Vallecular Cysts as a Rare Cause of Failure to Thrive with Obstructive Breathing in Infants

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Keywords

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Abstract

A vallecular cyst is a rare cause of upper airway obstruction in neonates and infants. Symptoms include stridor, failure to thrive, obstructive breathing and respiratory distress. We present a case of a 2-month-old infant with poor weight gain and feeding difficulties. The diagnosis was initially missed due to a concomitant viral infection. The patient underwent transoral marsupialisation with complete resolution of symptoms. Vallecular cysts should be included in the differential diagnosis of children with stridor and dysphagia. Diagnosis is primarily by laryngoscopy. The preferred treatment is marsupialisation, which is less invasive than complete excision and has a low risk of recurrence.

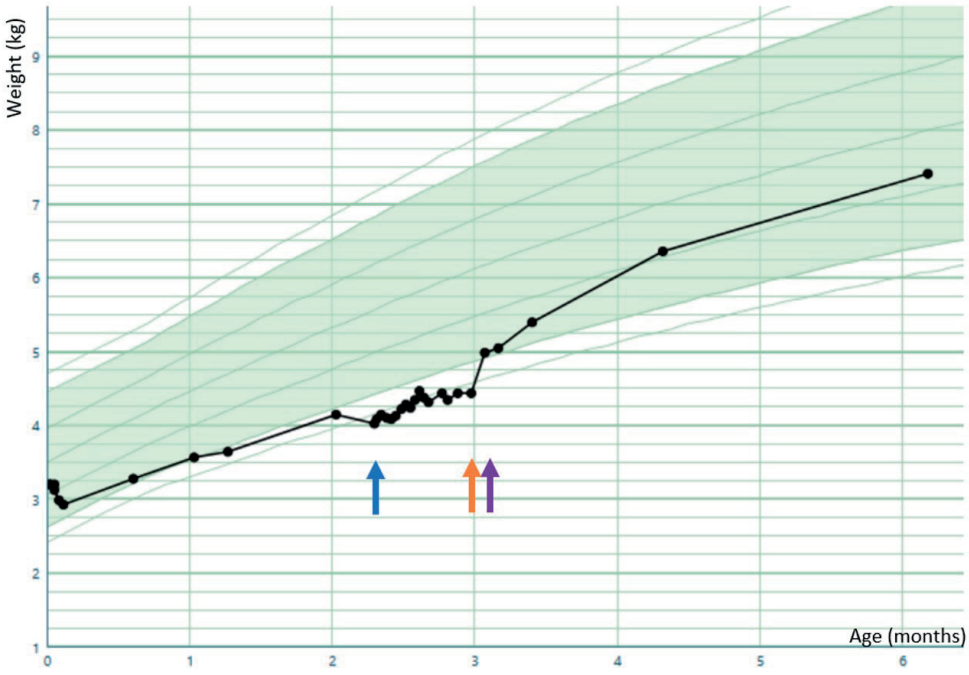
Introduction

A vallecular cyst is a unilocular cyst on the lingual surface of the epiglottis containing clear, non-infected fluid (1). Other terms used to describe the same lesion include mucus retention cyst, epiglottic cyst, base of tongue cyst and ductal cyst (2).They are thought to arise from obstructed ducts of submucosal glands in the vallecula (3). It is a rare cause of upper airway obstruction in neonates and infants. A vallecular cyst can cause stridor, dysphagia, failure to thrive, obstructive breathing, respiratory distress and, in rare cases, life-threatening upper airway obstruction (1, 4). The median age at diagnosis is one to two months, with a range of birth to 11 years reported in the paediatric literature (1, 4-7).The uncommon nature and often initially mild and non-specific symptoms often lead to delayed and misdiagnosis. We describe the case of an infant in whom the diagnosis was initially missed because of a concomitant viral infection thought to be responsible for the respiratory symptoms. Informed consent was obtained from the parents to publish the case.

Clinical report

A 2-month-old male infant presented to the paediatric outpatient clinic with poor weight gain and feeding difficulties since one month of age. The symptoms were previously attributed to cow's milk protein intolerance, for which a maternal diet was started. However, there was no improvement and the weight decreased from the standard deviation of -1 to -2 (Figure 1). He continued to drink very slowly and cried during feeding. When he returned to the outpatient clinic, he had a rhinitis and mild cough for a few days, with the appearance of diarrhoea. The mother had never reported any respiratory problems before. Clinically, we saw a thin, pale infant with signs of dehydration. He had nasal obstruction but no respiratory distress. He was admitted to hospital for additional tube feeding and oral rehydration solution to replace losses due to diarrhoea. A diagnosis of rhinovirus infection was made.

