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TOPÇU

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Congenital laryngeal cyst presenting with severe respiratory distress in a newborn

Yenidoğan bebekte ciddi solunum sıkıntısı ile kendini gösteren konjenital larinks kisti

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ABSTRACT

In the newborn, congenital laryngeal cysts can cause life-threatening respiratory distress. Mortality can be prevented by early diagnosis. Therefore, a congenital laryngeal cyst should be considered in the differential diagnosis of an infant who develops respiratory distress and stridor. Flexible nasopharyngolaryngoscopy is a valuable tool in diagnosis. The treatment of the cyst is total surgical excision. This article reviews the literature and presents a case of congenital laryngeal cyst that caused respiratory distress at birth and was treated surgically.

Key words: Larynx, cyst, congenital, newborn, stridor

ÖZET

Konjenital larinks kistleri yenidoğanda hayatı tehdit edebilecek solunum sıkıntısına neden olabilmektedir. Erken teşhis ile mortalite engellenebilir. Bu nedenle solunum sıkıntısı ve stridor şikayeti olan bir bebekte konjenital larinks kisti ayırıcı tanıda düşünülmelidir. Esnek nazofaringolaringoskop çok değerli bir tanı aracıdır. Tedavisi kistin total cerrahi eksizyonudur. Bu yazıda; doğumdan itibaren solunum sıkıntısı oluşturan ve cerrahi olarak tedavi edilen konjenital larinks kisti olgusu literatür eşliğinde tartışılmıştır.

Anahtar kelimeler: larinks, kist, konjenital, yenidoğan, stridor

INTRODUCTION

Congenital laryngeal cysts are rare, but they can cause severe respiratory distress and stridor immediately after birth. Therefore, they require early diagnosis and treatment [1-3], as they can cause obstruction of the larynx, which is potentially fatal for the newborn [4]. The pathogenesis of laryngeal cysts involves possible obstruction of the submucosal glands or atresia of the laryngeal saccule [5]. Laryngeal cyst should be considered in the differential diagnosis of a newborn with stridor. Flexible nasopharyngolaryngoscopy is very helpful for making the diagnosis [6,7]. These cysts tend to recur if not excised completely. Consequently, these cysts should be excised completely under general anesthesia. Here, we present a 2-month-old baby who underwent transoral surgery two times to treat a congenital laryngeal cyst.

CASE REPORT

A full-term, 3350 gram male neonate developed stridor immediately after birth. Our otolaryngology

department was consulted 2 days after birth. Transnasal flexible laryngoscopy revealed a large cystic mass that seemed to arise from the right aryepiglottic fold and epiglottis. Direct laryngoscopy was performed under general anesthesia the next day. The cystic mass wall was punctured with a needle and serous liquid was aspirated.

Subsequently, the mass shrank, and the vocal cords could be seen. The baby was discharged from the neonatal intensive care unit (NICU) with no problems 3 days postoperatively. Five weeks later, the baby was admitted to the NICU because of respiratory distress that had increased gradually over the last 4 days. Magnetic resonance imaging (MRI) of the neck showed a 2×1 cm cystic mass at the epiglottis level (Figure 1). The baby was taken to the operating room, and intubated with a small tube. The cystic mass was larger than previously (Figure 2). The entire cystic mass was excised (Figure 3). The diagnosis of a congenital laryngeal cyst was confirmed histopathologically by the presence of normal squamous epithelial cells. The baby was

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discharged from hospital 5 days after total excision of the cystic mass.

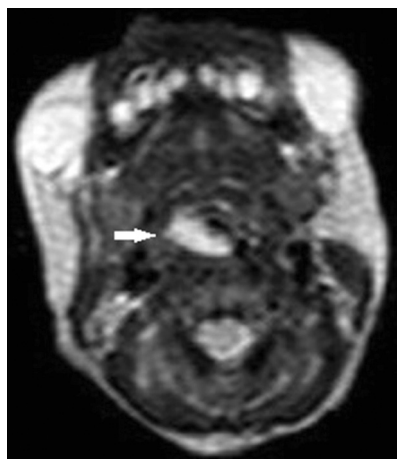


Figure 1. A mass in the larynx partially obstructs the airway on axial MRI (white arrow)

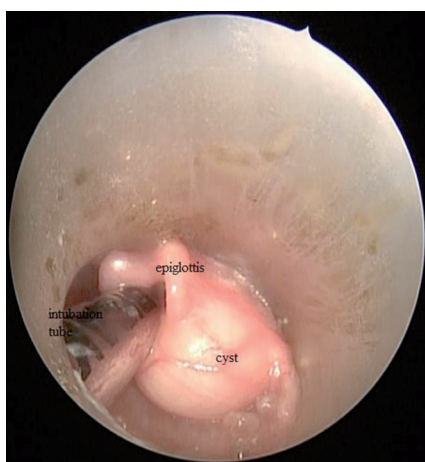


Figure 2. Endoscopic view of the cyst in the larynx



Figure 3. Laryngeal view after surgical excision of the cyst

DISCUSSION

Stridor is a common finding in the neonatal period, and it should be evaluated urgently [8]. Congenital laryngeal cysts are uncommon lesions and an infrequent cause of stridor in the neonatal period [2,3]. The incidence of congenital laryngeal cysts is 1.82 per 100,000 live births [4]. Usually, the patient presents with stridor and dyspnea, but can also present with cyanosis, feeding difficulties, and abnormal breath sounds [6,7]. Sometimes, a congenital laryngeal cyst can be misdiagnosed as laryngomalacia because of the similarity of symptoms [9]. Larger cysts cause respiratory distress immediately after birth, whereas smaller ones can cause respiratory symptoms that increase over time.

Laryngeal cysts mostly arise from the supra-glottic region and rarely from the glottis and sub-glottic region [1,3]. De Santo et al. [2] classified laryngeal cysts into thyroid-cartilage, saccular, and ductal cysts. This generally accepted classification has two main limitations: the classification was based on adults, and it does not consider cyst location. The differential diagnosis should include hemangioma, cystic hygroma, dermoid cyst, teratoma, lingual thyroid, laryngocele, laryngomalacia, laryngeal web, and vocal cord paralysis. Flexible nasopharyngolaryngoscopy is very important in making the diagnosis [6,7]. Although computed tomography (CT) and MRI are helpful in the diagnosis, a patient in distress should be taken to the operating room without obtaining images first [10].

Total surgical excision is the treatment of choice in congenital laryngeal cysts. The surgeon should be prepared to perform a tracheotomy if intubation fails, and the family should be informed about this possibility preoperatively [1,11]. As intubation can be challenging, an experienced anesthetist is required. Endoscopic excision is adequate for small cysts, but an external approach might be required for larger cysts [4,6,10]. After marsupialization, the cyst can recur.

In conclusion, laryngeal cysts should be considered in the differential diagnosis of a newborn with stridor, and complete surgical excision via an endoscopic or external approach should be performed urgently.

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