Vallecular cysts: a differential diagnosis to consider for neonatal stridor and failure to thrive

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Vallecular cysts are an important differential diagnosis in neonates and infants who present with stridor, respiratory distress and failure to thrive.

DECLARATIONS

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Patient presentation

Case 1

Patient 1 was born via lower segment caesarean section (LSCS) and weighed 3.254 kg. Even though soft inspiratory stridor had been present since birth, she was able to be discharged home on breast feeds. She was seen by her community paediatrician at four weeks of age for poor weight gain and was encouraged to increase the frequency of her feeds. At six weeks of age she presented to the emergency department with mild to moderate respiratory distress, increasing stridor and failure to thrive. She was breast feeding regularly, had wet nappies with normal stools but was noted to have frequent small posits. On examination in the emergency department she had a low pitched stridor, tachypnoea with use of accessory muscles, dry mucous membranes and weighed less than her birth weight (3.0 kg).

She was admitted under the general paediatricians and given supportive management including oxygen and intravenous fluids. During this period she was noted to have several brief apnoeic episodes with desaturations to 74%. This was associated with increased upper airway secretions requiring intermittent pharyngeal suctioning. She was tolerating her feeds however her weight gain remained poor. She continued to have increased work of breathing despite supportive therapy.

She was investigated with a barium swallow and follow through which demonstrated a normal oesophagus and pylorus, prompt gastric emptying, no episodes of gastro-oesophageal reflux and no evidence of malrotation. A dynamic magnified airway screen and abdominal ultrasound US showed no airway collapse to suggest laryngomalacia or tracheomalacia. There were no sonographic features of hypertrophic pyloric stenosis identified on abdominal ultrasound.

Otolaryngology (ENT) consultation was requested on Day 4 of her admission and a formal laryngo-broncho-oesophagoscopy under general anaesthesia was arranged. She was induced with a spontaneous inhalational technique. Of concern, the larynx could not be easily identified on initial laryngoscopy by the anaesthetist due to a large mass (Figure 1) in the tongue base obscuring the view. The ENT surgeon was required to emergently intubate using a fibreoptic telescope. Fortunately the lesion was soft and the endotracheal tube was able to be passed posterior to the mass. The lesion was identified as a *vallecular cyst*, which was initially decompressed with a needle and subsequently marsupialized during the same operation. The airway structures became clearly apparent only after decompression (Figure 2). Postoperatively her work of breathing and stridor improved and she had no further desaturation/apnoeic episodes. Histopathology confirmed a cyst lined by thin squamous epithelium consistent in appearance with a vallecular cyst.

Her growth chart showed significant improvement commencing immediately after her surgery and remarkably she crossed several centiles within a few weeks. At follow-up in the outpatient clinic 11 months postoperatively, she remained well with no recurrent stridor.

Figure 1
Endoscopic view of the larynx prior to intubation showing obstructing cystic lesion arising from the vallecula

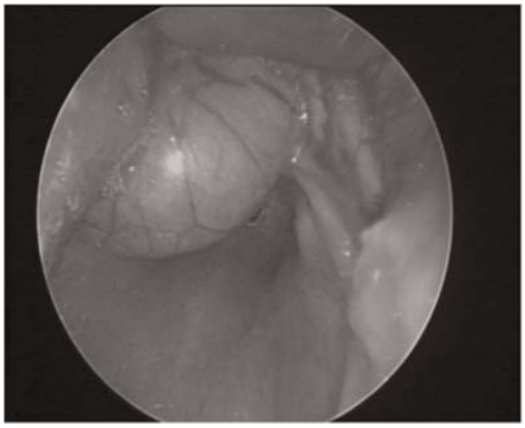
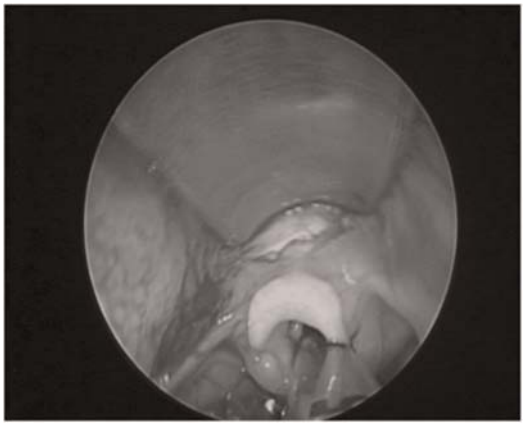


Figure 2
Endoscopic view of the larynx after decompression of the vallecular cyst revealing normal epiglottis and left posterior vocal cord



Case 2

Patient 2, a dizygotic twin, was born at 35 + 2 gestation via LSCS. She remained in hospital for 10 days for feeding difficulty, irritability and regurgitation postfeeding. After her discharge she was seen by her paediatrician for possible gastro-oesophageal reflux and commenced on ranitidine. Her twin remained well throughout.