Figure 1: Growth chart.
Blue arrow: beginning of hospitalization, orange arrow: evacuation puncture, purple arrow: marsupialisation



However, he did not improve during hospitalisation but developed an obstructive breathing pattern with stridor. This was attributed to nasal congestion and viral laryngitis. Nasal decongestants and budesonide aerosols were started. However, the obstructive breathing worsened and weight gain remained suboptimal despite additional tube feeding. Episodes of desaturation with cyanosis were noted during drinking and deep sleep. Dexamethasone was given orally without improvement of stridor. The enteral feed was changed to an amino acid formula because of a suspected severe cow's milk protein intolerance (due to crying, convulsions, suboptimal weight gain and mild persistent diarrhoea). Additional investigations were carried out to search for an anatomical cause of the obstructive breathing.

Figure 2: Endoscopic view of the vallecular cyst during diagnostic laryngoscopy.

a: posterior displacement of the epiglottis (note: a nasogastric tube is in place), **b:** image of the cyst immediately before aspiration, **c:** image of the cyst at the end of the aspiration.

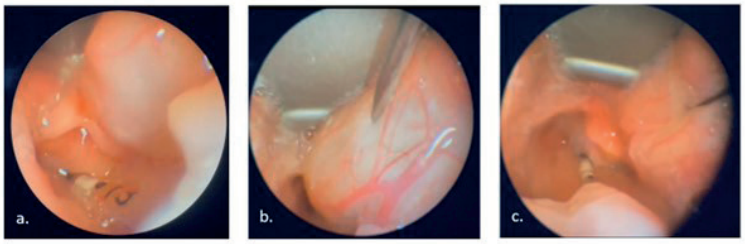
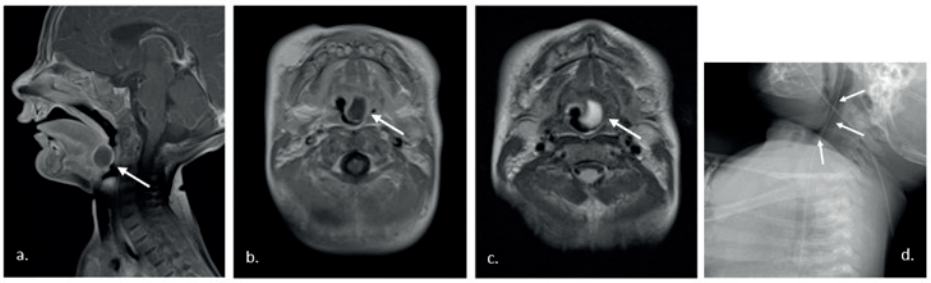


Figure 3: Radiologic findings

a-c: Magnetic resonance images (a : sagittal T1, b : coronal T1, c : coronal T2) showing a cystic mass at the left vallecula.
d: Lateral radiography showing a radiopaque mass in the laryngeal region.



Cardiac ultrasound showed no evidence of a vascular ring or arteria lusoria. An initial flexible laryngoscopy failed to show any evidence of laryngomalacia or laryngeal mass due to the presence of a nasogastric tube and poor cooperation. A lateral radiograph and barium oesophagography were reported as normal. Direct laryngoscopy under general anaesthesia revealed a cystic mass at the base of the tongue. A diagnostic evacuating puncture was performed with aspiration of 5 mL of clear fluid (Figure 2). Further radiological assessment by magnetic resonance imaging (MRI) was deemed necessary prior to removal of the cyst. Immediately after the evacuating puncture, an improvement in feeding technique and respiratory pattern was noted. The MRI showed a retention cyst of the left vallecula measuring 1.3-0.9m with a 50% reduction of the oropharyngeal surface (Figure 3 a-c). The thyroid gland had a normal appearance, so thyroid function tests were not performed. In retrospect, the cyst was already visible on the lateral radiograph (Figure 2d). Transoral marsupialisation was performed using a carbon dioxide (CO2) laser. Histopathology revealed a cyst lined by a non-keratinising stratified squamous epithelium. The patient left the hospital the following day with complete resolution of symptoms. Good weight gain was subsequently noted. Three months after the procedure, there has been no recurrence.

