Case Report

Managing airway distress in a 2-month-old infant with epiglottic cyst

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ABSTRACT

Background: Epiglottic cysts, although rare, are significant causes of neonatal airway obstruction, necessitating prompt recognition and intervention. **Purpose:** To emphasize the importance of accurate diagnosis and management of airway distress. **Case report:** A two-month-old infant exhibited persistent tachypnea, inspiratory stridor, and intercostal retractions. Initially misdiagnosed as severe pneumonia, the infant's condition did not improve with antibiotics. Video-assisted laryngoscopy revealed an obstructive cyst on the epiglottis, which was successfully managed by aspiration, resulting in significant clinical improvement. **Clinical question:** What are the most effective diagnostic and therapeutic approaches for managing epiglottic cysts in neonates? **Method:** Thirteen published cases of epiglottic and other laryngeal cysts in neonates, were analyzed, focusing on their clinical presentations, diagnostic methods, management strategies, and outcomes. Data were extracted and summarized in a table for comparison. **Result:** The review identified common symptoms, including respiratory distress and feeding difficulties. Initial misdiagnosis was prevalent, with many cases attributed to pneumonia or laryngomalacia. Video-assisted laryngoscopy emerged as the gold standard for diagnosis, supplemented by CT and MRI. Management varied from cyst aspiration to endoscopic ablation and microsurgical excision, with positive outcomes in most cases. The objective was to summarize the findings in a review manner, highlighting trends and commonalities among the cases. **Conclusion:** The successful management of this case through cyst aspiration highlighted the importance of timely and accurate diagnosis, and the necessity for standardized diagnostic and therapeutic protocols. Future research should aim to refine diagnostic criteria and develop consensus guidelines, to enhance patient outcomes in this vulnerable population.

Keywords: epiglottic cysts, neonatal airway obstruction, respiratory distress, video-assisted laryngoscopy

ABSTRAK

Latar belakang: *Kista epiglotis, meskipun jarang, merupakan penyebab signifikan obstruksi saluran napas pada neonatus, yang memerlukan pengenalan dan intervensi yang cepat.* ***Tujuan:*** *Untuk menekankan pentingnya diagnosis dan penatalaksanaan akurat.* ***Laporan kasus:*** *Bayi berusia dua bulan menunjukkan takipnea persisten, stridor inspiratorik, dan retraksi interkostal. Awalnya salah didiagnosis sebagai pneumonia berat, dan kondisi bayi ini tidak membaik dengan pemberian antibiotik. Video-laringoskopi mengungkapkan adanya kista obstruktif pada epiglotis, yang berhasil dikelola dengan aspirasi, dan menghasilkan perbaikan klinis yang signifikan.* ***Pertanyaan klinis:*** *Apa pendekatan diagnostik dan terapeutik yang paling efektif untuk mengelola kista epiglotis pada neonatus?* ***Metode:*** *Tigabelas kasus yang dipublikasikan tentang kista epiglotis dan kista laring lainnya pada neonatus dianalisis, dengan fokus pada presentasi klinis, metode diagnostik, strategi penatalaksanaan, dan hasilnya. Data diekstraksi dan dirangkum dalam tabel untuk perbandingan.* ***Hasil:*** *Tinjauan ini mengidentifikasi gejala umum, termasuk kesulitan bernapas dan kesulitan pemberian makan. Diagnosis awal kerap kali salah, dengan banyak kasus yang dianggap berhubungan dengan pneumonia atau laringomalasia. Video-laringoskopi muncul sebagai standar emas untuk diagnosis, dilengkapi dengan CT dan MRI. Penatalaksanaan bervariasi dari aspirasi kista hingga ablasi endoskopik dan eksisi bedah-mikro, dengan hasil positif pada sebagian besar kasus. Hasil temuan dirangkum dalam bentuk tinjauan, menyoroti gejala klinis dan kesamaan di antara kasus-kasus tersebut.* ***Kesimpulan:*** *Penatalaksanaan yang berhasil dari kasus ini melalui aspirasi kista, menitikberatkan pentingnya diagnosis yang tepat waktu dan akurat, serta kebutuhan akan protokol diagnostik dan terapeutik yang*

terstandarisasi. Penelitian di masa depan harus bertujuan untuk memperbaiki kriteria diagnostik, dan mengembangkan pedoman konsensus, untuk meningkatkan keberhasilan menolong pasien dalam populasi yang rentan ini.

