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Case Report

Idiopathic cervical tracheal stenosis in an 11-year-old male

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ABSTRACT

Idiopathic laryngotracheal stenosis is a fibroinflammatory stenosis in persons without alternative explanation such as trauma, intubation, or autoimmune disease. Patients are usually females of child-bearing age. We report on an 11-year-old male who developed progressive dyspnea and stridor. Bronchoscopy revealed 90% stenosis of the cervical trachea. He underwent serial balloon dilation with steroid injection; stenosis decreased to 20%. He had no history of intubation or trauma. Histologic and laboratory workup for autoimmune disease was negative. This report highlights occurrence of a well-known disease in an uncommon population, and alerts providers to consider this when seeing new pediatric patients with symptoms of progressive airway restriction.

1. Introduction

Idiopathic laryngotracheal stenosis (ITS) is a fibroinflammatory disease resulting in narrowing of the airway at the cricoid and/or proximal trachea [1,2]. It was initially described by Brandenburg in 1972 [3] and represents a diagnosis of exclusion when common etiologies such as intubation, granulomatosis with polyangiitis (formerly Wegener's disease), trauma, or prior surgery have been excluded [4]. Etiology is not known, but may potentially be related to estrogen [5], gastroesophageal reflux disease [6], or disruption of local blood supply secondary to telescoping of the first tracheal ring within the cricoid cartilage [7].

The vast majority of cases occur in females of childbearing age [8]. In reviewing multiple prior patient series, 181 of 184 patients were female with typical mean age of 40–50 years [4,7,9–13]. Notably, there were two outlying female patients aged 13 and 15 years [4,9]. These patients were not separately discussed.

Laryngotracheal stenosis occurs in children, but is typically congenital or related to prior intubation or trauma. Idiopathic laryngotracheal stenosis in this population is extremely rare. In addition to the two aforementioned patients in the larger case series, three single patient case reports are described of idiopathic subglottic stenosis occurring in children. Jazbi et al. reported on a 2.5-year-old male in 1977 who presented with one year of progressive cough, stridor, and mild respiratory distress [14]. Bronchoscopy showed an isolated subglottic stenosis. Rigid dilation was performed. The patient developed recurrent symptoms requiring tracheotomy, but was able to be decannulated five months later with no further sequelae. There is some suspicion, given the young age at presentation, the stenosis may have been secondary to a developmental defect of the cricoid cartilage or conus elasticus [13]. Bodart et al. reported on a 9-year-old male who presented with progressive dyspnea with exertion [15]. Bronchoscopy showed false membranes with inflammatory granulomas at the upper trachea. The granulomas were removed and systemic steroids were administered. Histology demonstrated non-specific inflammatory granulation tissue with epidermoid metaplasia, in contrast to the non-inflammatory pearl-like appearance seen in congenital subglottic stenosis [15]. He underwent carbon dioxide laser incision of the stenosis two months later for progressive symptoms followed by budesonide nebulizers for recrudescent symptoms with good effect. Modgil et al. reported on a 12-year-old female who presented with six months of progressive inspiratory stridor and dyspnea with exertion not relieved by bronchodilators and inhaled steroids [16]. Interestingly, symptoms began with menarche and worsened with menstruation. Bronchoscopy showed a 75% concentric subglottic stenosis. Omeprazole was given with no improvement in symptoms. Tracheotomy was then performed, followed by carbon dioxide laser incision and mitomycin-C injections. The patient was decannulated six weeks later. There were no further reported complications.

In addition to idiopathic subglottic stenosis, idiopathic tracheal stenosis can also occur. This is a rare condition in pediatric patients with few prior published reports. Beddow et al. reported on two 11-year-old patients with symptoms of progressive airway restriction who were found to have tracheal stenosis without identifiable cause and were managed with open surgery [17]. Watson et al. described the case of an 8-year-old with progressive cough, dyspnea, and stridor managed

