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Case Report

A case of congenital vallecular cyst associated with gastroesophageal reflux presenting with stridor, feeding cyanosis, and failure to thrive

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= Abstract =

Vallecular cyst is an uncommon but potentially dangerous condition causing stridor and has been associated with sudden airway obstruction resulting in death due to its anatomical location in neonates. It may also present with various degrees of feeding problems resulting in failure to thrive. When a vallecular cyst is suspected clinically, endoscopic laryngoscopy is necessary to visualize it. Other conditions leading to neonatal stridor such as laryngomalacia and other laryngotracheal abnormalities should be ruled out. Marsupialization with a CO_2 laser is the most effective and safest treatment to prevent recurrence. We report a case of a 1-month-old male infant with a vallecular cyst synchronous with gastroesophageal reflux, and failure to thrive. He was referred to our hospital because of hoarseness, inspiratory stridor, feeding-cyanosis, chest retraction and failure to thrive. Diagnostic workup revealed a cyst at the tongue base, suggesting a vallecular cyst. The cyst was removed by laryngomicrosurgery with CO_2 laser. After the surgery, the symptoms improved and the body weight increased steadily. We report a successfully treated case of neonatal vallecular cyst with symptoms of upper respiratory obstruction, gastroesophageal reflux, and failure to thrive. **(Korean J Pediatr 2008;51:775-779)**

Key Words : Vallecular cyst, Gastroesophageal reflux, Failure to thrive, Feeding cyanosis, Inspiratory stridor, Marsupialization

Introduction

Vallecular cyst is rare but potentially dangerous cause of stridor in neonates and infants[1-5]. When seen in adults, vallecular cyst is usually asymptomatic or there are only subtle symptoms such as voice change or a lump in the throat[4]. In contrast, cyst may lead to stridor and/or respiratory distress in neonates and young infants because of their relatively small airway and cause severe airway obstruction and even death[1].

Furthermore, it has the potential to excite the retching reflex, which may induce gagging and vomiting. These feeding difficulties lead to failure to thrive[1, 2, 5, 6].

Since vallecular cyst with failure to thrive and gastroesophageal reflux (GER) have been rarely reported in newborn infants in Korea, we describe a case with a review of the relevant literature.

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이 Case는 제 57차 소아과 추계학술대회에 지상발표 된 논문입니다.

Case Report

An 1-month-old male infant was referred to the neonatal intentive care unit of Seoul National University Children's Hospital for the evaluation of inspiratory stridor, feeding difficulty, cyanosis, suprasternal retraction and failure to thrive.

He was born at 39^{+5} weeks of gestation by vaginal delivery, weighing 2.95 kg (20th percentile) without perinatal problem at a local hospital. After oral feeding began, he started to vomit frequently. There was hoarseness when he cried, but he didn't have significant respiratory distress. On the third day of life, he was discharged without further

evaluation. After discharge, his vomiting recurred and he showed respiratory difficulty. He was brought to the emergency room of a nearby university hospital. In the emergency room, his cyanosis lasted over 5 minutes and intubation was done immediately, followed by ventilator care for 2 days in the neonatal intensive care unit (NICU). He was treated under the suspicion of congenital pneumonia for 7 days and was discharged.

Three days after discharge, respiratory difficulty and noisy breathing on feeding developed again. He was brought to the emergency room and was admitted to the NICU again. After nasal continuous positive airway pressure (CPAP) support, severe respiratory distress was relieved and nasal CPAP could be discontinued within 3 days. However, symptoms such as hoarseness, noisy breathing and feeding difficulty persisted. He was evaluated with several studies including bronchoscopy, chest computed tomography (CT) and esophagogram under the suspicion of laryngotracheobronchomalacia, but there was no abnormal finding.

Ten days after weaning from nasal CPAP, there was a sudden desaturation episode again, which was relieved by positive pressure ventilation via manual mask bagging. For evaluation of these repetitive conditions, he was transferred to our hospital under the suspicion of neuromuscular disease.

On admission, his body weight was 2.95 kg (<3rd percentile), height was 51 cm (3-10th percentile) and head circumference was 35.6 cm (10-25th percentile). His body temperature was 36.8℃, pulse rate was 166/min, respiratory rate was 37/min and blood pressure was 67/46 mmHg. Conjunctivae were not anemic, and sclerae were not icteric. The pharynx was not injected and the oral cavity was clear. Neither cervical masses nor enlarged lymph nodes were detected. The chest wall expanded symmetrically, but there was inspiratory stridor and suprasternal retraction. Heart beats were regular without any audible murmur. The abdomen was soft and flat with normoactive bowel sounds. Liver and spleen were not palpable. There was no rash or petechiae on skin exam. He was alert and both pupils were 3 mm in size with normal light reflexes. There was no abnormality on cranial nerve exam. Muscle bulk was hypovolemic but strength and power was normal. The remainders of the neurologic examination were normal.