Kata kunci: *kista epiglotis, obstruksi saluran napas neonatus, kesulitan bernapas, video- laringoskopi*

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INTRODUCTION

Neonates represent a highly susceptible demographic for respiratory emergencies, attributable to a multitude of anatomical and physiological factors. Their airways, significantly smaller in diameter compared to adults, exhibit heightened airway resistance, especially under conditions of generalized airway narrowing such as edema.[1] Furthermore, distinct anatomical features in neonates, including a relatively larger tongue, a natural propensity for neck flexion in the supine position, and a longer, omega-shaped, and less flexible epiglottis, predispose them to airway obstructions.[1–3] These factors, collectively render respiratory management in neonates particularly challenging, and potentially life-threatening.

Epiglottic cysts, though a rare pathological entity, are notably implicated in neonatal airway obstruction. The pathophysiology of epiglottic cysts remains inadequately understood, and there is a lack of standardized diagnostic and therapeutic protocols.[4,5] As such, documenting case reports of epiglottic cysts becomes essential for elucidating the clinical presentation, enhancing diagnostic accuracy, and refining management strategies.

The literatures review indicated that while the occurrence of epiglottic cysts in neonates is uncommon, their impact on airway patency necessitated prompt recognition and intervention. Various studies had highlighted the anatomical predispositions and clinical manifestations associated with epiglottic cysts. For instance, a review by Vijayasekaran et al.[1] emphasized the significant anatomical differences in neonatal airways that contributed to increased vulnerability to obstruction. Additionally, Park et al.[2] and Mandal et al.[3] elaborated on the structural characteristics, such as the relatively larger tongue and unique epiglottic morphology, which exacerbated the risk of airway blockage.

Despite these insights, the literature revealed a gap in standardized approaches to the diagnosis and management of epiglottic cysts in neonates. Baljosevic et al.[4] and Guo et al.[5] emphasized the variability in clinical practice, highlighting the need for more consistent guidelines. This case report aimed to contribute to the existing knowledge by detailing a specific instance of an epiglottic cyst in a neonate, describing the diagnostic challenges encountered, and the management strategies employed.

CASE REPORT

A two-month-old infant was referred to the Emergency Department with persistent respiratory difficulties. The infant presented with tachypnea, inspiratory stridor, and suprasternal and intercostal retractions. A chest X-ray revealed perihilar infiltrates. The patient had previously been treated with antibiotics for presumed severe pneumonia at another hospital, but her respiratory status showed no improvement.

The infant was born via spontaneous vaginal delivery with a birth weight of 2.580 grams, and had no prior surgical history. An

echocardiogram revealed an atrial septal defect (ASD); however, this was deemed unrelated to her respiratory symptoms.

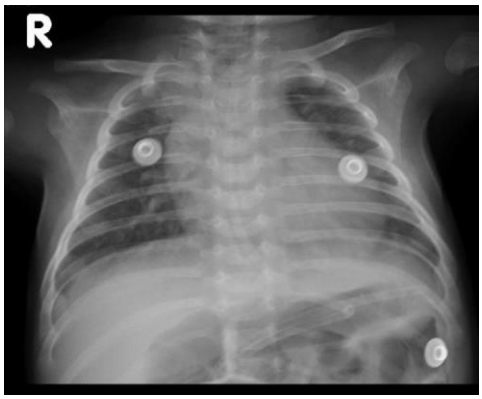

Figure 1. Chest X-ray of the patient

A video-assisted laryngoscopy was performed to assess the need for intubation. The procedure revealed a cyst obstructing the epiglottis during inspiration. To prevent impending respiratory failure, the initial management involved puncturing the cyst with a spinal needle to alleviate pressure. Approximately 3 mL of mucinous fluid was aspirated, leading to a significant reduction in the cyst's size and a marked improvement in the patient's clinical condition. The respiratory distress subsided substantially following the aspiration. An elective procedure was then scheduled for further definitive management.

