



An unusual cause of stridor: congenital laryngeal web

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Abstract

Respiratory distress and stridor are some of the common presenting symptoms for children in Pediatric Emergency Department. Most of these children are diagnosed as having common illnesses such as laryngitis, croup or laryngomalacia. However, Pediatric Emergency physicians must keep in mind other rare respiratory diseases other than laryngitis or croup in the differential diagnosis

of stridor. Stridor may occur due to congenital and acquired diseases. Laryngeal web is one of the rare congenital causes of stridor, which usually presents in the first weeks of life; however, it is very rarely diagnosed in the later period. Herein, we report a one-year-old boy who was evaluated for croup and was diagnosed as having laryngeal web.

Keywords: Child, croup, laryngeal web, laryngotracheobronchitis, stridor

Introduction

Stridor is generally an inspiratory, harsh, high-pitched sound that occurs as a result of upper respiratory tract obstruction. It may occur in relation to many congenital or acquired diseases. Stridor related to congenital causes is generally present at birth, but it may also occur during the days, weeks or months following birth. History is essential for the diagnosis. History taken in accordance with age may be helpful for physicians in the differential diagnosis of conditions leading to stridor. Laryngeal webs frequently lead to stridor in the first months of life (1). However, patients diagnosed in the late period have also been reported (2-4).

In this article, we present a one-year-old male patient who presented to the emergency department because of stridor was considered to have laryngotracheobronchitis (croup), and was subsequently diagnosed as having laryngeal web.

Case

A previously healthy one-year-old male patient presented to the emergency department because of cough and hoarse voice. In the history, it was learned that he had presented to our hospital two days previously with the same symptoms, and was diagnosed as having croup after evaluation and given inhaled epinephrine, salbutamol, and oral dexamethasone treatment and was discharged when his symptoms improved. The patient, who had no pathology in his personal and familial history, was born by vaginal delivery with a birth weight of 3050 g and was discharged from hospital after delivery without development of any adverse events. There was no history of intubation or ventilator treatment. His neurodevelopmental steps were compatible with his age. On physical examination, his general status was moderate, his consciousness was clear and his cooperation was full. Oxygen saturation was found to be 97% at room temperature, his body temperature was found to be 36.5°C, his respiratory

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Picture 1. The patient's anteroposterior lung graphy

rate was 30/min, and his blood pressure was 100/60 mm Hg. He had inspiratory stridor and subcostal, intercostal and suprasternal retractions. He had no cyanosis. A cardiac examination was found to be normal. The patient was given 1/1 000 inhaled L-epinephrine (0.5 cc/kg, maximum 5 mg) and oral dexamethasone treatment (0.6 mg/kg). However, investigations directed to the differential diagnosis of stridor were initiated because he presented to the emergency department again with the same symptoms. Lung radiography was obtained to exclude foreign body aspiration because there was no history of fever, his symptoms started suddenly at the second presentation, and treatment given was not helpful. Increased aeration, infiltration, atelectasis or pen-point appearance was not found on lung radiography (Picture 1).

The division of otolaryngology was consulted for a fiberoptic evaluation of the upper respiratory tract because there was no response to the inhaler and antiinflammatory treatment. With this procedure, it was planned to exclude infectious and congenital diseases involving the supraglottic region. The patient, who was diagnosed as having laryngeal web as a result of evaluation under general anesthesia, was internalized in the pediatric ward. Laryngeal web or additional airway anomaly had not been found on cervical tomography performed before surgery. He underwent surgery one day after hospitalization. On direct examination with laryngoscope during surgery, a web was found just below the vocal cords leading to obstruction in the subglottic region. It was opened with the assistance of forceps and scissors and bleeding control was provided. The patient, who

had no problems after the procedure, was discharged on the second day of hospitalization. The patient's family was informed that the patient was going to be presented as a case presentation and written informed consent was obtained.

