Persistent unrecognized congenital laryngomalacia as the cause of dyspnea in a 48 year-old woman

Melanie Giesler D.O.[1], Libby Smith D.O.[2]

Affinity Medical Center, Massillon, Ohio [1]
University of Pittsburgh Medical Center, Pittsburgh, Pennsylvania [2]

INTRODUCTION

Unrecognized symptomatic laryngomalacia diagnosed in adulthood is extremely rare.

Laryngomalacia is the most common congenital malformation of the larynx.[1] It is an anatomic disorder of the supraglottic structures characterized by a flaccid larynx and inward prolapse of supraglottic structures in infants with congenital inspiratory stridor. Laryngomalacia can also be acquired or exercise-induced. Acquired laryngomalacia rarely occurs in patients without a history of neurologic injury, radical neck dissection or trauma.[2,3]

For congenital laryngomalacia, the vast majority of these patients outgrow the condition by 18 months, necessitating only conservative treatment of prone positioning and proton pump inhibitors to protect the larynx from gastric acid during the negative pressure generated while inhaling against the supraglottic tissue.[4,6] Only a small percentage (10%) require supraglottoplasty for apnea or failure to thrive.[5,6]

CASE HISTORY

A 48-year-old female was referred to a tertiary care center for treatment of suspected idiopathic vocal fold paralysis. She had dyspnea at rest, with exertion, as well as with speaking She denied dysphagia. She recalled the dyspnea symptoms were present as a child, precluding her from "keeping up" with the other children. She did not need special positioning as a newborn. As an adult, she avoided exertion and speaking due to her persistent dyspnea. Additionally, her husband described her gasping for air while sleeping, although she had never been evaluated for obstructive sleep apnea. She also had complaints of vocal fatigue and decreased volume. She had been treated unsuccessfully for asthma for years. Recent pulmonary function tests were normal.

EXAMINATION

A fiberoptic flexible laryngoscopy (FFL) revealed severe left arytenoid mucosa prolapse anteriorly, leading to glottic obstruction, and preventing clear visualization of her true vocal folds. Close review of the exam demonstrated mild right true vocal fold paresis with decreased abduction and normal adduction, contrary to her previous diagnosis of vocal fold paralysis. (Figure 1)

It was thought at this point that the dyspnea was due to airway obstruction from laryngomalacia rather than vocal fold paralysis.

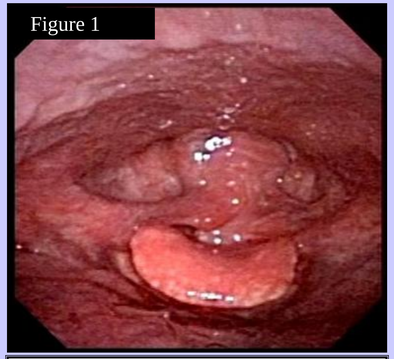
TREATMENT

The patient took twice daily proton pump inhibitors in anticipation for microsuspension laryngoscopy with CO_2 laser supraglottoplasty, with excision of redundant left arytenoid tissue. (Figure 2) Surgical treatment was performed without complications.

One month post-operatively, her dyspnea (at rest, exertional, and obstructive while sleeping), and vocal fatigue had resolved. An FFL revealed a widely patent airway. (Figure 3)

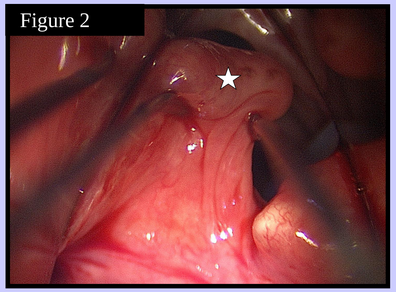
At 6 months after surgery, the patient's exercise tolerance continued to improve. She lost 60 pounds due to increased physical activity. An FFL showed a completely healed surgical site and patent airway. (Figure 4, 5)

Figure 1



FFL, inhalation, demonstrating collapse of redundant supraglottic tissue over airway.

Figure 2

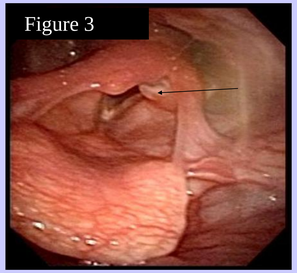


Intraoperative photo at time of supraglottoplasty showing redundant left arytenoid tissue (☆) being retracted anteriorly.

Figure 4: FFL, inhalation, 6 months post-op.

Figure 5: FFL, phonation, 6 months post-op.

Figure 3



FFL, inhalation, 1 month post supraglottoplasty. Arrow – left arytenoid post-operative eschar.

Figure 4

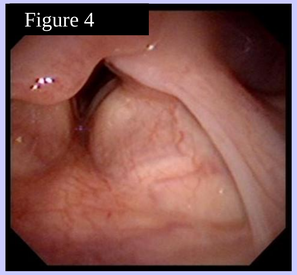
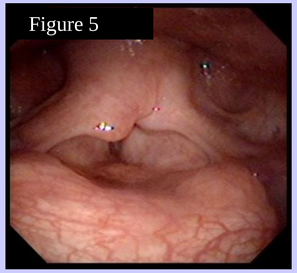


Figure 5



DISCUSSION

This patient's quality of life had been significantly decreased secondary to the burden of misdiagnosis and therefore delayed treatment of laryngomalacia.

This patient had been misdiagnosed as having vocal fold paralysis and asthma as the etiologies of her dyspnea. The anterior arytenoid prolapse may have been mistaken for a sign indicating vocal fold paralysis. In addition, it was difficult to observe vocal fold motion due to this finding.

Acquired laryngomalacia is usually present in the face of neurologic injury, radical neck dissection, post-radiation therapy or trauma. She had none of these risk factors. It is more likely that she had congenital laryngomalacia which was never diagnosed in childhood, worsened with time, and was mistaken for recalcitrant asthma and vocal fold paralysis.

CONCLUSIONS

Laryngomalacia must remain in the differential diagnosis for adult patients with respiratory distress not amenable to medical therapy.

REFERENCES

1. Archer, S. Acquired Flaccid Larynx: A Case Report Supporting the Neurologic Theory of Laryngomalacia. *Arch Otolaryngology Head Neck Surg*. 1992;118: 654-657.
2. Richter, G, et al. Late-onset laryngomalacia - a variant of disease. *Arch Otolaryngology Head Neck Surg*. 2008;134(1):75-80.
3. Siou, G, Jeannon, P, Stafford, F. Acquired idiopathic laryngomalacia treated by lase aryepiglottoplasty. *Journal or Laryngology and Otology* 2002;116:733-735.
4. Woo, P. Acquired laryngomalacia:epiglottis prolapse as a cause of airway obstruction. *Ann Otol Rhinol Laryngol*. 1992; 101(4):314-320.
5. Kay, D. et al Laryngomalacia: A classification system and surgical treatment strategy. *Ear, nose & throat*. 2006; 85: 328-331.
6. Olney, D, et al. Laryngomalacia and its treatment. *Laryngoscope*. 1999: 109(11): 1770-1775.
7. Hitchings, A, Crampsey, D, Kubba, H. A variant of laryngomalacia in the neurologically normal older child. *International Journal of Pediatric Otorhinolaryngology*. 2007; 71: 571-574.