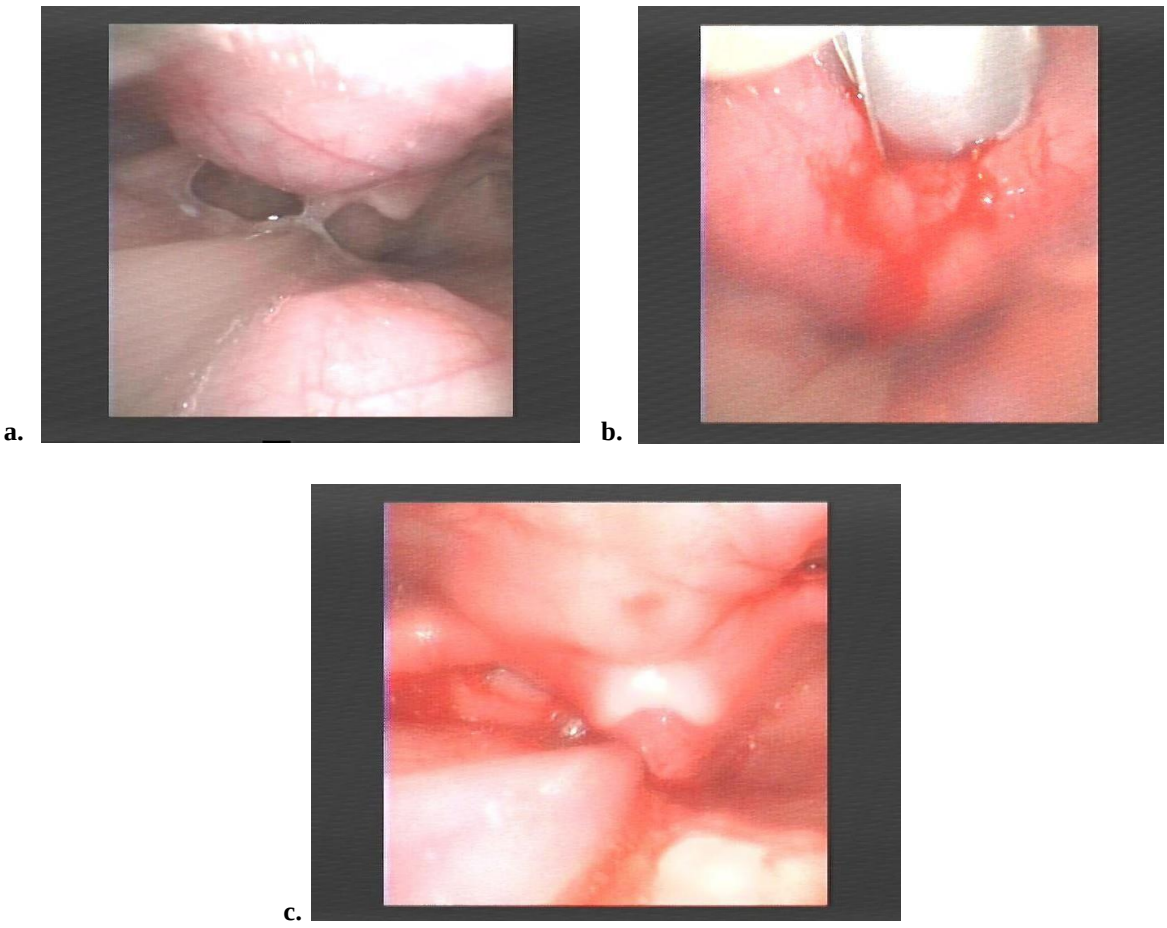

Figure 2. a. The visualization of the cyst before aspiration; b. Aspiration of the cyst by spinal needle; c.The post-aspiration visualization of the larynx.

CLINICAL QUESTION

What are the most effective diagnostic and therapeutic approaches for managing epiglottic cysts in neonates?

METHOD

Thirteen published cases of epiglottic and other laryngeal cysts in neonates were analyzed, focusing on their clinical presentations, diagnostic methods, management strategies, and outcomes. Data were extracted from various case reports to create a comprehensive summary of the current understanding of this condition. Each case was organized into a table format for easy comparison, facilitating a clearer understanding of the diagnostic and therapeutic approaches used across different reports.

RESULT

The review identified 13 cases of neonates diagnosed with epiglottic and other laryngeal cysts. Clinical symptoms commonly included respiratory distress, stridor, and feeding difficulties. Initial misdiagnosis of these cases was prevalent, as symptoms were often attributed to more common conditions such as pneumonia or laryngomalacia. Diagnostic approaches varied, with fiberoptic laryngoscopy emerging as the gold standard for direct visualization, supplemented by imaging techniques like CT and MRI to assess cyst size and location.

The management strategies also varied, ranging from conservative approaches such as cyst aspiration, to more invasive methods including endoscopic ablation using low-temperature plasma radiofrequency (LPRF), and microsurgical excision. Positive outcomes were observed in most cases, with symptoms resolving after appropriate interventions. The outcomes, management techniques, and any associated anomalies for each case were summarized in Table 1.

