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CLINICAL RECORD

Congenital vallecular cyst in an infant: case report and review of 52 recent cases

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Abstract

Objectives: Vallecular cyst is uncommon in infants. We treated a female infant with vallecular cyst, and curious magnetic resonance imaging findings. We also review 51 other cases of vallecular cyst in infants reported over the past 23 years.

Case report: A three-month-old female infant presented with congenital inspiratory stridor and failure to thrive. Flexible laryngoscopy and ultrasonography revealed a cystic mass in the vallecula. Magnetic resonance imaging findings were initially curious because of artefacts from breathing and swallowing. Marsupialisation of the cyst was performed. Post-operatively, the patient was immediately free of symptoms.

Conclusion: Magnetic resonance imaging presents various difficulties in infants, but has the best diagnostic effectiveness. We recommend the use of magnetic resonance imaging, flexible fibroscopy and ultrasonography to enable extensive examination of suspected vallecular cysts in infants. Marsupialisation has a recurrence rate of only one in 39 cases, and its safety and effectiveness are well balanced. Thus, prompt marsupialisation of vallecular cyst is the recommended surgical procedure.

Key words: Vallecular Cyst; Cyst; Oropharynx; Infant; Radiology; Diagnosis; Surgical Procedures, Operative

Introduction

Vallecular cysts are uncommon in infants. The first case was reported in 1881.[1] They can cause respiratory distress, dysphagia, failure to thrive,[2–5] and sometimes life-threatening airway obstruction.[6] The differential diagnosis includes thyroglossal duct cyst, dermoid cyst, adipose tumour, lymphangioma, haemangioma and lingual thyroid.[5,7] Post-operative airway management is difficult if an invasive surgical procedure is required, because of the small respiratory tract in infants. Therefore, pre-operative diagnosis and planning of the treatment strategy are very important. Vallecular cyst in infants is associated with a low morbidity rate, so few clinical studies have been reported in recent years.[5,8] Consequently, the appropriate diagnostic procedure and treatment strategy continue to be discussed.

We treated a female infant with a vallecular cyst manifesting as congenital stridor and failure to thrive, with curious magnetic resonance imaging (MRI) findings. We describe this case, and also review 51 other cases of vallecular cyst in infants reported over the past 23 years,[4,5,7–23] focussing on diagnostic methods and surgical procedures.

Case report

A three-month-old female infant presented with congenital inspiratory stridor and failure to thrive.

She had been born uneventfully at 40 weeks via normal spontaneous delivery, weighing 3.342 kg, and had had no perinatal problems. She had suffered inspiratory stridor since birth, but a misdiagnosis of laryngomalacia had been made, without a detailed examination. Feeding difficulty and failure to thrive had persisted since the age of five weeks. She was referred to our hospital at the age of three months for detailed investigation.

On admission, physical examination revealed the following: body temperature, 36.6°C; heart rate, 150/minute; respiratory rate, 40/minute; body weight, 5.018 kg; and body height, 60 cm.

Chest radiography and routine laboratory investigations (including thyroid hormone levels and blood gas analysis) showed no abnormalities.

Ultrasonography (US) revealed a round, low, echogenic mass at the base of the tongue, and a normal thyroid gland. Flexible laryngoscopy revealed a smooth, cystic mass in the vallecula, and excluded airway abnormalities.

A T1-weighted MRI scan demonstrated a 15-mm diameter, well circumscribed, spherical mass in the vallecula. A T2-weighted MRI scan showed the mass contents as hyperintense and its margin as slightly hypointense (Figure 1).

We had initially suspected a partly solid tumour. However, endoscopic and MRI findings were inconsistent, so we decided to repeat the MRI scan using a high-speed scan mode (utilising the Magnetom Avanto system; Siemens, Munich, Germany). Sagittal images indicated that the homogeneous lesion moved up and down slightly with breathing and swallowing, in the absence of larger body movements (Figure 2). This confirmed that the lesion was a homogeneous, cystic mass, and established the diagnosis of vallecular cyst.

