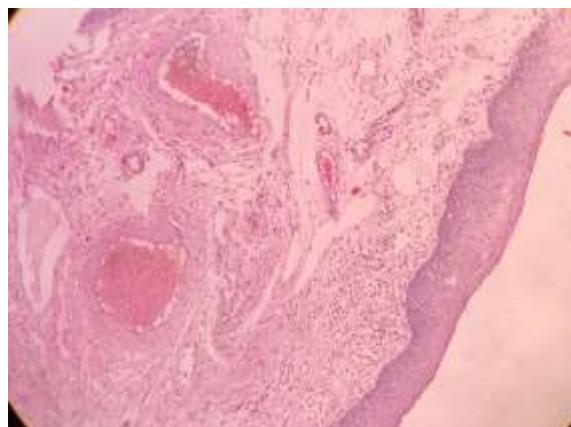
**Figure 2:** Flexible fibro-optic laryngoscopic view of left sided laryngeal cyst.



**Figure 3:** X-ray of lateral view of soft tissue neck (A) post-operative; and (B) pre-operative.



**Figure 4:** Post-operative picture of the baby in the follow-up day 14.



**Figure 5:** Histopathological picture of excised cyst.

## DISCUSSION

Congenital laryngeal cysts are rare but can be unfortunately fatal.<sup>3</sup> The majority of congenital laryngeal cysts arise from the vallecula, the saccule of the ventricle, and the aryepiglottic fold, and rarely from the epiglottis.<sup>4</sup> Laryngeal cysts are categorized as saccular or ductal types as per the classification given in 1970 by Santo et al.<sup>5</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>6</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. In case of endoscopic procedure, they are two choices. The first is needle aspiration considered insufficient because of the high risk of re-fitment of the cyst. The second possibility recommended is endoscopic de-roofing or complete excision with micro-instrument or CO<sub>2</sub> laser. Congenital laryngeal cysts are responsible of upper airways obstruction which can be potentially life-threatening. The most common symptoms are inspiratory stridor, dyspnea, cyanosis, abnormal voice and difficulty with feeding. The

severity and timing of appearance of symptoms depends on the size and location of the cyst.<sup>7</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 the 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>8</sup> Fiber-optic laryngoscopy and direct laryngoscopy are used to confirm and show the exact site of the cyst if there is no severe respiratory distress. In case of severe respiratory distress, tracheostomy can be necessary. Other modalities, as CT scan, MRI may be performed for further evaluation.<sup>9</sup> The best treatment consists in the entire removal of the cyst.<sup>10</sup>

For which many surgical procedures are proposed. They include endoscopic excision, de-roofing, needle aspiration, lateral pharyngotomy; and this after securing the airway generally with endotracheal intubation or with a tracheostomy sometimes.<sup>11</sup> In case of large cyst or recurrence, cervical approach is recommended, since it offers a good exposure of the pharyngeal space.

## CONCLUSION

Congenital laryngeal cysts are a rare and uncommon cause of airway obstruction leading to inspiratory stridor during the neonatal period or early infancy. It should be differentiated from laryngomalacia. Trans-nasal flexible laryngoscopy is useful in establishing the diagnosis as well as for the following up. Treatment of choice is complete excision by an endoscopic approach or through an external cervical procedure.

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