Case Report of a 30 year old man with a Congenital Laryngeal Web

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Abstract

Objective
To present a rare and interesting case of a 30 year old man with a congenital laryngeal web.

Methods
We reviewed the chart for this case and conducted a literature review in OVID medline (1966 to 2010). This study was deemed exempt by the Western University, Office of Research Ethics.

Results
A 30 year old man presented with hoarseness since birth. He had dysmorphic facial features, fissured tongue, irregular articulation, orthopaedic abnormalities of the hands and elbows, and mental retardation. It was unclear why the patient did not seek medical attention earlier. He was born in the Bahamas and had a weak cry and hoarseness since birth. He also had “breathing troubles”, especially at night and during exercise. Videostroboscopy showed a glottis web obstructing up to ¾ of his airway. He was treated with CO2 laser lysis of this glottis web and triamcinolone injection. Postoperatively, his voice and breathing symptoms resolved completely. Cytogenetics testing was negative for 22q11 microdeletion syndrome.

Conclusion
This 30 year old man had a congenital laryngeal web. A review of the literature showed that it is rare for congenital laryngeal webs to present so late in life. We present this interesting case to highlight the importance of keeping this diagnosis in the differential for hoarseness in adults.

Introduction
A congenital web is abnormal fibrous tissue that forms between two structures in the larynx. Congenital laryngeal webs are rare, accounting for about 5% of congenital anomalies of the larynx [1,2]. The most common location is at the level of the glottis extending across the anterior one third of the vocal cords. It can also extend to the posterior glottis and inferiorly to the subglottis. This congenital anomaly forms during the tenth week of embryogenesis due to incomplete recanalization of the primitive larynx [1]. There is a spectrum of anomalies ranging from a congenital laryngeal web to laryngeal atresia [3]. Although anterior laryngeal webs are usually congenital in origin, they may also be acquired secondary to surgery, intubation, or infection (Corynebacterium diphtheriae or Bacillus cereus) [4].

Clinical presentation
There is a variety of symptoms, depending on the severity of the laryngeal web. The presenting complaint is usually an abnormally weak, soft, husky, or absent cry since birth [4]. Other symptoms include stridor (usually biphasic and worsened in exertion), recurrent croup, and episodic cyanosis [5]. In extreme cases, newborns may present in respiratory distress.

Most congenital laryngeal webs present in early childhood. It is extremely rare in adults [6]. Some cases of adults with small laryngeal webs have been reported [4]. For example, it the web is small enough, it can be found during anesthetic intubation during adulthood. The oldest reported case in the literature is a 45 year old man who presented with a 1-month history of cough and hoarseness and his symptoms worsened with a cold. He was found on videostroboscopy to have a anterior laryngeal web and treated endoscopically with lysis and topical application of mitomycin-C. [6]

Association with deletions of 22q11
Up to 65% of patients with anterior glottis webs were positive for chromosome 22q11.2 deletion [7]. This deletion causes a wide range of phenotypes including DiGeorge’s syndrome, velocardiofacial (Shprintzen) syndrome, conotruncal anomaly face syndrome, heart defects, and CATCH 22 (cardiac defect, abnormal facies, thyim hypoplasia, cleft palate, hypocalcemia, and chromosome 22 deletion). It is strongly recommended that all patients diagnosed with anterior glottic webs undergo fluorescence in situ hybridization evaluation for this chromosome deletion.

Case Presentation
A 30 year old man presented with hoarseness since birth. He had “breathing troubles”, especially at night and during exercise. It was unclear why the patient did not seek medical attention earlier. He was born in the Bahamas and had recently immigrated to Canada. He had no previous history of surgery, intubation, or severe infection.

Physical exam showed dysmorphic facial features, fissured tongue, irregular articulation, orthopaedic abnormalities of the hands and elbows, and mental retardation. Videostroboscopy showed an anterior glottis web obstructing up to ¾ of his airway. Cytogenetics testing was performed and was negative for 22q11 microdeletion syndrome.

He was treated with CO2 laser lysis of this glottis web and triamcinolone injection. Postoperatively, his voice and breathing symptoms resolved completely. He is now symptom free 1 year post surgery.

Treatment options
The first treatment goal is to stabilize the patient who may be in respiratory distress. This may require a tracheotomy. The second goal is to preserve function of the larynx (breathing and voice). Depending on the severity of the laryngeal web, there are three options: 1) Observation. This is an option if the patient is relatively symptom free. 2) Endoscopic. The web can be lysed with cold steel or a CO2/KTP laser. Topical mitomycin C can be applied. A silastic keel can be used. 3) Open. Laryngotracheal reconstruction can be performed with or without a cartilage graft placed anteriorly or posteriorly. A stent or keel with a tracheostomy may be required during the healing process [8,9].

Conclusion
A congenital laryngeal web is a rare anomaly that can present with a variety of symptoms ranging from a weak dry since birth to respiratory distress. Due to the high association with 22q11 deletion, all patients should be tested . Treatment options include observation, endoscopic and open. Due to the range or severity of the web, some patients may present in adulthood. We present this interesting case to highlight the importance of keeping this diagnosis in the differential for hoarseness in adults.

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References