Marsupialisation of the cyst was performed under general anaesthesia, using electrical cautery under direct laryngoscopic visualisation with an operating microscope.

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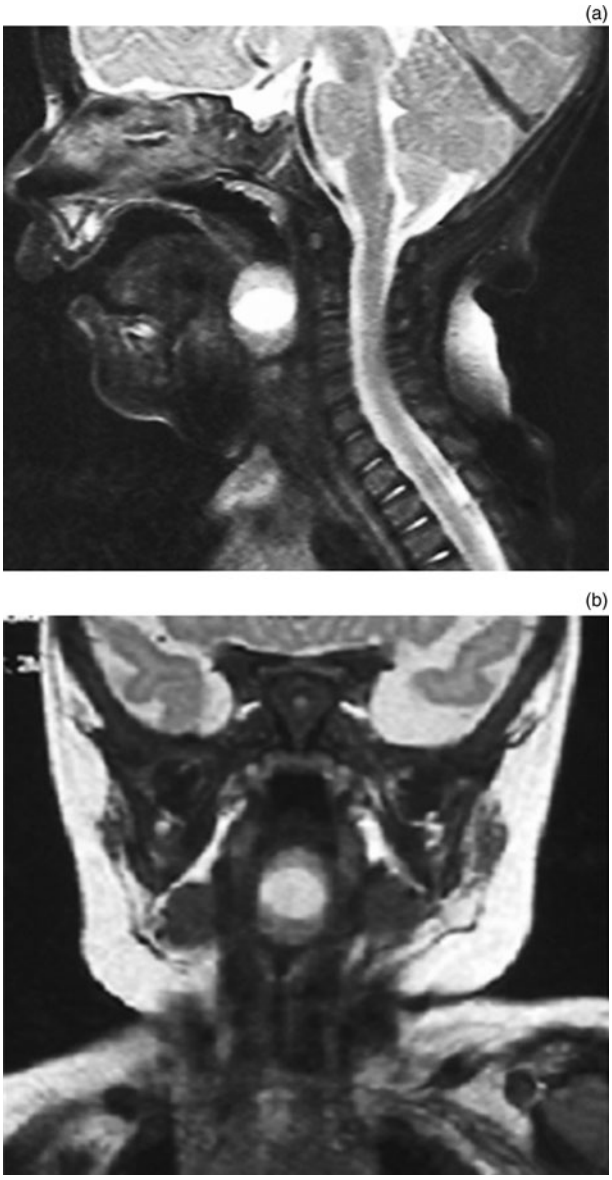

FIG. 1

(a) Sagittal and (b) coronal T2-weighted magnetic resonance images demonstrating a 15-mm diameter, well circumscribed mass in the vallecula, with hyperintense contents and a slightly hypointense margin.

