Diagnosis and Treatment of Pediatric Vallecular Cysts and Pseudocysts

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ABSTRACT

Objectives/Hypothesis: Review of common presenting symptoms, diagnostic work-up, and management of vallecular cysts and pseudocysts with an evaluation of optimal treatment and risk factors for recurrence.

Study Design: Retrospective review from a tertiary care children's hospital.

Methods: We review our experience with vallecular cysts and pseudocysts from 1997 to 2009.

Results: Twelve patients (age 3 days to 13 years, mean 19 months) were included in the study. The most common presenting symptoms were stridor (8/12, 67%), respiratory distress (7/12, 58%), and feeding difficulties with or without failure to thrive (4/12, 33%). Symptoms of gastroesophageal reflux disease were present in 67% of patients and 17% carried a concurrent diagnosis of laryngomalacia. Eleven patients required operative intervention, the majority of which were a transoral endoscopic procedure. Three patients (3/11, 27%) recurred. Two of the patients who recurred required only a second procedure, but one patient required multiple procedures and continues to have a small, asymptomatic laterally based vallecular cyst. Fifty percent (2/4) of the patients 2 years or older experienced a recurrence, while only 14% (1/7) of the patients less than 2 years old had a recurrence, a difference which was not statistically significant (P=0.49). Pseudocysts tended to recur more frequently than vallecular cysts (p=0.13). Surgical approach (marsupialization versus total excision) did not affect recurrence rate. One patient with a small, asymptomatic cyst was observed and continues to be symptom-free. There were no surgical complications.

Conclusions: Vallecular cysts and pseudocysts are rare congenital lesions of the upper aerodigestive tract. Vallecular pseudocysts are more likely to recur than vallecular cysts. Surgery is the treatment of choice for symptomatic patients; smaller cysts may be followed closely.

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INTRODUCTION

Vallecular cysts (VC) (including true vallecular cysts and vallecular pseudocysts) are rare, but well known causes of upper airway obstruction and death in newborns and infants. They consist of a cyst or pseudocyst arising beneath the mucosa of the vallecula, either on the lingual surface of the epiglottis or the base of tongue [Figure 1 and 2]. Clinical presentation may include feeding difficulties and/or respiratory distress. Given the rarity of these lesions, there is no consensus in the literature as to the ideal diagnostic work-up and management. While most authors recommend flexible laryngoscopy for initial diagnostic evaluation [1,2], others have utilized lateral X- ray [1,3] and barium esophagram [4]. Treatment options include conservative medical management [2], cyst aspiration, marsupilization, surgical debulking, and laser excision [1-3].

In this study we review our experience with the diagnosis and treatment of vallecular cysts and pseudocysts, emphasizing diagnostic evaluation, surgical procedure performed, underlying pathology (pseudocyst versus cyst), recurrence rate and complications. We also review the presence of concurrent upper aero-digestive tract disorders.

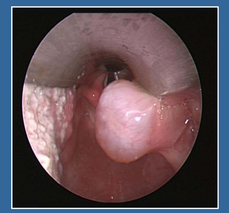

Figure 1: Vallecular cyst viewed with a O-degree endoscope. Lesion is laterally based.

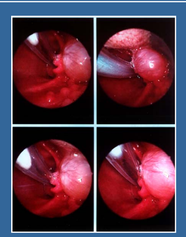

Figure 2: Vallecular cyst viewed with a 0-degree endoscope. Note compression and posterior deviation of the epiglottis

METHODS AND MATERIALS

The primary aim of this study was to determine the optimal surgical treatment of VCs. The primary outcome was surgical success, defined as no recurrence of symptoms or lesion requiring further surgery (except endoscopy for follow-up evaluation). Secondary outcomes evaluated included whether recurrence rates were impacted by type of pathology, surgical technique, penetration of the cyst during the procedure or age at presentation. We also evaluated the impact of concurrent laryngomalacia (LM) or gastrointestinal reflux disease (GERD) in patients with feeding difficulties and respiratory symptoms.

