Case report

Subglottic bronchogenic cyst presenting as neonatal asphyxia—Case report and literature review

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ABSTRACT

Bronchogenic cysts are an uncommon congenital anomaly, usually found in the lungs or the mediastinum of a child or a young adult with progressive dysphagia or recurrent pulmonary infections. Its occurrence in the neck is rare, and its antenatal identification in that region has never been described. Here we report a case of a neonatal asphyxia requiring prompt intubation, which was found during bronchoscopy to be caused by a laryngeal cystic lesion. We discuss the steps of the patient’s workup and management resulting in the resection of a laryngeal bronchogenic cyst and reconstruction of the airway. The relevant literature is reviewed.

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1. Introduction

The occurrence of a bronchogenic cyst in the neck is rare, as this uncommon congenital anomaly is more often found in the lungs or in the mediastinum, and sometimes in the abdomen. The few reported cervical locations were reviewed by Ustundag [1] who pointed out that in 70% of cases, the cyst is located in the midline region, whereas in about a quarter of cases the cyst can be found in the lower lateral neck.

The clinical picture is typically that of an asymptomatic neck lump, but according to the cyst’s location and degree of aero-digestive tract compression, children can present with stridor [2], dyspnea [3] and dysphagia [4]. The radiological picture is not specific, as ultrasonography, computerized tomography (CT) and magnetic resonance (MRI) demonstrate a homogenous, fluid-filled cyst. The definitive diagnosis is made by the histological findings of bronchial elements.

There exist a few neonatal anomalies that can lead to acute respiratory distress at birth, but are compatible with life if the

airway is secured by prompt intervention, particularly if anticipated by the antenatal screening. These include bilateral choanal atresia, laryngeal webs, bilateral vocal folds paralysis, severe Pierre–Marie–Robin syndrome, large cervical masses (teratomas, lymphatic malformations) and other laryngeal or tracheal malformations. According to current publications [2], bronchogenic cysts are less likely to belong to this group of critical lesions, excepted for rare cases where lesions were confined to the thorax.

The aim of this paper is to report that a bronchogenic cyst can be located in the larynx and be the cause of a neonatal asphyxia, as well as to discuss the diagnosis and management of these specific lesions.

2. Case report

At the 39th week of gestation, a woman presented with a premature detachment of a normally positioned placenta (PDNPP). Fetal monitoring showed bradycardia and an urgent cesarean section was performed. The newborn boy was hypotonic and asphyxiated, with an Apgar score of 2–2–3 and intubation was quickly undertaken. The intubation was difficult due to undefined anomalies preventing the visualization of the vocal cords, and the passage of the tube succeeded only at the 4th forceful attempt. The child was then transferred to our institution for 72 h of fetal cooling, in order to minimize anoxic brain damage. Antenatal history revealed only polyhydramnios from an unknown cause. Chest radiography (Fig. 1) showed an air filled cavity at the lower part of the neck, suggesting a traumatic intubation. The

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otolaryngology team was asked to perform a rigid bronchoscopy. The endoscopy, using a 0°-4 mm telescope, showed a near total obstruction of the lumen by an air-filled, epithelium-lined cystic lesion on the left side of the lower sub-glottic area, continuing in the direction of the esophagus through a cartilage defect at the level of the left inferior cricoid and 1st tracheal ring (Fig. 2 a). The cyst’s wall had a perforation toward the laryngeal lumen (Fig. 2b), which we attributed to the intubation. After deflation of the cyst using a gentle pressure with the suction, the first 3 tracheal rings showed a dysmorphic appearance with a 90° angulation of the anterior part of the cartilage rather than the usual round contour, with a mild secondary stenosis. The rest of the endoscopic exam including the supra-glottic and glottic areas and the trachea down to the carina was normal. Proximal rigid esophagoscopy did not show a connection to the cyst. A nasogastric feeding tube was inserted and the patient was reintubated for further investigation.

Barium contrast (Fig. 3a) injected via the feeding tube while the tube was gradually pulled out did not demonstrate any esophageal diverticula or connection to the cyst, but showed an abnormally tortuous distal esophagus with impaired motility. A CT scan with contrast demonstrated an air collection to the left of the trachea mildly pushing it to the right, and extending in the mediastinum just above the aortic arch (Fig. 3b).

Surgical excision of the cyst was performed via a cervical approach by a joined otolaryngology and general surgery team. The cyst was dissected from the esophagus and the left lobe of the thyroid gland, with complete preservation of both structures. The dissection was made close to the structures to avoid damage to the left recurrent laryngeal nerve (RLN), which was not identified during the surgery. The cyst’s collar was dissected as close as possible to the cartilaginous defect on the left side of the cricoid and the first 2 tracheal rings before resecting it. The underlying laryngeal mucosa was preserved and sutured to the external tracheal rings to avoid bulging into the airway. The 12 mm hole in the cricoid and trachea was reconstructed with conchal cartilage externally sutured both to the surrounding cartilage and to the underlying layer of respiratory mucosa. The patient was extubated at the 8th post-operative day, after a control bronchoscopy at day 6 (Fig. 2c) showed a 3 mm area of sub mucosal inflation at the level of the reconstruction along with a 40% stenosis at the level of the reconstruction. The day after extubation, another rigid endoscopy was undertaken after an episode of dysnea and stridor. Adequate adherence between the mucosa and the wall and mild subglottic stenosis were confirmed, but suspicion of a left vocal cord paralysis was raised, which was later confirmed by a flexible endoscopy at the clinic.