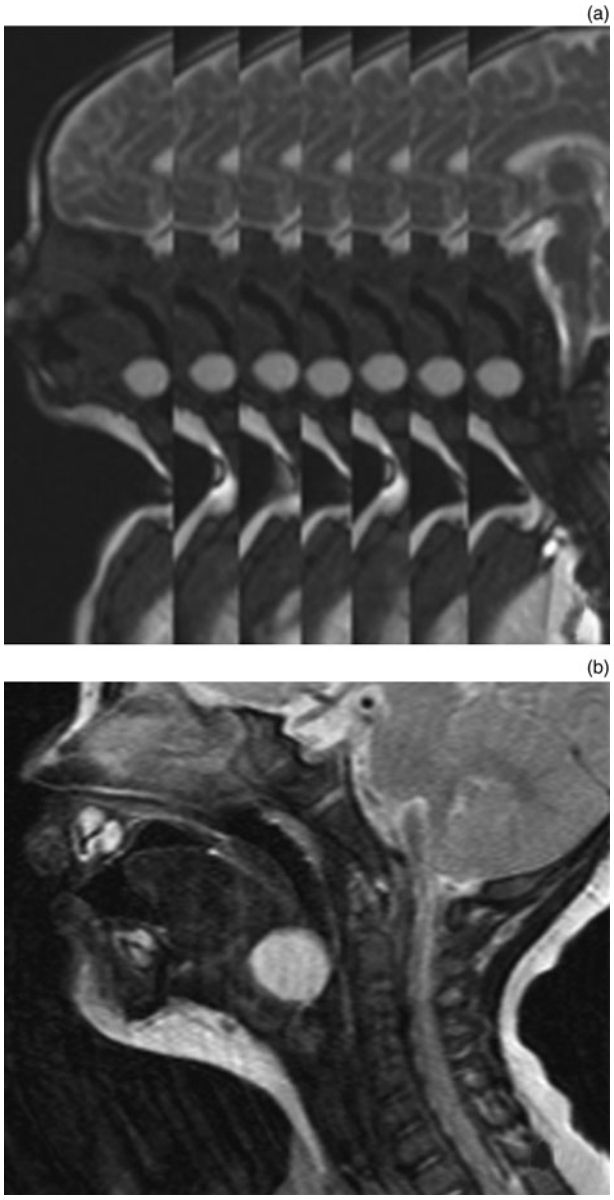

FIG. 2

(a) Sagittal, high-speed scan mode magnetic resonance image (MRI) scans demonstrating slight rising and falling movement of the lesion with breathing and swallowing, in the absence of large body movements. (b) Sagittal, T2-weighted MRI scan showing the lesion to be homogeneously hyperintense.

Histological examination of the operative specimen showed laryngeal epithelium with squamous metaplasia, connective tissue with mild inflammatory cell infiltration, and respiratory epithelium with squamous metaplasia. No thyroid tissue was found.

The post-operative course was uneventful. The patient's symptoms immediately resolved, and she was discharged 12 days after surgery.

Eight months after surgery, flexible laryngoscopic examination showed no abnormality in the larynx. The patient was in perfect health and had reached a normal weight.

Discussion

Congenital stridor is a relatively common symptom in infants, but vallecular cyst is a rare cause. The major differential diagnosis of congenital stridor includes laryngomalacia, subglottic stenosis and vocal fold paralysis;[24,25] in such cases, the incidence of vallecular cyst is only 0.9–2.0 per cent.[24,25] Vallecular cyst carries the risk of sudden airway obstruction and death, because of the anatomical location of the cyst and the small respiratory tract in infants.[6] Therefore, all otolaryngologists should consider this uncommon condition in the differential diagnosis of congenital stridor in infants.[14]

The diagnosis of vallecular cyst may be assisted by lateral radiography, flexible laryngoscopy, direct laryngoscopy, computed tomography (CT), MRI, US, barium oesophagography and thyroid scanning examinations. Figure 3 shows the use of diagnostic investigations in the 52 cases published to date. Radiography was used in 39 cases, but made no contribution to the diagnosis in 15 of these 39. Radiography is convenient and sometimes helpful,[7,12,13] but it is difficult to interpret and has many diagnostic pitfalls.[7,8] These

