Varied Clinical Presentation and Management of Pediatric Vallecular Cyst

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Abstract

The aim of this article is to highlight the varied presentation and management of vallecular cysts. Three children with diagnosis of vallecular cyst presented in our department were reviewed retrospectively for clinical presentation, diagnostic tools and treatment options. All three cases presented with respiratory and feeding difficulty. Diagnosis in all the three cases was made with laryngoscopy and imaging. All three patients were treated by transoral approach aimed for excision of cyst using cold instruments. Postoperative period was uneventful and no recurrences were observed on the long-term follow up. Vallecular cysts should be considered as one of the differentials in children with respiratory distress and dysphagia in spite of being a rare anomaly. Direct transoral approach for excision of the vallecular cyst is recommended as a safe and reliable method with no recurrences till date.

Keywords: Vallecular Cyst; Respiratory Distress; Dysphagia; Laryngoscopy

Introduction

Vallecular cysts are a unilocular cystic mass which contain clear and non-infected fluid. Vallecular cysts are rare anomalies and are presented with respiratory distress, dysphagia and failure to thrive and can cause potentially life-threatening condition causing sudden airway obstruction. Presentation with symptoms like chest retraction, apnoeic/cyanotic, choking, postprandial vomiting, coughing, and hoarse cry also seen occasionally.
Any infant presenting with such symptoms should be considered to have laryngomalacia, upper airway anatomical anomalies, or upper airway obstructive lesions like hemangioma, cystic hygroma, teratoma, hamartoma, dermoid cyst, thyroglossal duct cyst or thyroid remnant.  

Nasopharyngolaryngoscopy and imaging are required to confirm the diagnosis and to see the extension of cyst. Treatment options include cyst aspiration, marsupialization, deroofing of cyst wall and complete excision.

Here, three cases of vallecular cysts in pediatric group are shared and their presentation and management is described.

**Case One**

A 4-year-old boy presented in our ENT clinic with progressive deglutition problems, intermittent odynophagia and persistent dry cough for the last one year. Oral examination was normal. Flexible naso-pharyngo-laryngoscopy showed a pink pale coloured swelling over the lingual surface of epiglottis [Figure 1A]. Contrast enhanced Computed Tomographic scan (CECT) showed ovoid non enhancing cystic lesion at the level of epiglottis [Figure 2].

Endoscopically assisted cyst excision was performed via trans-oral route under general anaesthesia, using cold instrument and electrical cautery [Figure 1B]. Histopathological examination [Figure 1C] revealed cyst lined by pseudostratified columnar epithelium with dense lymphocytic infiltrate within the sub-epithelium. Analgesics, antibiotics and dexamethasone were administered in postoperative period and the patient was discharged after 72 hours. No sign of local recurrence was seen after one year follow-up.

**Case Two**

An 11-month-old male infant presented to our hospital with respiratory distress, choking spells and frequent regurgitation of feeds. The child had noisy breathing for one month of age and keeping laryngomalacia in mind, conservative management was planned in the primary hospital. The patient was doing well until 8 months of age. During examination, child got agitated and again had an apnoeic spell requiring resuscitation and was subsequently intubated. Urgent CT [Figure 3] was done to evaluate upper airway and a cystic mass 17 mm × 8.9 mm was seen embedded in the base of tongue. The patient was shifted to OT, general anaesthesia given, direct laryngoscopy was performed and large cystic swelling was found at
the base of tongue. Molar retractor was positioned and retraction sutures were placed to retract the tongue. Deroofing of cyst wall done and remnant of cyst wall was cauterized. The patient was extubated and observed overnight in the intensive care unit (ICU). Histopathological examination confirmed diagnosis of benign cystic lesion. Child recovered well with no recurrence at two-year follow-up.

Case Three
A full term three-day-old neonate was referred from Neonatal ICU with a history of inspiratory stridor, choking and difficulty in feeding since birth. On examination, the neonate was in respiratory distress. The patient was kept on continuous positive airway pressure (CPAP) and maintained saturation above 95%. Oral examination was normal. Gentle laryngoscopic examination showed huge vallecular cyst obscuring the endo-larynx. Urgent CT confirmed the extension and origin of the cyst [Figure 4A].