During the post-operative period the baby had oral feeding difficulties, and was not capable of drinking more than 10 ml per meal, despite normal swallowing on video fluoroscopy. Sufficient intake was assured through the nasogastric tube, and weight gain was normal. A mild biphasic stridor and costal retraction persisted, but the patient was eupneic and was discharged at 6 weeks of age (1 month after surgery) in good general condition. The histological exam showed a cyst lined by ciliated respiratory mucosa with a submucosa containing mucous glands, elements of smooth muscle and islets of hyaline cartilage, all compatible with a bronchogenic cyst (Figs. 4–6).

The patient was last seen 10 months postoperatively. He was hospitalized for severe viral laryngitis the preceding month, which resolved with medical treatment. He no longer has stridor or chest retractions, but with significant swallowing difficulties do persist. His left vocal cord now shows partial motion at flexible endoscopy.

3. Discussion

The occurrence of a bronchogenic cyst at the level of the larynx is rare, and to our knowledge, this is the first case report of such a cyst as a cause of asphyxia at birth. Lazar et al. described in 1991 a case [2] of a stridor and dyspnea in a neonate necessitating intubation and thoracotomy for resection of a mediastinal bronchogenic cyst deviating the carina to the right and the esophagus to the left. Goswamy et al. [5] have also reported on a cervico-thoracic bronchogenic cyst causing stridor in a 6 month-old child, due to compression of the trachea down to the level of the carina. In our case, the presumed mechanism of obstruction is related to the pressure applied by that fluid-filled cyst on the laryngeal mucosa through a lack of cricoid and tracheal cartilage, possibly protruding between the vocal cords. The cyst’s wall is believed to have ruptured during intubation, allowing the fluid to drain, and the child to be intubated and ventilated.

The presence of the cyst was not suspected before birth, although a prenatal detection of a bronchogenic cyst, at least in the chest, is possible as early as in the 15–19th week of gestation [6,7]. The sonographic appearance is that of a single, unilocular, echo-free cystic structure with thin walls [8,9], while on MRI the cyst appears homogenous and hyper intense on T2. A judicious use of the prenatal information was described by Levine et al. [7] who performed an extra uterine intrapartum (EXIT) bronchoscopy, CT scan and eventually thoracotomy for the resection of a bronchogenic cyst at the level of the carina at the 33rd week of gestation.

The left (ipsilateral to the cyst) vocal cord’s mobility was found to be impaired in our patient, and lacking a pre-operative assessment, a damage to the RLN during the resection and...
reconstruction cannot be ruled out. Still, the cyst was located at the level of insertion of the RLN to the larynx and could have, in theory, mechanically interfered with its embryological attachment. A common developmental anomaly is less likely since the RLN originates from the 6th pharyngeal arch, whereas the cricoid from the 4th arch and the trachea and possibly the cyst from the respiratory primordium. Similarly, Teissier et al. [10], who have published the largest series of cervical bronchogenic cysts,
Among the different diagnoses considered after the initial rigid endoscopy were an unusual tracheo-esophageal fistula or esophageal diverticulum, but the barium feed clearly showed the lack of connection to the esophagus. Hence, it seemed more reasonable to suspect either a tracheocele or a fluid-filled cyst that burst. The differential diagnosis of such a congenital cyst includes a thyroglossal duct cyst and a 2nd or 3rd branchial cleft cyst, although a tract in continuity with the tongue/thyroid or with the tonsillar fossa/piriform sinus would be expected and an imbedding in the larynx or upper trachea is not typical. Other types of malformations to consider are laryngeal cysts such as saccular cysts, laryngoeceles or duc tal cysts (mucous retention), but these are not reported to cross through the cartilage, but rather stay endolaryngeal or course via the thyro-hyoid membrane [11,12]. Regarding the tracheal involvement, tracheal or bronchogenic cysts are also possible, since in addition to their more typical extra-luminal location, trans-mural cysts have been described in the mid-trachea [3] or larynx [10]. Generally, a CT scan of a bronchogenic cyst is not specific and shows a unilocular homogenous cyst with a density varying between fluid and soft tissue [10,13]. In our case the images of an air-filled cavity did not help in establishing the diagnosis, and only provided anatomical information to exclude other organ involvement. The final diagnosis, determined histologically, was based on the presence of a well-defined wall containing hyaline cartilage, smooth muscle and mucinous glands underneath a ciliated respiratory epithelium (Figs. 4-6). Cartilage and muscle are not seen within the wall of a tracheocele.

A surgical resection of the cyst had to be performed in order to remove the airway obstruction and extubate the patient. The idea to use an alternative approach, i.e. an endoscopic marsupialisation was rejected for two main reasons. First, there was a risk of an eventual formation of a mucus-accumulating pocket open to the airway. Also, a fistular tract to the neck could not be excluded. The cervical approach allowed a safe dissection from the esophagus, and was convenient for the airway reconstruction with a conchal cartilage. This thin and malleable cartilage is particularly suitable for small tracheal defects. Reviewing the literature regarding the treatment of a bronchogenic cyst, we observe that all authors recommend a surgical resection in order to avoid future complications. In cases of a mediastinal or pulmonary parenchymal lesions the bronchogenic cyst can cause recurrent infections and bronchiectasis, while in extra-thoracic location the arguments in favor of a resection include the risk of malignant transformation [14,15] and of progressive growth [1].

The paucity of such cases makes it difficult to establish strong recommendations for future cases. Nevertheless, we suggest that: the diagnosis of a bronchogenic cyst should be considered among the differential diagnosis of a congenital laryngeal cyst; an MRI is helpful in defining the location and extensions; a rigid endoscopy provides precious information for planning the definitive resection; the latter should take into consideration the need for an airway reconstruction.

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**References**