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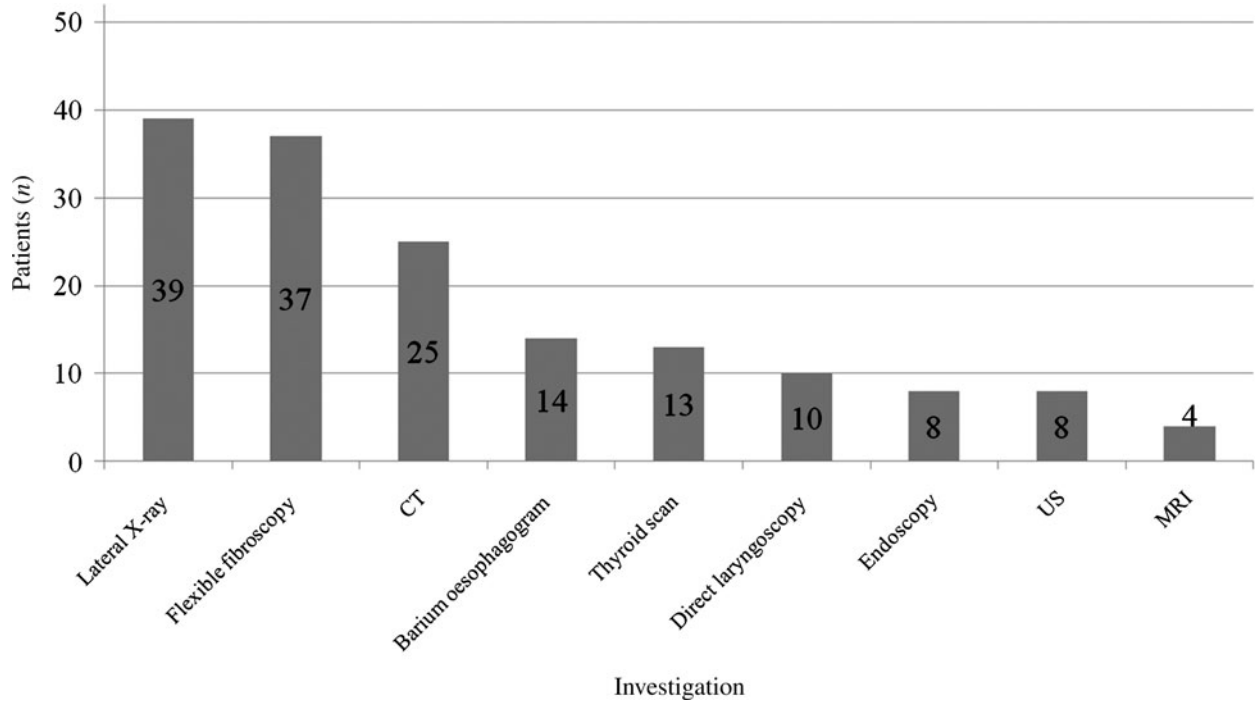

FIG. 3
Use of diagnostic investigations in the 52 cases reviewed.

findings suggest that radiography is insufficient as the sole investigation for congenital stridor.

Flexible endoscopy (using a laryngoscope or bronchoscope) was used in 37 cases, and successfully identified the lesion in all cases. Flexible endoscopic examination is quick, easy and minimally invasive,[4] and allows dynamic assessment of the larynx, evaluation of swallowing and detection of congenital oropharyngeal lesions.[13,16] However, it cannot make fine distinctions between the various differential diagnoses of vallecular cyst.[16] Therefore, we propose that flexible endoscopy should be replaced by direct laryngoscopy, performed by a skilled operator under cardiopulmonary monitoring, to enable methodical examination of such cases.[12,26] Flexible endoscopy is recommended for initial screening.

Computed tomography was used in 25 of the reviewed cases, and may be the most common type of radiographic imaging. Computed tomography may be indispensable for the diagnosis of lingual thyroglossal duct cyst in neonates;[26] however, vallecular cyst, thyroglossal duct cyst, dermoid cyst, haemangioma and lymphangioma all have a similar, low density appearance on CT.[27] In addition, CT involves a radiation risk for infants.

Magnetic resonance imaging is the diagnostic modality of choice,[20,21] and detailed knowledge of the distinct MRI characteristics of common vallecular masses will enable correct diagnosis.[16] However, the use of MRI was described in only four of our reviewed cases. This modality requires sufficient sedation to exclude motion artefacts, together with cautious airway management in infants;[28] it is also costly. Therefore, MRI is not widely used. Magnetic resonance imaging under sedation was performed in our patient. At first, we thought her lesion was partly solid, because of the above-described MRI findings in the apparent absence of motion artefacts. The interpretation of pharyngolaryngeal MRI in infants should consider the influence of breathing and swallowing, which cannot be prevented by sedation. High speed scanning should be used if findings are not logically consistent.

