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Original Scientific Paper

TREATMENT OF CONGENITAL LARYNGEAL CYSTS IN NEWBORNS

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SUMMARY – Congenital laryngeal cysts are rare lesions that may occur in newborns, characterized by symptoms of respiratory obstruction and severe dyspnea. The aim of this study was to indicate optimal surgical treatment of congenital laryngeal cysts in newborns. We present a case series of five neonates treated for congenital laryngeal cysts during the 2011-2017 period at our pediatric tertiary care hospital. Patient age ranged from one day to 14 days. All patients had unilateral cysts. After surgical excision, burning of the walls of the cysts was performed by a CO2 laser. In one case, after four months, a recurrent cyst appeared, and re-operation was performed. Our relatively small case series indicates that symptoms such as stridor and labored breathing can occur already during the first days of life and potentially endanger the patient's life. Complete excision of the cyst and burning of its walls with CO2 laser leads to complete cure and prevent recurrence of the lesion.

Key words: *Cyst, congenital; Endoscopy; Larynx; Newborns*

Introduction

Stridor and difficulty breathing in newborns should always raise doubts about the presence of a congenital anomaly of the larynx, especially laryngomalacia and congenital laryngeal cyst. Congenital laryngeal cyst is a rare lesion that in a newborn can lead to respiratory obstruction and severe dyspnea[1-5]. Cyst growth can be rapid and can cause airway obstruction that potentially compromises the patient's life. The aim of this case series was to indicate optimal surgical treatment of these conditions in newborns. This is the first study concerning the diagnosis and treatment of congenital laryngeal cysts in newborns in our country.

Patients and Methods

This retrospective study was performed according to the Declaration of Helsinki at our pediatric university hospital during the 2011-2017 period. The study was approved by the institutional Review Board (approval No. 24/31) and written informed consent was obtained from all parents of treated newborns to use their patient data. All patients were in neonatal age, aged from one to 14 days. Patient data were processed by gestational week, sex, age, clinical presentations, diagnosis and treatment method (Table 1). The follow up period was 12 months after surgical treatment. All patients were admitted to the hospital due to stridor and difficulty

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Table 1. Clinical characteristics of treated patients

Age	Sex	Gestational week	Symptoms	Localization of cysts	Preoperative intervention	Postoperative intubation	Compli-cations	Follow up period	Recu-rrence	Associated anomalies
10 days	M	37	Stridor, dyspnea	Epiglottis and left ventricular fold	No	24h	No	12 months	No	No
1 day	F	37	Stridor, dyspnea	Right ventricular fold	No	48h	No	12 months	No	Atresia of esophagus
14 days	F	38	Stridor, dyspnea	Epiglottis and left ventricular fold	No	24h	No	12 months	Yes	Syndrome Cru de Chat
7 days	M	38	Stridor, dyspnea	Left ventricular fold	No	24h	No	12 months	No	No
10 days	M	38	Stridor, dyspnea	Right ventricular fold	No	24h	No	12 months	No	Polydactyly

