CASE REPORT

A case of acquired laryngomalacia in an infant, with endoscopy before and after establishing the diagnosis for the first time

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SUMMARY

A 12-week-old infant girl born at 27 weeks gestation, with multiple comorbidities, was referred to the paediatric otolaryngology team due to stridor and feeding difficulties. Initial airway endoscopy was performed, showing no abnormal findings. The infant underwent frequent hospitalisations for recurrent cyanotic spells. Despite initial laryngoscopy and bronchoscopy not showing abnormalities, a repeat endoscopy demonstrated laryngomalacia, significant enough to require a supraglottoplasty. This is the first report to confirm, with video evidence, that some cases of laryngomalacia are acquired rather than congenital, even in infancy.

BACKGROUND

Laryngomalacia (LM) is the most common congenital laryngeal anomaly, however, its pathophysiology is not very well understood. Mechanical,¹ anatomical,² neurological^{3–6} and multiple aetiological theories have been suggested.^{7–10} Recently, some authors introduced terms such as state dependent, late onset and occult LM, to describe an atypical onset or a state that precipitated the supraglottic collapse. Others reported on atypical primary clinical presentations of LM, such as sleep disordered breathing (SDB) and swallowing dysfunction (SWD).^{11–12} It is unclear if these reports pertain to one or different varieties of LM, casting doubt on the generally accepted notion that LM is universally a congenital condition.

Only 11 cases of acquired LM have been described in children.^{13–16} We present this case of acquired LM in an infant with persistent symptoms of stridor, SDB and SWD, despite having early normal upper and lower airway examinations.

CASE PRESENTATION

A 12-week-old infant girl born at 27 weeks gestation, the surviving twin of a diamniotic–dichorionic twin pregnancy, was referred to the paediatric otolaryngology team due to airway congestion, stridor, dusky spells and feeding difficulties. At that time, she was in the neonatal intensive care unit feeding orally, supplemented by nasogastric (NG) tube. Her medical history was significant for stage 1 retinopathy of prematurity in the left eye, apnoea of prematurity, respiratory distress syndrome, for which she received surfactant and was intubated, stage 1 necrotising enterocolitis, two small atrial septal defects and mild pulmonary stenosis.

On examination, there was evidence of stridor, noisy breathing with feeding, desaturations and intermittent bradycardia. Nasal congestion was also apparent. Otherwise, the rest of the head and neck examination was within normal limits.

INVESTIGATIONS AND TREATMENT

Under general anaesthesia (allowing spontaneous respiration using intravenous propofol and remifentanyl), an upper airway examination using a neonatal flexible laryngoscope demonstrated significant oedema of the larynx and hypopharynx and pooling of secretions. Rigid laryngoscopy and bronchoscopy showed vocal cord sulci bilaterally and oedema. No subglottic stenosis, malacia or fistula was seen (video 1). Budesonide (125 µg q12 h) and lansoprazole (2.5 mg NG q12 h) were advised for her laryngeal oedema and possible gastro-oesophageal reflux disease (GORD). The speech-language pathology service clinically assessed swallowing, and recommended continuous work on breast feeding and supplementing it by gavage.

Within 4 months following discharge, the infant had been hospitalised on three occasions due to increased problems with feeding, and coughing, retching, vomiting, and exhibiting increased work of breathing and desaturations. With persistence of symptoms and signs of aspiration, a multidisciplinary team restricted feeding to the alternate route, and advanced her NG tube to nasojejunal, and

Video 1 Initial suspension laryngoscopy and bronchoscopy with no abnormal findings.

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eventually a gastrostomy and Nissen fundoplication were undertaken.

At 7 months of age, the baby was hospitalised once again due to problems with recurrent choking during sleep and while awake, during which she would become cyanotic. Paediatric otolaryngology was consulted again and a repeat suspension laryngoscopy and bronchoscopy were planned. On that occasion, LM was found causing obstruction of the laryngeal inlet and glottis (video 2). At that time, it became evident that further intervention would be necessary. Therefore, the parents were consented for supraglottoplasty (SGP). The redundant supra-arytaenoid tissue was trimmed bilaterally using microscissors, and access to the glottis opening was improved with no complications.

At 10 months of age, a home overnight oximetry was performed for possible SDB. It was consistent with obstructive sleep apnoea (OSA), showing low baseline saturation value (mean saturation 93.6%) with increased variability, frequent clusters of desaturations and presence of frequent hypoxaemia (minimum saturation 75%; oxygen desaturation index 17.5 events/h).

Following that, an overnight split-night study (overnight polysomnogram) was performed. The baby was noted to have 37 central, 25 obstructive, 3 mixed and 65 total apnoeas, and her apnoea-hypopnoea index was 36.6 events/h. At this point, she was started on bilevel ventilation, with a final pressure of inspiratory positive airway pressure 9 and expiratory positive airway pressure 5, and a backup rate of 14. At these pressures, she achieved slow-wave and rapid eye movement sleep. Baseline oxygen saturations were maintained above 95%, and transcutaneous CO₂ below 40 mm Hg recommended pressures.

At 17 months of age, a repeat suspension laryngoscopy and bronchoscopy due to salivary aspirations was performed, showing normal vocal cord mobility and profuse salivary secretions penetrating the larynx. No adhesions were seen and no residual supraglottic prolapse was evident. Bilateral clipping of the parotid and submandibular salivary ducts was performed to control salivary aspiration.

OUTCOME AND FOLLOW-UP

The patient had continued on bilevel non-invasive positive airway pressure and started glycopyrrolate to reduce the

secretions for 3 days postoperatively. She remained stable with no further hospitalisations and no further symptoms. She has regular outpatient follow-ups at the paediatric otolaryngology clinic and has shown marked improvement.