Ultrasonography is non-invasive, easy to perform, and familiar to otolaryngologists and paediatricians. It provides a good distinction between solid and cystic masses, as well as confirmation of a normal thyroid gland.[18,22] Ultrasonography was used in only eight of our reviewed cases. We propose that US can provide a useful screening method for congenital stridor, especially for paediatricians who have no experience with flexible fibroscopy.

Lingual thyroid may present as a mass lesion at the base of the tongue, and thyroid scanning and thyroid function tests are necessary to confirm a normally located and functioning thyroid gland.[5] Thyroid scanning was performed in 13 of our review cases. However, it carries a radiation risk and is difficult to administer in infants. We suggest that US and thyroid function tests, as used in our patient, are sufficient for confirmation of a normal thyroid gland.

Table I summarises the 52 published cases of infantile vallecular cyst (including the present case). The mean age at presentation was 2.5 months (range, 2 days to nine months; 38 cases), and the female:male ratio was 1:1.3. The most common diagnostic strategy was a combination of CT, flexible fibroscopy and other techniques (22 cases); this strategy has become standard practice over the last 22 years. The use of MRI has various difficulties in infants, but has the greatest diagnostic effectiveness.[16] Hence, we recommend that a combination of MRI, flexible fibroscopy and US should be used to enable extensive examination of possible cases of vallecular cyst.

Surgical treatment for vallecular cyst in infants includes aspiration, marsupialisation (i.e. de-roofing) and extirpation (i.e. resection and excision). Surgery was performed in 51 of our 52 reviewed cases (Table II).

In the reviewed cases, aspiration had a high rate of recurrence and was of limited use.[3,16] Small cysts can be

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TABLE I
REPORTED CASES OF INFANTILE VALLECULAR CYST (PAST 23 YEARS)

Year	Author(s)	Pt age	Sex	Main investgn	Cyst size (mm)	Coexisting abnormality	Surgery	FU (mth)	Recurrence	
									Time	Treatment
1988	Myer[9]	8 wk	F	CT + DL			Mar	3		
1989	LaBagnara[10]	2.5 mth	M	X-ray	20		Asp	6	6 mth	Mar
1992	Gluckman *et al.*[4]	12 d	M	Flex + DL	15		Exc	6		
1995	Wang & Lim[11]	2 mth	F	CT + DL			Mar (laser)	1		
1995	Wang & Lim[11]	2 mth	F	CT + DL			Mar (laser)	1		
1995	Wong *et al.*[7]	2 mth	F	CT + flex	10	LM	Exc (laser)			
1995	Wong *et al.*[7]	1 mth	M	Flex	25	LM	Exc			
1996	Oluwole[12]	5 mth	M	DL			Mar			
1999	Gutierrez *et al.*[8]	1 d to 16 wk[†]	5 M, 3 F	Endoscopy[‡]	4–12[**]		Mar (8 pts)[§]	3–36	8 wk (1 pt)[#]	Mar (laser)
2000	Hsieh *et al.*[5]	<2 mth	8 M, 6 F	CT + flex[‡]		LM (9 pts)	Mar (12 pts), asp (1 pt), con (1 pt)	6–48		
2000	Ku[13]	6 wk	F	CT + flex + DL	10	LM	Mar			
2000	Ku[13]	6 wk	M	DL	15		Mar			
2000	Ku[13]	19 d	F	Flex			Exc (laser)	13		
2000	Ku[13]	2 d	M	Flex + DL	17.5		Mar			
2002	Tuncer *et al.*[14]	3 mth	M	CT + flex	15		Mar (laser)	6		
2002	Chow *et al.*[15]	25 d	M	Flex		LM	Ext	11		
2003	Tibesar & Thompson[16]	4 mth	F	MRI + flex + DL	7		Ext	12		
2004	Yao *et al.*[17]	11 wk	F	Flex	20	LM, GER	Exc (laser)	12		
2004	Ahrens *et al.*[18]	3 mth	F	MRI + flex + US	8	LM	Mar	0.5		
2008	Yang *et al.*[19]	1 mth	M	CT + flex + US	13	GER	Mar (laser)	6		
2008	Mahajan *et al.*[20]	3 mth	M	CT + DL	20	GER	Ext			
2009	Grasso *et al.*[21]	4 mth	M	MRI + flex	20		Mar (laser)			
2009	Breysem *et al.*[22]	2 mth	F	Flex + US	15		Res (laser)	~2–12 mth[¥]		
2009	Breysem *et al.*[22]	9 mth	M	CT + flex	20		Mar (laser)	~2–12 mth[¥]		
2009	Breysem *et al.*[22]	3 mth	M	Flex + US	8		Mar (laser)	~2–12 mth[¥]		
2009	Breysem *et al.*[22]	1 mth	M	CT + flex + US	20		Mar	~2–12 mth[¥]		
2009	Breysem *et al.*[22]	3 mth	M	Flex + US			Res (laser)	~2–12 mth[¥]		
2009	Sands *et al.*[23]	5 wk	F	Flex	11 × 9	LM	Mar	1		
2009	Sands *et al.*[23]	6 wk	M	CT + flex + EUA	9 × 5	LM	Mar	1		
2009	Sands *et al.*[23]	2 wk	F	CT + flex + US	20 × 9		Mar	10		
2009	Sands *et al.*[23]	8 mth	F	Flex + EUA	6 × 6	LM	Mar	1		
2011	Present case	3 mth	F	MRI + flex + US	15		Mar	9		