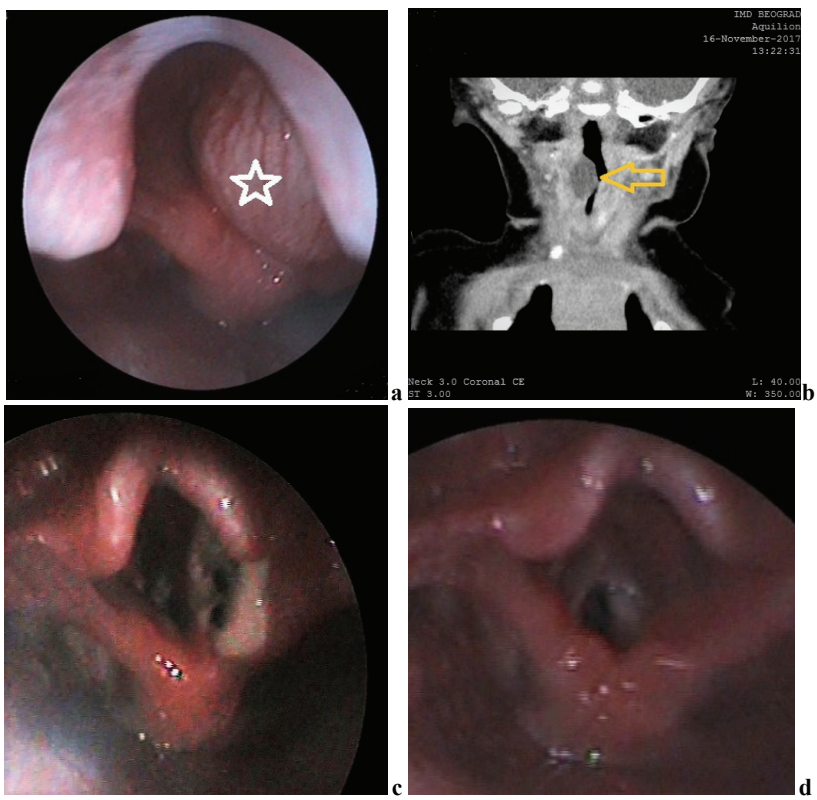

Fig. 1. (a) Endoscopic view of the congenital laryngeal cyst before surgery. Note the presence of laryngeal cyst (asterisk inside) on the right ventricular fold; (b) coronal plane of the computed tomography scan of the throat. Note the presence of a laryngeal cyst on the right lateral wall of the larynx (arrow head); (c) endoscopic view, 7 days after excision of the cyst from the right ventricular fold. Note the presence of two breaches on the inner surface of the larynx; (d) endoscopic view in the same patient 12 months after surgical treatment. Note the absence of breaches.

breathing. Fiberoptic laryngoscopy was the main procedure for setting the diagnosis (Fig. 1a, c, d). In four patients, computed tomography (CT) scan of the neck was performed to determine the exact localization and extension of the cysts (Fig. 1b).

In all patients, surgical interventions were performed under general anesthesia. Endotracheal tubes were inserted in all patients and treatment was carried out under the microscope (Leica F40, Leica Microsystems, Wetzlar, Germany). Laryngeal cysts were excised by microsurgical instruments. After removal of the cyst walls, burning of the walls and bases of the cyst was performed by a CO2 laser (AcuPulse™ Strength 3W, Lumenis Ltd., Yokneam, Israel). Patients were extubated 24 to 48 hours after the surgery. Postoperatively, they were treated with intravenous antibiotics and corticosteroids for the next three days. There were no postoperative complications and all patients were released from the hospital in good general and local condition. Control fiber laryngoscopy was performed seven days after discharge from the hospital (Fig. 1c), and then at one month, three, six and 12 months after the surgery (Fig. 1d).

Results

Out of five neonates, three were male and two female. The mean gestational week was 34±2. Symptoms appeared immediately after birth and the most common were difficulty breathing, intense stridor, cyanosis, and difficulty swallowing. Patient age ranged from one day to 14 days. All patients had unilateral cysts. Two children had anterior laryngeal cysts localized in the ventricular folds and epiglottis, two patients had cysts situated on the right ventricular fold extending towards the right pyriform sinus, and one had a cyst on the left ventricular fold. Three neonates had associated anomalies, including one cri du chat syndrome with a congenital heart defect (ventricular septal defect), polydactyly, and congenital atresia of the esophagus each. The patient who was operated on the first day of life had congenital atresia of the esophagus and an urgent surgical intervention was done. During intubation, a laryngeal cyst was found on the right ventricular fold, significantly narrowing the aditus of the larynx. It was estimated that it could complicate postoperative recovery, so the cyst was removed first and then esophageal surgery was performed. There were no early postoperative complications. In one case, four months after the treatment, a recurrent cyst was found, and re-operation was performed, also by using a CO2 laser. Histopathologic analysis showed that the inner surface of each cyst was covered by respiratory pseudostratified cylindric epithelium with small islands of seromucinous glands, which confirmed the diagnosis of congenital laryngeal cysts (Fig. 2).