The patient was taken to OT but endo-larynx was difficult to visualize with awake fibreoptic intubation due to presence of large cyst and anteriorly placed larynx. 2ml of fluid was then aspirated from the cyst which aided in successful intubation. Surgery was planned via transoral route. Tongue retracted with sutures (silk 2-0) and Hopkins 30-degree endoscope was used. Cyst was grasped with forceps and excised [Figure 4B]. Complete excision could not be ascertained due to the attachment of cyst to lateral wall of vallecula and inadequate exposure. The base of cyst was cauterized and wound left to heal by secondary intention. The child was shifted back to ICU and observed overnight. Antibiotics, dexamethasone and histamine 2 blockers were given in preoperative period. The very next day the child was extubated and significant improvement of respiratory distress was observed. Cyst with columnar epithelial lining was confirmed on biopsy. The Child is remained on follow up for the last 10 months and planned for periodic laryngoscopic examination to rule out any recurrence.

Discussion
In 1881, Abercrombie described vallecular cyst. Vallecular cysts have been given various names such as congenital and ductal cyst, mucus retention cyst, epiglottic cyst and base of tongue cyst.
The incidence of laryngeal cysts is approximately 5% of all benign laryngeal lesions and vallecular cyst makes 10.5% of all congenital laryngeal cyst. Thus, vallecular cyst is a rare entity. Laryngeal cysts are classified into ductal, saccular and thyroid foraminal subtypes by De Santo. Ductal cysts are most common and are frequently found in vallecula and the true vocal cords. Later, classification based on histology and location described cysts as congenital, retention, inclusion and lymphoepithelial cysts.

Vallecular cysts can be found at any age, but their presentation differs according to age and size. They are usually asymptomatic when small in size; however, with increase in size or when infected, they may cause respiratory, feeding and speech difficulties. Inspiratory stridor and dyspnea are most commonly seen after birth and failure to thrive with feeding difficulties is more commonly noted among older children. Symptoms like chest retraction, apneic/cyanotic, choking, postprandial vomiting, coughing, and hoarse cry are also seen occasionally. Vallecular cysts may cause supraglottic obstruction due to mass effect and more importantly by displacement of the epiglottis posteriorly and inferiorly which may present as episodes of apnea and cyanosis. Increased size progressively interferes with swallowing, subsequently leading to poor weight gain and failure to thrive.

Due to the anatomical location of the cyst and small airway in infants, higher risk of sudden airway obstruction is there. Around 12–45% of patients with laryngomalacia may have coexisting airway abnormality such as laryngeal cyst. Subsequent increase in inspiratory negative pressure leads to supraglottic prolapse and development of laryngomalacia. Hence, vallecular cyst should be kept in the differential diagnosis of congenital stridor in infants along with laryngomalacia, vocal cord palsy and subglottic hemangioma.

Three cases of pediatric vallecular cyst with varied presentation were presented above. One newborn presented with inspiratory stridor and failure to thrive, one child was on conservative treatment for laryngomalacia and a toddler with chief complaints of dysphagia. The combination of detailed history, radiological and endoscopic assessment provides information necessary to confirm the diagnosis. Lateral radiography, flexible nasopharyngolaryngoscopy, direct laryngoscopy, computed tomography (CT), magnetic
resonance imaging (MRI) and ultrasonography (USG) of the neck, are helpful tools in the assessment of vallecular cyst.³

Flexible laryngoscopy is an initial screening tool. Direct laryngoscopy is used in securing the airway, detection of other lesions and surgical management.³ Contrast enhanced CT or MRI are essential to find out the size, site, anatomic dimensions and extension of a cyst to plan for surgical resection. MR is diagnostic modality of choice but is time consuming, requires sedation and costly, due to which it is not widely used in children with compromised airway.¹⁵ We performed Laryngoscopy (flexible/ direct) in all of our three patients to confirm the diagnosis and contrast enhanced CT before planning surgery to determine the dimensions and location of the cyst. Prenatal diagnosis of this condition is feasible with ultrasound scanning or fetal MRI which helps surgeons to secure airway during birth.³