DISCUSSION

This case report presented a two-month-old infant experiencing severe respiratory distress due to an epiglottic cyst, a rare yet significant cause of airway obstruction in neonates. The infant exhibited persistent tachypnea, inspiratory stridor, and intercostal retractions; symptoms initially misattributed to severe pneumonia. Diagnostic evaluation through video-assisted laryngoscopy revealed an obstructive cyst on the epiglottis, which was successfully managed by aspiration, resulting in marked clinical improvement.

Epiglottic cyst, also known more generally as congenital laryngeal cysts, is a rare pathology causing neonatal airway obstruction. It is estimated that the incidence of laryngeal cysts in neonates as a whole is 1.8 to 3.5 per 100,000 live births.[4–6] Despite their rarity, late detection could result in fatality in up to 40% of cases.[7] The associated risk of fatality underscores the importance of timely diagnosis of epiglottic cysts as a critical step in ensuring better outcomes for patients.

The etiology of epiglottic cysts is not fully understood. However, it is known that epiglottic cysts can occur congenitally during the first days and weeks of a neonate's life, or due to acquired causes, most commonly as a result of endotracheal intubation.[8]

Diagnosing epiglottic cysts in neonates presents a significant challenge due to the rarity of the condition, and its potential overlap with other causes of airway obstruction. The diagnostic process typically involves a combination of clinical evaluation, imaging, and direct visualization techniques.

The initial step in diagnosing an epiglottic cyst involves a thorough clinical evaluation. Neonates with epiglottic cysts often present with symptoms of airway obstruction, including stridor, tachypnea, and respiratory distress. As highlighted by Marseglia et al.[9], a detailed clinical history and physical examination are essential to identify

obstructive symptoms and differentiate them from other common neonatal airway anomalies, such as laryngomalacia.

Imaging plays a crucial role in the diagnostic workup of epiglottic cysts. Computed tomography (CT) scans and magnetic resonance imaging (MRI) are frequently utilized to evaluate the size, location, and extent of the cyst, as well as to assess its impact on the airway.[10] Namshikar et al.[11] emphasized the importance of preoperative imaging to guide anesthesia and surgical planning, highlighting the use of CT scans in their case to delineate the cyst's characteristics, and plan for safe airway management.

Direct visualization through laryngoscopy remains the gold standard for diagnosing epiglottic cysts.[9,12] Video-assisted laryngoscopy allows for real-time assessment of the airway and direct visualization of the cyst. This method does not only confirm the diagnosis, but also helps determine the most appropriate intervention. In our case, video-assisted laryngoscopy revealed a cyst obstructing the epiglottis during inspiration, and guiding the subsequent aspiration procedure.

Fiberoptic laryngoscopy is another valuable diagnostic tool, providing detailed visualization of the laryngeal structures. It can be particularly useful in identifying cysts that may not be visible during initial examination. Marseglia et al.[9] underscored the importance of repeated laryngoscopy if symptoms persisted or worsen, as congenital laryngeal cysts might not be immediately visible, and could be misdiagnosed as other conditions like laryngomalacia.

Differentiating epiglottic cysts from other causes of neonatal stridor is critical. Conditions such as laryngomalacia, vocal cord paralysis, and other congenital laryngeal anomalies must be considered.[13] Detailed imaging and direct visualization help exclude these conditions and confirm the presence of an epiglottic cyst. The literature indicated that epiglottic cysts could co-exist with other laryngeal abnormalities, further complicating the diagnostic process. For instance, Marseglia et al.[9] reported cases where laryngomalacia co-existed with vallecular cysts, highlighting the need for comprehensive airway evaluation.

Management strategies for epiglottic cysts vary and the lack of standardized protocols contributing to variability in clinical practice. For example, Guo et al.[5]discussed the use of endoscopic ablation by low-temperature plasma radiofrequency (LPRF) as a novel and effective approach for treating congenital epiglottic cysts. This technique, which involves the precise removal of the cyst with minimal thermal damage to surrounding tissues, had shown promising results in terms of efficacy and safety.[14]

Anesthetic management of these cases is particularly challenging due to the risk of airway occlusion and the need for careful handling to prevent cyst rupture and pulmonary aspiration. Namshikar et al.[11] described the challenges faced during laryngoscopy, where the size and extent of the cyst could obscure critical anatomical landmarks, necessitating a gentle and precise approach. In our case, the successful aspiration of the cyst provided immediate relief from respiratory distress, underscoring the importance of timely and accurate diagnosis.