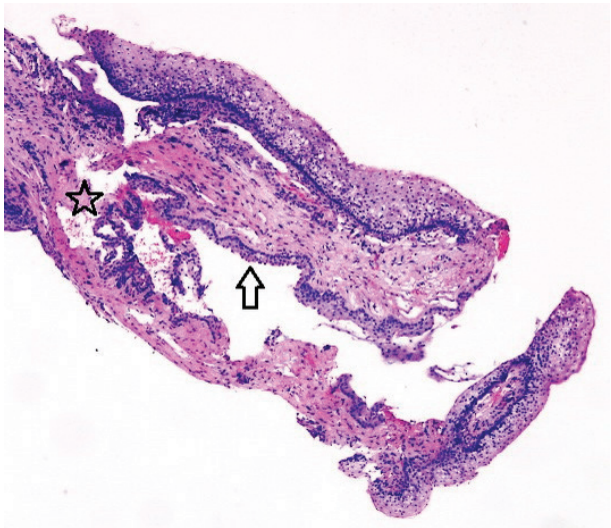

Fig. 2. Photomicrograph of an excised congenital laryngeal saccular cyst of the ventricular fold. The inner surface of the cyst is lined by pseudostratified respiratory epithelium (arrow head). Note the presence of seromucinous glands (asterisk) (hematoxylin-eosin staining, magnification X50).

Discussion

Laryngeal cysts represent rare lesions that can be found in 1.87 *per* 100,000 live-born children[2]. In neonates, these cysts are manifested by severe symptoms such as stridor, dyspnea, hoarseness, dysphagia, and dysphonia, and can cause mortality in up to 40% of cases if the diagnosis is not set timely[3]. Laryngeal cysts can be congenital and acquired. Congenital laryngeal cysts occur in the first days and weeks of life and are potentially fatal because they cause airway obstruction. Some laryngeal cysts are acquired, most often as a result of endotracheal intubation[4]. De Santo *et al.*[1] proposed the first classification of laryngeal cysts. They defined two types of laryngeal cysts, saccular and ductal. Saccular cysts are submucosal cysts caused by accumulation of mucus within the laryngeal sacculus. They occur in the supraglottic region, including aryepiglottic fold, epiglottis, vallecula and laryngeal

ventriculus or pyriform sinus. Ductal cysts are formed from liquids accumulated in a secondary manner because of obstruction of submucosal glands. Forte *et al*.[5] suggested the origin of the innate laryngeal cyst to occur due to the blockage of the saccular duct or atresia of the sacculus itself, and this typical localization allows the cyst to expand through structurally weaker areas of the throat, such as the thyrohyoid membrane. It can be assumed that it has embryonic origin if endoderm derivatives are found, such as respiratory epithelium and seromucinous glands[5]. In our study, histopathologic examination showed the presence of a mucous membrane lined by respiratory cylindric epithelium with small seromucinous glands in which acini were moderately dilated and filled with mucus. Regarding the level of extension, laryngeal cysts can be divided into type I cysts, which can be removed by endoscopic approach, and type II cysts that are spreading beyond the larynx and require an external surgical approach[5]. We removed all cysts endoscopically.

Typical symptoms of congenital laryngeal cysts are respiratory stridor that increases during cry, respiratory distress during feeding that leads to regurgitation, and cyanosis. Other laryngeal anomalies such as laryngocele have similar symptoms[6]. Symptoms depend on the size and localization of the cyst. They begin several hours after birth in 40% of patients and within 6 months after birth in 95% of patients[7]. Saccular laryngeal cysts cause 40% mortality if not diagnosed on time[8]. Nevertheless, in 50% of cases, they are without symptoms and are only detected on autopsy. Some authors note that as many as 35.7% of patients with congenital cysts are treated as laryngomalacia due to the similarities of their symptoms[9].