The results of laboratory test were hemoglobin, 9.9 g/dL; white blood cell count, 7,500/uL with a normal differential cell count, and a platelet count, 517,000/uL.

The chest x-ray was normal. For further evaluation, specific examinations were repeated despite of normal findings of previous hospital data.

Esophagography showed GER. On the flexible laryngoscopic examination, an anteriorly displacing cystic mass on tongue base midline was detected. On the neck ultrasonography, 1.3×1.1×1.1 cm sized unilocular cyst was located on the mouth floor without communication to the hyoid bone (Fig. 1A). The CT film taken at the previous hospital also showed the cystic mass upon retrograde review by our radiologist (Fig. 1B).

Under general anesthesia, endoscopic laryngoscopy confirmed the cystic mass between the base of the tongue and the epiglottis (Fig. 2). After the milky content of the cyst was aspirated, the cyst wall was then removed using CO_2 laser. Histopathologic examination showed a cyst lined by squa-

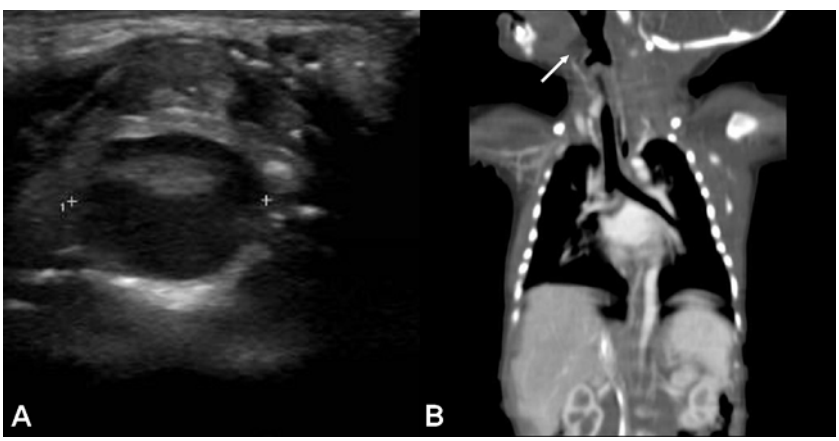

Fig. 1. (A) On ultrasonography, a 1.3×1.1×1.1 cm sized mass and unilocular cyst were located at the base of the mouth. (B) The CT film taken at the other hospital also showed the cystic mass, suggesting a vallecular cyst.

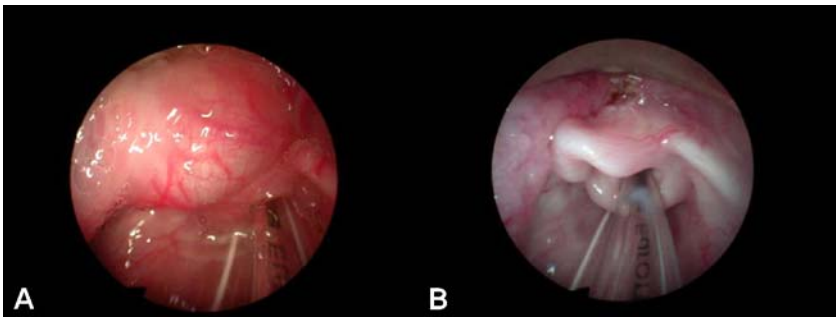

Fig. 2. Endoscopic findings of vallecular cyst. (A) Before marsupialization; the cyst deviates the epiglottis downward. (B) After marsupialization; the position of the epiglottis is recovered.

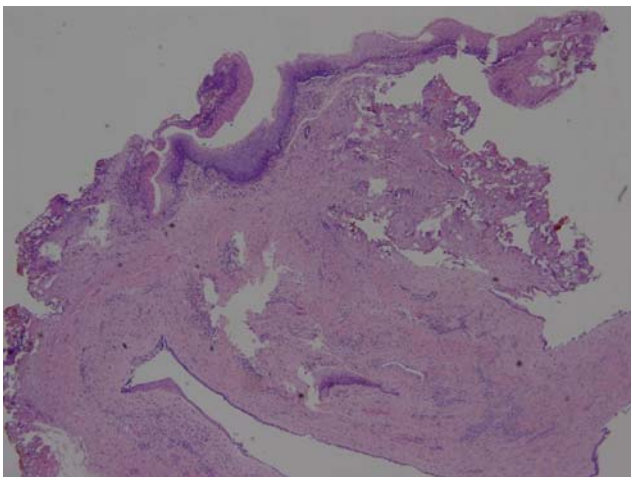

Fig. 3. The pathologic report, shows the presence of a benign cyst lined by squamous epithelium with chronic active inflammation (H&E, ×40).