At seven weeks of age she presented with an apnoeic and cyanotic episode that her parents described occurring 15–20 min after feeding. Her septic screen was negative and empirical antibiotics were ceased. While on the ward, she was observed to have soft airway noise and intermittent desaturations at the time of feeding. After a prolonged apnoeic episode she required bag and mask ventilation and was transferred to the intensive care unit.

In the intensive care unit she had multiple investigations including barium swallow, head ultrasound, electro-encephalography and transthoracic echocardiogram – all of which were unremarkable. The ENT team were asked to review her and a laryngo-broncho-oesophagoscopy was performed on day six of her admission. A midline vallecular cyst that was not as obstructing as Case 1 was identified, causing posterior displacement of the epiglottis. Thin serous fluid was aspirated from the cyst before formal marsupialization. Her trachea, bronchi and oesophagus were normal.

Following the marsupialization of the vallecular cyst, she made excellent progress with no further desaturations. She was alert, feeding and saturating well on room air, and was able to be discharged on the second postoperative day. Histopathology of the cyst demonstrated mildly chronically inflamed non-keratinizing squamous mucosa and small minor salivary glands in the submucosa. Cytology of the aspirated fluid consisted of degenerate histiocytes and cystic debris. No malignant cells were identified. She remained asymptomatic and thriving on follow-up at six months.

Discussion

Vallecular cysts are a form of supraglottic cyst accounting for 10.5–20.1% of all laryngeal cysts.[1] They are typically benign, unilocular cystic masses of variable size that arise from the lingual surface of the epiglottis and usually contain clear, non-infected serous fluid. Histologically, the cyst has an external lining of squamous epithelium and may contain respiratory epithelium with mucous glands.[2] The cause of vallecular cysts are thought to be due to either an embryological malformation or ductal obstruction of a mucous gland.[1] Other cystic lesions to consider in this site include a lingual thyroid, thyroglossal duct cyst, lymphatic malformation and cystic tumours such as teratomas.

Patients with vallecular cysts can present with feeding difficulty, failure to thrive, stridor and cyanotic episodes.[3,4] Infrequently patients present with sudden upper airway (supraglottic) obstruction, which can result in death.[2] Stridor is a relatively common symptom during the neonatal period with laryngomalacia being the most probable cause of *inspiratory* stridor.[4] However, inspiratory stridor is also the most common presentation of vallecular cysts. To complicate the clinical picture, laryngomalacia often coexists with vallecular cysts as the cyst itself can compress the adjacent epiglottic cartilage and cause supraglottic prolapse.[3,4] Interestingly, the airway noise heard in Case 1 was present from birth and had a low pitched quality. To an experienced ear, this may have raised a suspicion of an alternate diagnosis, as laryngomalacia is classically high pitched and has a later onset.

Diagnosis of vallecular cyst requires a high index of clinical suspicion. Antenatal diagnosis via magnetic resonance imaging and ultrasound has been reported;[1] however this was an incidental finding and would not be routinely recommended. Where a vallecular cyst or other mass airway lesion is suspected in infancy, a lateral airways X-ray may show an alteration in the airway contour. Definitive diagnosis is made by inspection of the tongue base with direct laryngoscopic examination. This can be achieved with a flexible nasendoscope or under general anaesthesia with rigid instrumentation. These investigations are straightforward and do not require unnecessary exposure to radiation.

As illustrated in Case 1, it can be challenging to manage the airway in a patient with a vallecular cyst, even for experienced anaesthetic and surgical staff. Assessment is best performed in a controlled environment as rapid intervention may be required. The laryngeal anatomy can be distorted making endotracheal intubation near impossible and the need for a surgical airway likely. The rate of complications in an emergency infant tracheostomy is potentially high as the small and easily compressed structures may make anatomical landmarks difficult to identify. If either of our patients presented had deteriorated in the ward setting and required intubation by a less experienced clinician, an adverse outcome might have ensued.

Surgical decompression is the treatment of choice for vallecular cysts and can be done at the same setting as the diagnostic study in the operating room. This treatment, however, may not be appropriate in certain centres or without commitment to a high-dependency unit, therefore Specialist Hospital transfer may be required. Marsupialization rather than total excision is usually sufficient and results in minimal long-term sequelae.[3–5] Simple aspiration of the vallecular cyst however has a high chance of recurrence.

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