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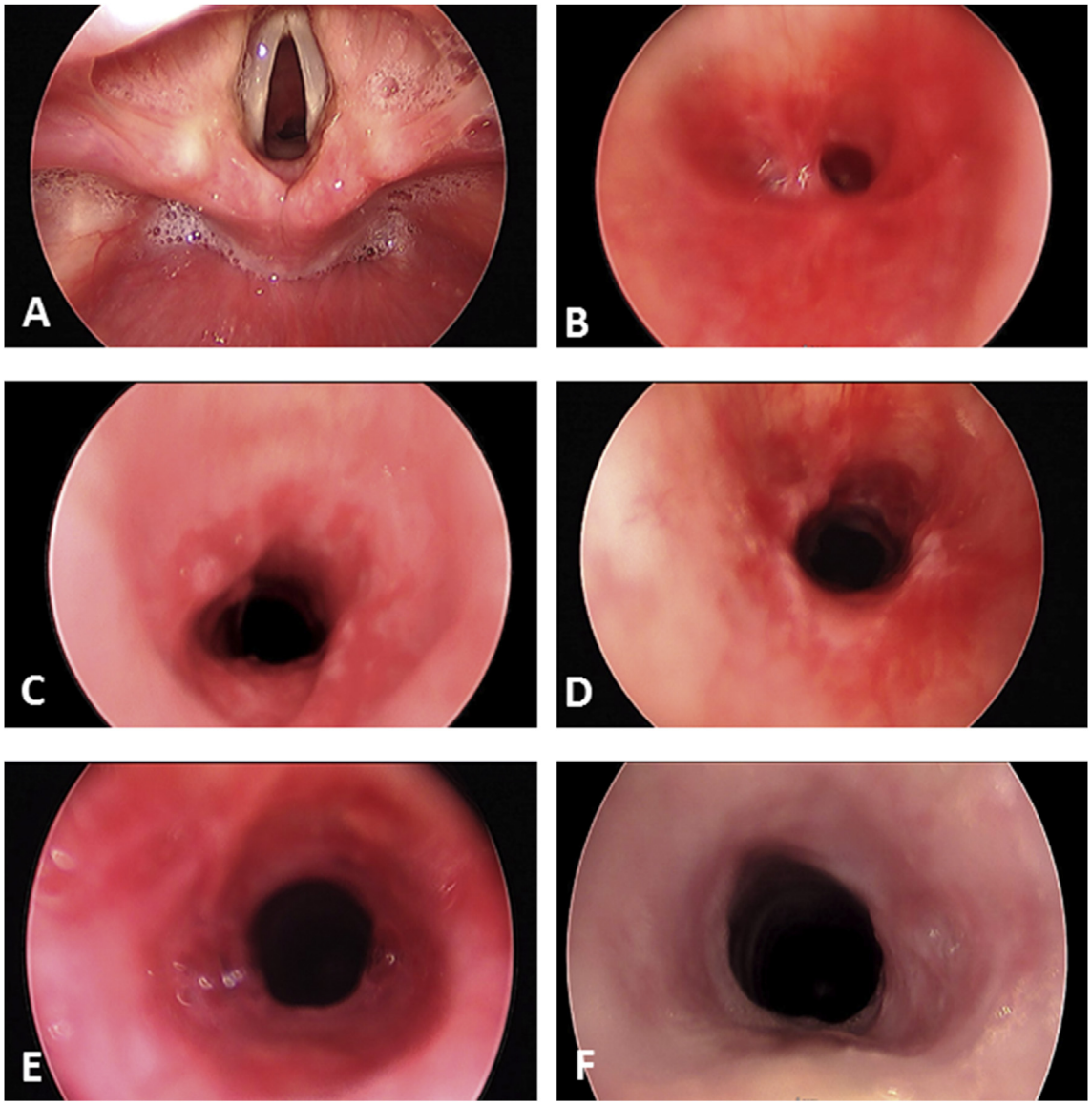

Fig. 1. Rigid telescopic views demonstrating glottis at time of first exam (A), proximal cervical trachea at time of first exam with 90% stenosis (B), at conclusion of first procedure (C), at beginning of second procedure (D), at conclusion of second procedure (E), at two-week follow-up (F), and at six-week follow-up (G).

with endoscopic dilations and high-dose steroids [18].

We report on an 11-year-old male who presented with progressive dyspnea and stridor and was found on bronchoscopy to have 90% concentric stenosis of the cervical trachea on bronchoscopy with negative workup for alternative etiologies for subglottic stenosis. This report adds to the small body of literature on pediatric idiopathic laryngotracheal stenosis and raises awareness of this disease entity.

2. Case report

An 11-year-old male presented to his family practitioner with one month of progressive dyspnea and two weeks of noisy breathing. He had no history of prior intubation, neck trauma, recent infectious disease, or autoimmune disease. Expiratory peak flow was 100 L/min (normal value for height: 370 L/min) that did not change after an albuterol nebulizer. He was prescribed albuterol, prednisone, and fluticasone nasal sprays. Symptoms did not improve over the next three weeks. A chest x-ray was unremarkable. A referral to otolaryngology was made. Five days later, symptoms worsened acutely and he presented to the emergency room with labored breathing. He was given albuterol, intravenous steroids, racemic epinephrine, and antibiotics without significant improvement. He was transferred to the university hospital where otolaryngology was consulted. In the emergency room, he exhibited audible biphasic stridor with tracheal tugging. There was no significant dysphonia, but speech cadence was interrupted. Flexible fiberoptic laryngoscopy did not reveal any supraglottic or glottic abnormalities. A white blood cell count was normal. The patient was taken urgently to the operating room for direct microlaryngoscopy and bronchoscopy.

Rigid bronchoscopy revealed a concentric stenosis (Fig. 1), obstructing approximately 90% of the cervical trachea just inferior to the cricotracheal junction (Fig. 1). Given the degree of stenosis, no formal airway sizing was performed prior to dilation. The patient was placed into suspension and serially dilated with 5, 7, and 10 mm balloons. After dilation, approximately 0.4 ml triamcinolone (40 mg/ml) was injected into the area of fibrosis. At the conclusion, there was a leak at 20 cmH_2O with a 4.0 endotracheal tube and no leak with a 4.5 endotracheal tube. The patient was extubated and taken to the intensive care unit for monitoring. He was given intravenous steroids, budesonide nebulizers, dual anti-reflux therapy, and humidified oxygen.