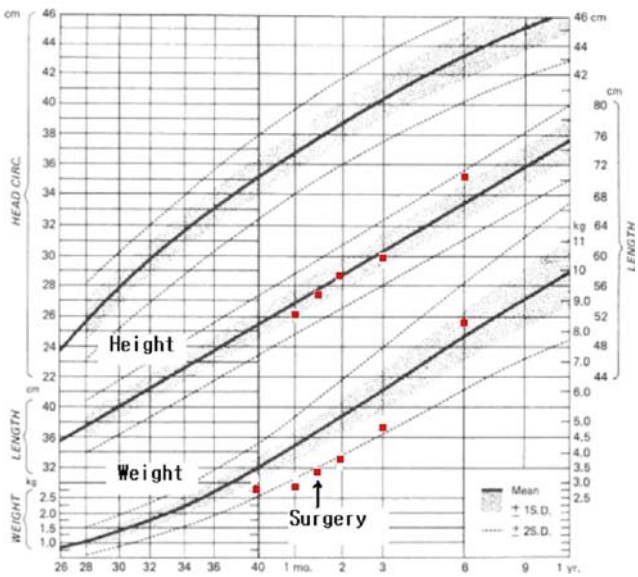

Fig. 4. Serial changes of weights relating to the 10th, 50th and 90th percentiles.

mous epithelium with chronic active inflammation (Fig. 3).

Immediately after surgery, inspiratory stridor, suprasternal retraction, and feeding cyanosis continued, but O_2 could be withheld, and feeding difficulty improved gradually.

1 week later, follow-up flexible laryngoscopy showed no abnormal finding except episodic influx of epiglottis. He was discharged and followed up in out patient clinic. He was healthy at home, had no airway and feeding problems, and gained weight steadily. Fig. 4 shows his weight and height from birth.

Discussion

Laryngeal cysts are rare in infants and children[1-17]. Aryepiglottic cyst is the most common laryngeal cyst in children, followed by vallecular cyst, ventricular cyst, and subglottic cyst[3, 17]. The presenting symptoms of vallecular cysts are usually related to upper respiratory tract obstruction. Stridor is the most common symptom of the vallecular cyst[1-5]. About 60% of children with stridor have laryngeal obstruction such as laryngomalacia, vocal cord paralysis, subglottic stenosis, hemangioma, or laryngeal cysts; 25% have lesions in the upper airway, including choanal atresia, macroglossia, facial anomalies; 15% are due to tracheobronchial lesions such as tracheomalacia or vascular compression, and others [3, 18].

In 1881, Abercrombie provided the first description of a laryngeal cyst[5, 14, 17]. In 1987, Mitchell et al. published the largest single series consisting of 20 cases experiencing of the Hospital for Sick Children, London over a 15-year period [17]. In 1980, Holinger[19] reported on 219 children presenting with stridor to two Chicago pediatric units over four years. Two of these cases (0.9%) were due to laryngeal cysts. In 1984, Wood[20] studied 225 children with stridor and found

that 4 patients had supraglottic cysts and 2 had subglottic cysts. But, there were no vallecular cysts in these two large studies of patients with upper airway anomalies.

Althouth the vallecular cyst is rare, it is a potentially life threatening condition causing sudden airway obstruction by its location[1-9]. Furthermore, 12-45% of laryngomalacia presents with synchronous airway abnormality such as laryngeal cyst[3]. Thus, in neonatal stridor, evaluation of the airway anatomy and differential diagnosis from other causes of stridor are important to prevent any mortality and morbidity from this cause.

Vallecular cysts have been reported in case files under different names, which has led to some confusion[2]. Terms used have included *mucous retension cyst*, *epiglottic cyst*, *base of the tongue cyst, congenital cyst*, and, more recently, *ductal cyst*. The name "ductal cysts" originates from the classification of DeSanto et al[10]., in which they grouped laryngeal cysts according to their location and surface mucosa. Three major categories were proposed: thyroid cartilage foraminal cysts, saccular cysts, and ductal cysts. This classification has become very popular but has the limitation that it was largely based on observation in adults, and did not recognize different anatomical sites and variations in clinical presentation[2, 11]. Making up for these defects, Newman classified laryngeal cysts as epithelial, tonsillar and oncocytic cysts (1984: Modified working classification)[11, 12].

Vallecular cyst is an unilocular cystic mass of variable size arising from the lingual surface of the epiglottis, containing clear and noninfected fluid[2]. Two major hypotheses to explain the pathogenesis of vallecular cyst are that this cyst is a consequence of either ductal obstruction of mucus glands or an embryological malformation[10]. Histologically, the cyst contains respiratory epithelium with mucous glands, with an external lining of squamous epithelium[2, 4, 13].