A limited number of case reports and series of vallecular cysts have been published stating different treatment modalities. Surgical options for vallecular cyst in infants and children include cyst aspiration, marsupialisation (i.e. deroofing) and extirpation (i.e. resection and excision).¹¹ However, aspiration of cyst is not advocated for its high recurrence rate.⁸ Hence, aspiration should be either considered as a palliative procedure, or as an initial step in cases of difficult intubation for decompression of the cyst.⁸,¹¹ Definitive treatment of vallecular cyst is marsupialisation or extirpation.⁴⁻⁸ In retrospective analysis, Li found successful outcomes from marsupialisation using micro-instruments, electrocautery, KTP laser and micro-debrider at 15% recurrence rate.³ However, Gutierrez et al., reported a success rate of 87.5% with marsupialisation using cup forceps and CO² laser.¹

The use of CO² laser and micro-debrider are recommended because they are less invasive and provide fast healing. Hsieh et al. used CO² laser in 33 patients for treating vallecular cyst with successful outcomes and no recurrence.¹⁶

Vallecular cyst may be approached often via direct transoral route or using suspension laryngoscopy. Very rarely external approach is required for extirpation of recurrent lesions and large cysts.⁸

In this article, a simple, direct transoral approach for managing vallecular cyst without any specialized equipment is described. This approach is safe in infants and has minimal recurrence rate. Chen found no recurrence in seven patients who underwent complete cyst
excision via transoral surgical approach. Similarly, we have not encountered any recurrence so far with a follow-up period from 10 months up to 2 years.

In the given series, oral fiberoptic intubation was done in Case One and Case Two was intubated in emergency. In Case Three, fiberoptic intubation was difficult, so the cyst was aspirated with large bore needle, following which the child was intubated orally. However, in few cases tracheostomy may be needed to secure airway.

**Conclusion**

Vallecular cysts are rare and the challenge in making an early diagnosis keeping laryngomalacia in mind has been ascertained in our case series. Each case requires an individualized approach. Direct transoral approach for excision of the vallecular cyst is recommended as a safe and reliable method with no recurrences till date. Further randomized studies are needed to determine the best diagnostic and treatment options for this entity.

**References**

Figure 1A: Flexible nasopharyngolaryngoscopy for a 4-year-old boy presented in our ENT clinic with progressive deglutition problems, intermittent odynophagia and persistent dry cough for the last one year, showing a pink pale coloured swelling over the lingual surface of epiglottis. **Figure 1B:** Post-op picture of cyst excised with cold instrument and base cauterized. **Figure 1C:** Microscopic section showing a cyst lined by stratified squamous epithelium with cyst wall showing mucous glands. No nuclear atypia seen.

Figure 2: Contrast enhanced CT scan of Case One (a 4-year-old boy presented in our ENT clinic with progressive deglutition problems, intermittent odynophagia and persistent dry cough for the last one year) revealed ovoid cystic mass (star 5 point) at level of base of tongue and epiglottis. **Figure 2A:** Lateral cut showing minimal narrowing of airway. **Figure 2B:** Axial cut showing cyst on the lingual surface of the epiglottis.
**Figure 3:** CT images of Case Two, an 11-month-old male infant presented to the hospital with respiratory distress, choking spells and frequent regurgitation of feeds. **Figure 3A:** Axial CT image showing a cystic ovoid mass measuring 17mm × 8.9 mm, embedded in the base of the tongue. **Figure 3B:** Lateral CT cuts reveal compromised airway due to tongue base cystic lesion.

**Figure 4:** Images of Case Three, a full term 3-day-old neonate who was referred from Neonatal ICU with a history of inspiratory stridor, choking and difficulty in feeding since birth. **Figure 4A:** CT cut showing ovoid non enhancing cyst attached to right lateral wall of vallecula, causing airway narrowing. **Figure 4B:** Clinical image showing cyst removal via trans-oral approach, tongue is retracted and vallecular cyst (arrow head) is grasped with forceps.