Eligible study participants included all patients who had undergone surgery on a VC between 1997 and 2009 at The Children's Hospital of Philadelphia (CHOP). Because there is no specific procedure code for excision of VC, the following procedure codes were used to identify all potential cases: 31622, 31525, 31535, 31526, 31541, 41113, and 42870. After obtaining institutional IRB approval, all surgical records between 1/1/1997 and 6/30/2009 at CHOP were queried for those procedure codes listed above. After identification, the chart (written or electronic) was reviewed to ensure appropriateness for this study.

Inclusion criteria for this retrospective review included all children (aged 0-18 years) that had undergone surgery for a VC at CHOP over the previous 12 years. Exclusion criteria included those patients with an incomplete medical record or a medical record that was not available for review. No patients were excluded from the study. Appropriate data was analyzed for statistical significance using a two-tailed Fischer's test.

RESULTS

Twelve children were identified who met the inclusion criteria. Eight were male and four were female. Age at time of diagnosis ranged from 3 days to 13 years. Table 1 lists the most common presenting symptoms.

Concurrent Illness

The majority of patients (9/12, 75%) were full term at birth with an uncomplicated neonatal course. Seven (67%) of the patients had a diagnosis of gastroesophageal reflux disease (GERD) at time of presentation and were being treated medically with proton-pump inhibitor therapy. Only one patient's reflux symptoms improved postoperatively to the point that medical therapy was discontinued. The prevalence of concurrent laryngomalacia was 17% (2/12). Neither patient required a surgical procedure to correct the laryngomalacia.

Pre-op Diagnosis & Evaluation

Eighty-three percent (10/12) of patients were diagnosed with a VC pre-operatively based on NPL exam; however, NPL failed to definitively identify a VC in two patients (17%) who had ongoing symptoms necessitating intraoperative evaluation. In one patient no obvious mass was seen; in the other patient, the NPL exam was limited due to moderate laryngomalacia and GERD-related changes. Both patients underwent rigid laryngoscopy in the operating room, where both were confirmed to have vallecular masses present. Adjunct radiologic examinations included: chest X-ray, lateral neck X-ray, neck ultrasound, barium swallow, CT neck and MRI neck. Of note, the majority of radiologic tests were ordered by non-ENT physicians prior to ENT evaluation. Interestingly, one patient had a VC incidentally discovered on an MRI performed during the evaluation of a lymphangioma of the neck..

Presenting Symptom	Number of Cases (n=12)	Percentage
Stridor	8	67%
Respiratory distress/increase work of breathing (includes cyanosis, retractions, nasal flaring)	7	58%
Feeding difficulty/poor weight gain/FFT	4	33%
Sleep disruptive symptoms	3	25%
Incidentally noted	1	9%

Table 1: Presenting symptoms of vallecular cyst

Surgery

Ninety-two percent (11/12) of patients underwent surgery. Of the ten patients whose initial surgery was at our institution, 100% (10/10) underwent a transoral endoscopic approach. Six records indicated a complete excision, while 4 records indicated that some tissue remained. Refer to Table 2.

Patient	Surgery	Complete	Recurrence	Pathology (VC vs. PC)
1	Incision and drainage; marsupialization	No	Yes, 10 days	PC
2	Complete excision	Yes	No	VC
3	marsupialization	No	No	PC
4	marsupialization	no	No	VC
5	Complete excision	Yes	No	VC
6	Complete excision	Yes	No	VC
7	marsupialization	No	No	PC
8	In-Out technique	Yes	No	VC
9	In-Out technique	Yes	No	VC
10	complete excision; microdebrider excision	Yes	Yes, 6 weeks	PC
11	Multiple	N/A	Yes, Multiple	VC then PC

Table 2: Surgical approach and characterization of excision. Complete refers to surgeon's assessment of completeness of excision. Please note that patient 11 is included in this table, although the patient's initial surgery was at an outside institution. VC: vallecular cyst; PC: pseudocyst; N/A: data not available