DISCUSSION

We report a case of a premature infant, who presented with stridor, respiratory and swallowing difficulties, and whose initial examination with video documentation showed no evidence of LM. Owing to persistence of symptoms, repeat endoscopy demonstrated acquired LM.

Acquired LM has been reported in the literature in children,^{1 13–16} but more so in adults,^{5–10 17} mainly as individual case reports or short case series. Largely, two theories have been proposed to explain it: one neurological and the other mechanical. The neurological theory proposes that defective central neurological control of the muscular support of the supraglottic structures is to blame.⁴ The mechanical theory is based on the existence of a vicious cycle of increased negative pressure across the laryngeal inlet (eg, secondary to SDB), with recurrent inspiratory prolapse of the supraglottic structures leading eventually to oedema of mucosal and submucosal tissues, which ultimately precipitates the redundancy of supra-arytaenoid tissues and aryepiglottic folds.¹

In children, the majority of cases were described in association with some neurological impairment.^{13 15} Archer described an 11-year-old boy who developed LM following basilar artery thrombosis causing extensive pontine infarction.¹³ Peron *et al* reported the first series of acquired LM. Seven cases were described, in which LM was subsequent to: coma secondary to alcohol and drug overdose; microcephaly and cerebral palsy with seizure disorder; post-traumatic comas following a motor vehicle accident in three patients; prematurity with hydrocephalus; and seizure disorder; and bacterial meningitis that developed sequelae of hypotonia, seizures and hydrocephalus.¹⁵ All these reports supported the neurological theory. A minority postulated a mechanical theory. Cunningham *et al*¹ presented two children who demonstrated stridor and respiratory distress after adenotonsillectomies, and exhibited inspiratory collapse of oedematous supraglottic structures. Common to all of these, however, is the absence of an earlier endoscopy ruling out the pre-existence of the condition, thereby depending totally on circumstantial evidence and the absence of prior symptoms attributable to LM.

Meanwhile, the literature on adults has a good few more cases that are generally ascribed to either being a neurological^{5 6} or iatrogenic entity (after some form of neck or floor mouth surgery),^{6 7 9} or being idiopathic in nature.^{8 10} Again, none of these reports document how the larynx appeared before this diagnosis was discovered.

Several new terms have been lately introduced to describe LM. Authors used the terms 'state dependent',^{18–20} 'late onset',²¹ and 'occult LM'.^{22 23}

Ten cases of state-dependent LM have been reported.^{18–20} Sproson *et al*¹⁸ reported a 12-year-old child who developed state-dependent LM following anaesthesia with a laryngeal mask. Amin and Isaacson¹⁹ reported five children with state-dependent LM, while Smith *et al*²⁰ described four children. All these children developed their condition on induction of anaesthesia. Given the fact that these children had normal function of the larynx while awake and supraglottic collapse was only witnessed while they were spontaneously breathing under general anaesthesia, a proposed explanation of a neurogenic mechanism

Video 2 Repeat suspension laryngoscopy and bronchoscopy confirming the diagnosis of acquired laryngomalacia.

was justified.^{18–20} It is plausible that these reports are a variant of acquired LM, notwithstanding the proposed explanations.

Two recent studies have documented the variable primary presentations of LM in children, with stridor being the most common, followed by snoring-SDB and SWD.^{11 12} Olney *et al* described three different types of LM in 1999. Type 1 being prolapse of the mucosa overlying the arytaenoid cartilages. Type 2 presenting as foreshortened aryepiglottic folds. Type 3 exhibiting posterior displacement of the epiglottis.²⁴ According to Erickson *et al*, atypical presentations may represent a different category of LM depending on anatomical dysfunction or type of LM, as their study significantly correlated the type of LM with age and primary presentation. However, this study was performed in neurologically intact children.¹²

We postulate that pharyngeal and laryngeal trauma and increased negative pressure from SDB and SWD led to the development of LM in our patient. This may explain why there was no improvement subsequent to surgery in our patient. Also, Chan *et al* examined the outcomes after SGP in children with OSA and non-infantile occult LM. Their findings propose that significant improvement of sleep in children relies on surgical correction, yet, medical comorbidities diminished the efficacy of surgery.²²

It has been claimed that GORD is greatly associated with LM and, hence, related in its aetiology.²⁵ In 2007, Thompson studied the role GORD provides in causing LM. It has been portrayed that gastric acid causes functional denervation of the laryngeal afferent response. A decline in the sensitivity of the superior laryngeal nerve responsible for the patency of the larynx causes this abnormal sensorimotor integrative function,³ which may clarify the pathophysiology of acquired LM.

This report, with its documented video endoscopy, is a first in the literature of acquired LM. It shows the development of LM in an infant with an originally normal larynx since birth. It is highly suggested that acquired LM exists, unless congenital LM develops over time. However, it may not be the epicentre of clinical symptoms.

Learning points

- Laryngoscopy and bronchoscopy are the essential procedures required to diagnose laryngomalacia (LM).
- Sleep oximetry and polysomnogram should be routinely performed in cases of obstructive sleep apnoea.
- Gastro-oesophageal reflux disease is highly associated with LM and may be one of the major causes of acquired LM.

Contributors MJG was involved in substantial contributions to the conception of the work and acquisition of data; drafting the work; final approval of the version published and agreement to be accountable for all aspects of the work. HE-H was responsible for substantial contributions to the conception of the work and

interpretation of data; revising the work critically for important intellectual content; final approval of the version published and agreement to be accountable for all aspects of the work.

Competing interests None declared.

Patient consent Obtained.

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