Discussion

A vallecular cyst is a rare cause of neonatal stridor, found in only 0.9-2% of infants with this presentation (6). Therefore, the condition is not always well known among paediatricians and ENT specialists and may be overlooked. It should be suspected when stridor is associated with failure to thrive, dysphagia and/or obstructive airway. Flexible laryngoscopy is the preferred primary screening technique in infants with stridor. The importance of good visualisation of the base of the tongue during the procedure should be emphasised. Often it is not possible to obtain a clear and reliable assessment with flexible endoscopy in infants. In this case, direct laryngoscopy under general anaesthesia should be performed to differentiate between laryngomalacia, subglottic stenosis, vocal cord paralysis, a laryngeal mass or a laryngeal web (6, 8). If the image is strongly suggestive of a vallecular cyst (a unilocular cystic mass on the lingual surface of the epiglottis containing clear fluid), it has been recommended to proceed immediately to direct laryngoscopy under general anaesthesia. The diagnosis can be confirmed by cyst puncture (1). The distinction between a solid and a cystic mass is not always immediately apparent on laryngoscopy and further investigations are required to differentiate between a thyroglossal duct cyst, dermoid cyst, adipose tumour, lymphangioma, haemangioma, lingual thyroid or teratoma before proceeding with therapy (1, 6).

Several investigations may contribute to the diagnosis. MRI is the preferred diagnostic modality, as it has the best ability to differentiate between the various laryngeal masses. However, it is sometimes difficult to obtain in a short period of time. Ultrasound is more readily available and can also differentiate well between cystic and solid masses. It can also confirm normal thyroid anatomy. However, it is important to remember that this examination is more operator dependent. The cystic mass can sometimes be seen on lateral radiographs, but these are difficult to interpret and do not allow differentiation between the different types of mass. Computed tomography cannot properly differentiate between a vallecular cyst, thyroglossal duct cyst, dermoid cyst, haemangioma and lymphangioma, all of which have a similar low-density appearance (6). Thyroid scanning should only be performed if the mass has a solid appearance and a normal thyroid position cannot be confirmed.

Associated conditions are found in a significant number of patients and should be considered in management strategies. In a retrospective chart review, 68/156 (43,6%) children with a vallecular cyst had concomitant laryngomalacia and 9/156 (5,8%) had gastroesophageal reflux (4). Another review found even higher figures, with two thirds of the children also having laryngomalacia, and one third having gastroesophageal reflux (7). Infants with laryngomalacia were younger at symptom onset, had more severe symptoms and were more likely to have residual symptoms after treatment. They were also more likely to be admitted to intensive care and to need mechanical ventilation (7).

There are several options for the surgical treatment of vallecular cysts, including needle aspiration, marsupialisation (deroofing) and complete excision. Simple needle aspiration has a high recurrence rate and is therefore not the preferred treatment (4).Marsupialisation is generally the preferred initial treatment option as it is less invasive than complete excision and has a low risk of recurrence. Marsupialisation can be performed using micro-instruments, electrocautery, coblation, KTP laser or CO2 laser. During the procedure, after marsupialisation, the larynx and epiglottic cartilage should be reassessed to diagnose concomitant laryngomalacia (4). In a retrospective study of 156 patients with vallecular cysts, only two patients had recurrence after marsupialisation (1.2%). Both children were over a year old. In older children, a thicker cyst wall

and more viscous cyst contents were observed, which may explain this increased risk of recurrence (4). Another study reported a recurrence rate of up to 15% (3/20) after marsupialisation, of which two patients required revision marsupialisation. The third patient was closely followed and showed no symptoms (3).In the event of recurrence, complete excision of the cyst can be performed transorally with a CO2 laser. In children with severe laryngomalacia (type I and II only) it has been suggested that supraglottoplasty be performed at the same time to improve the outcome of the operation (4).

Definitive diagnosis is made by pathological evaluation. Histopathology of the resected specimen shows an internal mucosal lining of non-keratinising squamous or respiratory epithelium and a fibrous wall (3).

Conclusion

A vallecular cyst is a rare cause of upper airway obstruction in neonates and infants. Clinical symptoms are similar to those of laryngomalacia, including stridor, failure to thrive, obstructive breathing and respiratory distress. Flexible laryngoscopy is the preferred primary screening technique, but has a high risk of missing the diagnosis. Therefore, direct laryngoscopy must be performed if the base of the tongue and larynx cannot be properly assessed. Several investigations can contribute to the diagnosis, with MRI being preferred as it has the best ability to differentiate between the various laryngeal masses. Many surgical treatments have been described, including needle aspiration, marsupialisation (deroofing) and complete excision. The preferred treatment is marsupialisation, which is less invasive than total excision and has a low risk of recurrence.

Conflicts of interest

None of the authors have a conflict of interest to disclose. No funding was received for this paper.

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