Moreover, the potential co-existence of laryngeal cysts with other conditions such as laryngomalacia adds complexity to the management of these patients. Marseglia et al.[9] reported on the frequent co-occurrence of laryngomalacia with vallecular cysts, noting that the presence of both conditions could exacerbate respiratory symptoms and complicate treatment. This highlights the need for comprehensive airway evaluation in neonates presenting with stridor and respiratory distress to ensure all contributing factors are identified and addressed.

In conclusion, this case report highlighted the critical importance of timely recognition and intervention in managing epiglottic cysts in neonates, which were rare but significant causes of airway obstruction. The diagnostic approach, encompassing clinical evaluation, imaging studies, and direct visualization through laryngoscopy, is vital for accurate diagnosis and effective management. This case underscored the need for standardized diagnostic and therapeutic protocols, given the variability in clinical presentations and the challenges in differentiating epiglottic cysts from other neonatal airway anomalies. The successful management of the presented case through cyst aspiration, illustrated the potential for immediate clinical improvement and the necessity for comprehensive airway evaluation in neonates presenting with respiratory distress. Future research should aim to refine diagnostic criteria, and develop consensus guidelines to enhance patient outcomes in this vulnerable population.

APPENDIX

Table 1. Summary of several published cases of epiglottic and other laryngeal cysts in neonates

Case#	M/F	Age	GW	Symptoms	Diagnostic approach	Cyst location	Management	Follow up	Outcome	Associated anomalies	Reference
1	M	10 days	37	Stridor, dyspnea	Fiberoptic laryngoscopy, CT-scan	Epiglottis, left ventricular fold	Excision by microsurgical instruments and CO2 laser	12 months	Cured	No	4
2	F	1 day	37	Stridor, dyspnea	Fiberoptic laryngoscopy, CT-scan	Right ventricular fold	Excision by microsurgical instruments and CO2 laser	12 months	Cured	Esophageal atresia	4
3	F	14 days	38	Stridor, dyspnea	Fiberoptic laryngoscopy, CT-scan	Epiglottis, left ventricular fold	Excision by microsurgical instruments and CO2 laser	12 months	Recurrence	Cri-du Chat syndrome	4
4	M	7 days	38	Stridor, dyspnea	Fiberoptic laryngoscopy, CT-scan	Left ventricular fold	Excision by microsurgical instruments and CO2 laser	12 months	Cured	No	4
5	M	10 days	38	Stridor, dyspnea	Fiberoptic laryngoscopy, CT-scan	Right ventricular fold	Excision by microsurgical instruments and CO2 laser	12 months	Cured	Polydactyly	4
6	F	28 days	39	Feeding difficulty, stridor, respiratory distress, failure to thrive	Endoscopic laryngoscopy, MRI	Valecula	Marsupialization and excision	3 months	Cured	Laryngo-malacia type 3	9
7	M	2 months	n/a	Dyspnea, choking, coughing	Fiberoptic laryngoscopy	Epiglotis	Endoscopic ablation by low-temperature plasma radiofrequency (LPRF)	3 months	Cured	No	5
8	M	8 days	n/a	Dyspnea, poor feeding	Fiberoptic laryngoscopy	Left lingual epiglotis	Endoscopic ablation by low-temperature plasma radiofrequency (LPRF)	1 months	Cured	No	5
9	M	5 days	n/a	Dyspnea	Fiberoptic laryngoscopy	Right lingual epiglotis	Endoscopic ablation by low-temperature plasma radiofrequency (LPRF)	2 weeks	Cured	No	5

10	F	2 months	term	Respiratory distress, failure to thrive, stridor	Fiberoptic bronchoscopy, contrast CT scan, 99 Tc scan	Valecula	Excision using Arthrocare ENT coblator system	3 months	Cured	No	15
11	M	3 months	term	Stridor, respiratory distress	Fiberoptic bronchoscopy, contrast CT scan, 99 Tc scan	Valecula	Excision using Arthrocare ENT coblator system	2 months	Cured	No	15
12	M	40 days	term	Respiratory distress, stridor	Fiberoptic bronchoscopy, contrast CT scan, 99 Tc scan	Valecula	Excision using Arthrocare ENT coblator system	24 hours	Cured	Hypothyroidism	15
13	M	4 months	term	Failure to thrive, respiratory distress	Contrast CT scan	Valecula	Excision using Arthrocare ENT coblator system	n/a	Cured	No	15

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