Visualization of the larynx by flexible endoscopy is very important for differential diagnosis of conditions in newborns with stridor. Laryngoscopy in general anesthesia represents an alternative method for detailed examination[10]. It is necessary to do a CT scan of the neck, by which detailed localization and extension of the cyst can be determined[11].

Some authors note that aryepiglottic cysts are the most common laryngeal cysts in children, then vallecular cysts, ventricular, and then subglottic cysts[12]. In our study, we diagnosed saccular cysts localized in the area of aryepiglottic and ventricular folds in three cases, and anterior saccular cysts localized in the area of epiglottis and aryepiglottic folds in two cases. As in our patients, laryngeal cysts are most often unilateral, but some authors also describe bilateral cysts in 37% of cases[9].

Laryngeal cyst needs to be surgically removed. Attempts at aspiration of the cyst by a needle or incision are not sufficient and do not lead to healing. According to analysis by other authors, the main time for the appearance of recurrent cysts after needle aspiration is 5 to 10 days, and after removal of the cyst roof alone, it is one to 10 months[9]. Cyst marsupialization can lead to improvement in symptoms, but recurrent cysts after excision can also frequently occur[6]. Complete cyst excision is the best choice for cure. Smaller saccular cysts can be removed endoscopically. However, big, extralaryngeal cysts require removal by external approach, performed through the side of the neck, laryngophoresis or paramedial thyrotomy[13-15]. Xiao *et al*.[9] have described the largest series of patients with saccular laryngeal cysts treated in a 10-year period. They recommend complete removal of the cyst wall by endoscopic approach using CO2 laser and micro suture of the walls. There were no recurrent lesions[9]. In our patients, after endoscopic removal of the cysts, burning of cyst walls by CO2 laser was performed. In one case, recurrent cyst was diagnosed, which was completely removed 6 months later, also using CO2 laser. There were no recurrences of cysts.

Conclusion

Congenital laryngeal cysts are rare anomalies. Our relatively small series indicates that symptoms such as stridor and impaired breathing can occur already during the first days of life and endanger the patient's life. Serious upper airway obstruction requires urgent intervention. Flexible endoscopy is the best choice for differential diagnosis. Complete excision of cyst and burning of walls with CO2 laser leads to complete cure and prevents recurrence of lesions.

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Sažetak

LIJEČENJE PRIROĐENIH CISTA GRKLJANA U NOVOROĐENČADI

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Prirođene laringealne ciste rijetke su lezije koje se mogu pojaviti kod novorođenčadi, a obilježene su simptomima respiracijske opstrukcije i teškom dispnejom. Cilj ove studije bio je ukazati na optimalno kirurško liječenje prirođenih cista na grkljanu kod novorođenčadi. Predstavljamo seriju slučajeva od pet novorođenčadi liječenih zbog prirođene ciste grkljana tijekom razdoblja od 2011. do 2017. godine u našoj dječjoj bolnici tercijarne skrbi. Najmlađi bolesnik bio je jednodnevni, a najstariji 14 dana. Svi su bolesnici imali jednostrane ciste. Nakon kirurške ekscizije izgaranje zidova cista izvedeno je CO2 laserom. U jednom slučaju, nakon 4 mjeseca, pojavila se rekurentna cista i učinjena je ponovna operacija. Naša relativno mala serija slučajeva ukazuje na to da se simptomi kao što su stridor i naporno disanje mogu pojaviti već prvih dana života i potencijalno ugroziti život bolesnika. Potpuna ekscizija ciste i paljenje zidova CO2 laserom dovodi do potpunog izlječenja i sprječava ponovni nastanak lezije.

Ključne riječi: *Cista, prirođena; Endoskopija; Grkljan; Novorođenčad*