Most of affected infants have symptoms during the first week of life[1, 2]. Clinical manifestations consist of various degrees of upper airway obstruction such as inspiratory stridor, chest retraction, apnea, cyanosis and feeding difficulty[1-8].

Failure to thrive is an uncommon manifestation[1, 6, 14], but is reported up to 66% in one previous report[4] likewise our case. Infants with vallecular cysts may present a secondary form of laryngomalacia. This phenomenon can be explained by that altered airway dynamics caused by a progressively enlarging cyst may elevate inspiratory negative pressures, contributing to supraglottic prolapse and a secondary form of laryngomalacia[1]. Furthermore, GER is often associated to respiratory tract disease. It may result from severe pressure imbalances between the thorax and the abdomen caused by increased respiratory efforts[18].

Laryngomalacia and GER developed in infants with vallecular cysts, which explains major cause of failure to thrive. These two conditions from altered respiratory mechanics in infants with vallecular cyst interact each other, resulting in developing feeding difficulty and subsequent poor weight gain[1, 4, 5, 14]. Although, there was no anatomic laryngomalacia in our case, probably secondary functional laryngomalacia with vallecular cyst, resulted in failure to gain weight together with GER.

Flexible laryngoscopic or bronchoscopic exam is usually performed to diagnose the vallecular cysts[5-8]. On the exam, a cyst is seen on the lingual surface of the epiglottis obstructing the upper airway. When a suspected vallecular cyst is identified on the laryngoscopy, the diagnosis can be confirmed by cyst puncture and therapeutic marsupialization under general anesthesia[1, 2, 4-8]. If the mass is not localized or does not prove to be cystic, further evaluation by thyroid scan, CT or MRI should be performed. Lateral neck X-ray, neck sonogram and barium esophagogram can be helpful[2, 4]. Hemangioma, cystic hygroma, teratoma, hamartoma, dermoid cyst, lymphangioma, thyroglossal duct cyst and thyroid remnant cyst should be considered in the differential diagnosis of a vallecular mass lesion[2-4, 15-16]. Although surgical removal may be the treatment of choice[1-6], alternative modalities such as endoscopic marsupialization, excision and de-roofing have been developed recently. Marsupialization is a safe and definitive procedure, especially when performed by CO_2 laser[1-5]. Simple aspiration of the cyst is not advised because of its high recurrence rate[17].

In our case, a vallecular cyst was diagnosed in an 1-month-old infant with symptoms of upper airway obstruction accompanied with feeding cyanosis and failure to thrive. As in this case, misdiagnosis could be made unless careful laryngoscopic exam is performed. In the past, as many as 50% of vallecular cysts were discovered at autopsy[21]. Although we did not confirm the GER with 24hr pH monitoring, GER and cricopharyngeal incoordination were found with vallecular cyst on esophagogram.

We experienced a case of an 1-month-old infant with stridor, hoarseness, feeding cyanosis and failure to thrive caused by the vallecular cyst and GER through the flexible laryngoscopy and esophagography. The cyst was successfully removed with CO_2 laser in this case. Six months after

surgery, he was in perfect health without any airway problem and had gained weight.

Vallecular cysts, such as in this case, can be a disease presenting with serious symptoms and resulting in death. Therefore, if upper-airway problems are suspected, it is necessary to examine with careful laryngoscopy.

Since a case of vallecular cyst with GER and failure to thrive has been rarely reported in Korea, we report it with a review of literature.

한 글 요 약

협착음과 수유시 청색증 및 성장 장애를 보인 선천성 후두개 낭종(vallecular cyst) 1예

양미애 · 강민재 · 홍지나 · 신승한 · 김상덕
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후두개 낭종은 신생아 및 영아에서 협착음을 일으킬 수 있는 드문 질환이지만, 이 질환에 이환된 환아는 갑작스런 기도 협착을 일으켜 사망할 수도 있다. 따라서, 협착음을 보이는 환아에서 후두개 낭종을 감별해 냄으로써 갑작스런 기도 협착으로 인한 사망을 방지할 수 있다. 후두개 낭종은 협착음, 쉰목소리, 흉골 함몰, 무호흡, 수유시 청색증 및 성장 장애를 일으킬 수 있는 질환으로 이 질환이 의심되면 굴곡성 후두경으로 진단해야 한다. 이 질환의 치료는 CO_2 레이저를 이용한 후두미세수술이며, 적절한 치료를 통해 재발을 방지할 수 있다. 저자들은 협착음, 쉰목소리, 흉골 함몰, 수유시 청색증 및 성장 장애를 주소로 전원된 1개월 남아에서 굴곡성 후두경을 통해 후두개 낭종을 진단하였고, CO_2 레이저를 통한 후두미세수술 후 증상이 호전된 증례 1예를 경험하였기에 문헌 고찰과 함께 보고하는 바이다.

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