Discussion

Stridor or a high-pitched breath sound is one of the most important symptoms indicating upper airway obstruction. Whatever the underlying reason, the patient's clinical status may vary according to the place and severity of airway obstruction causing stridor. Therefore, these patients should be evaluated very rapidly in the emergency department. Stridor may develop as a result of many congenital or acquired diseases. In the emergency department, the most common cause of stridor is croup and foreign body aspiration. In addition, bacterial tracheitis, epiglottitis, peritonsillary abscess and retropharyngeal abscess, trauma, and subglottic hemangiomas may also lead to stridor. In young children, enlarged tongue, laryngomalacia, and congenital goiter syndrome are other causes of stridor (5).

In patients with stridor, the diagnosis is made with history and physical examination. Laboratory investigations are rarely used for the diagnosis. Croup is one of the most common infectious airway emergencies leading to upper airway obstruction. It frequently develops in relation to parainfluenza virus infections and occurs in children aged between 6 months and 3 years (5). It is important to interrogate the age of the patients presenting with stridor and to make the differential diagnosis of stridor by age. Making a diagnosis of croup without detailed history and physical examination in a patient aged below six months who presents with stridor or noisy breathing will lead to missing of the other diseases. In treatment, relieving the airway is of top priority in patients with respiratory distress. However, patients will be discharged from the emergency department after a transient clinical improvement and present to emergency department again with similar symptoms if the airway obstruction is treated with antiinflammatory drugs without a detailed history and physical examination. Knowing in which age groups the clinical conditions leading to stridor occur will be helpful in the differential diagnosis, in selecting the laboratory tests appropriate for the diagnosis, and in specifying treatment options.

Patients must be hospitalized and monitored following the initial treatment for croup in cases where the history indicates severe airway obstruction before presentation, previous episode of croup or known airway disease (for example, subglottic stenosis), readmission to the emergency department within 24 hours after the initial treatment, the patient's being aged below six months, presence of stridor at rest, inadequate fluid intake, inability to obtain adequate response to initial treatment, high anxiety level in parents, the patient's house being distant from hospital, and with uncertainty about the diagnosis (6). In our case, it was planned to exclude congenital causes leading to stridor because our patient's clinical status did not improve with treatment.

Congenital laryngeal webs, which constitute 5% of all laryngeal congenital lesions, are one of the rare airway anomalies. They occur with incomplete recanalization of the primitive larynx during embryogenesis. The frequency of laryngeal web is approximately one in every 10,000 births. Most congenital webs are diagnosed at birth or in the first few months of life. Approximately 75% are observed at the glottic level and the rest are in the supraglottic or subglottic region. Accompanying congenital anomalies, especially subglottic stenosis, which is mostly observed in the upper part of the airway, are present in 10% of patients. The webs frequently involve the anterior part of the glottis. The thickness of the tissue in this region may show a difference (1). It may extend towards the subglottic region. Thick webs may be observed on lateral airway radiography.

Before treatment, the airway should be endoscopically visualized both to determine the severity of the web and whether it extends to the subglottic region, and to make the diagnosis and plan treatment (1).

Treatment depends on the degree of airway obstruction at the glottic level. Treatment is necessary if the web closes more than 50% of the glottic gap (1). Webs can be treated by different ways. Endolaryngeal techniques including dilatation, lysis with laser and stent are used for treatment of glottic and subglottic webs in cases where subglottic stenosis is absent (7). The laryngeal web in our patient was opened using forceps and scissors after visualization with direct laryngoscopy and the patient was discharged from hospital two days after the procedure.

A small number of patients diagnosed as having laryngeal web in the late period have been reported in the literature. A portion of the patients were diagnosed with hoarse voice starting from birth or speech problems and others were asymptomatic and diagnosed with sudden respiratory distress (2, 4). In addition, there are also patients who were diagnosed with occurrence of problems during anesthesia while undergoing surgery for another reason (3, 8, 9). The acquired causes of laryngeal webs, especially at advanced ages, include gastroesophageal reflux, smoking, previous surgery, and a history of intubation or infection (10). However, our patient had no such history.

In conclusion, this patient was presented to emphasize that stridor may be a finding of many conditions involving the upper airway; not all patients with stridor have croup. The patient's age, presence of accompanying findings, and response to treatment administered should be the essential indicators for differential diagnosis. Although laryngeal webs are generally diagnosed in the first few months of life, they should also be kept in mind in older children.

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