[†]Mean = 40 days. [‡]All patients. [**]Mean = 8 mm. [§]With laser for 5 pts. [#]Forceps = this cyst was marsupialized by forceps at initial surgery. [¥]For series. Pt = patient; investgn = diagnostic investigation; FU = follow up; wk = weeks; mth = months; d = days; F = female; M = male; CT = computed tomography; DL = direct laryngoscopy; flex = flexible laryngo(broncho)scopy; MRI = magnetic resonance imaging; US = ultrasonography; EUA = examination under anaesthesis; LM = laryngomalacia; GER = gastroesophageal reflux; mar = marsupialisation; asp = aspiration; exc = excision; con = conservative; ext = extirpation

TABLE II
SURGERY AND RECURRENCE RATES: 51 PREVIOUSLY PUBLISHED CASES

Procedure	Pts (*n*)	Recurrence	
		n	%
Extirpation*	10	0	0
Marsupialisation (with laser)	39 (14)	1 (0)	2.6 (0)
Aspiration	2	1	50
Total	51	2	3.9

*Including resection and excision. Pts = patients

aspirated,[12] but aspiration should only be used as a palliative procedure, or as an initial manoeuvre if intubation is difficult.[3,10]

- **Vallecular cysts are uncommon in infants**
- **Magnetic resonance imaging presents various difficulties in infants, but has the highest diagnostic effectiveness for these lesions when used carefully**
- **The recurrence rate after marsupialisation is very low, so prompt marsupialisation is the recommended procedure for these lesions in infants**

In general, marsupialisation or extirpation is used for definitive treatment.[3–5,13,17] Extirpation is the ideal treatment but sometimes requires an external approach[3] with or without tracheostomy. A published review of nine patients with congenital laryngeal cyst reported no recurrence in four after excision and in five after marsupialisation.[29] In our review of 52 cases (including the presented case), recurrence was encountered in none of 10 patients after extirpation, and in one of 39 patients after marsupialisation. In particular, recurrence occurred in none of 14 patients following marsupialisation with laser instruments. Therefore, our study suggests that marsupialisation and extirpation are equally effective, and that marsupialisation, especially with laser instruments, is to be recommended because of its minimal invasiveness.

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