Recurrence

The recurrence rate in this series was 27% (3/11). 50% (2/4) of the patients 2 years or older experienced a recurrence, whereas only 14% (1/7) of the patients less than 2 years old had a recurrence (P=0.49, Fisher exact test). The recurrence rate was not affected by cyst penetration or completeness of resection (p=1). When evaluating only those patients with a single pathologic subtype (excluding patient #11), both recurrences occurred with pseudocyst pathology (p=0.13, Fisher exact test). Further, there was no statistically significant difference between cyst recurrence and surgical technique. Follow-up ranged from 4-31 weeks, while total follow-up time (defined as a re-referral back to our service for recurrence of symptoms) ranged from 1-31 months. Of the eight patients deemed free of recurrence, all had resolution of symptoms after operative intervention. Of those who did recur after the primary procedure, recurrences occurred at 10 days and 6 weeks. Both these patients remained asymptomatic after their second procedure. Only one patient, who had been first diagnosed at age 2, had multiple recurrences after a second procedure.

Complications & Post-Operative Course

There were no complications in any of the patients. The majority of patients were extubated with 24 hours of surgery without incident (7/11, 63%). The remainder of the patients remained intubated between one and four days postoperatively.

DISCUSSION

The differential diagnosis for vallecular masses in the neonate and child is limited, and includes lingual thyroid tissue, thyroglossal duct cyst, hemangioma, lymphangioma, and dermoid cyst. The majority of our patients presented with stridor and respiratory related complaints. Two patients presented with sleep obstructive symptoms. This finding has not been reported in the literature and may relate to a combination of decreased muscle tone and compression of the epiglottis by the cyst during sleep. We recommend flexible nasopharyngolaryngoscopy (NPL) as our initial diagnostic tool given the ease of performance. In cases where NPL is negative but clinical suspicion for a VC is high, we recommend direct laryngoscopy. Despite multiple reports of concurrent laryngomalacia and GERD occurring in patients with a VC [3,4,6,8], we report only a weak association with GERD and a 17% incidence of LM.

Primary treatment of vallecular cysts is surgical. A transoral endoscopic approach is preferred. Based on surgeon preference, different techniques were utilized such as marsupialization, inside-outside technique, complete excision, and incision and drainage. In all but one patient (90%), the cyst was opened during the procedure and this penetration did not affect recurrence rate. In addition, with the exception of cyst aspiration, the surgical technique used did not appear to affect recurrence rate. There appeared to be a trend towards a higher recurrence rate in older patients (>2 years old). We suspect that this may relate to the presence of increased inflammation and fibrosis. When recurrence rate was analyzed in terms of underlying pathology, pseudocysts were more likely to recur than true cysts in our series. Leuin et al reported a similar finding in 2 patients with vallecular pseudocysts, each of whom had multiple recurrences, requiring eventual transhyoid external approach. [10] Regardless of pathology, only one of our three patients who recurred needed more than a second endoscopic procedure to treat the cyst. This child continued to have recurrences with symptoms recurring between four months and three years after a procedure. Throughout those years, he was followed every three to six months. His longest symptom free interval occurred following transhyoid external resection. In reviewing his history, his symptoms usually returned after episodes of upper respiratory tract infections. More recent pathology reports have demonstrated the presence of actinomyces. After consultation with our infectious disease service, he has been treated with daily penicillin and has remained asymptomatic for eleven months. This suggests that exacerbations may be related to infections of the mass. Berger et al described similar findings in adult patients, many of whom have had VC that presented only after a primary upper respiratory tract infection [11].

CONCLUSIONS

In reviewing our results, most VCs can be successfully treated using a transoral endoscopic approach. Recurrence rate did not appear to depend on whether the cyst was excised completely or marsupialized. There was a trend towards higher rate of recurrence if the child was older or if the pathology was consistent with a pseudocyst. In only one of our patients who recurred after more than a second procedure, his pseudocyst appeared to be affected by inflammation or infection. This has been reported in the adult literature as well. Also, we found one patients with an incidental VC which has remained asymptomatic without treatment, indicating that there is a role for conservative management in some cases. Development of respiratory symptoms, weight loss, obstructive symptoms, or failure to thrive would indicate growth of the cyst, and obviate further evaluation.

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