CLINICAL VIGNETTE

Idiopathic Subglottic Tracheal Stenosis, an Unusual Cause of Dyspnea during Pregnancy

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Case Report

A 36-year-old female with refractory dyspnea was initially seen in her thirteenth week of pregnancy. She was referred by her allergist and had a history of asthma for at least six years, requiring occasional use of an albuterol inhaler. She noticed increasing dyspnea on exertion for the past five weeks, which she attributed to her pregnancy. The patient denied any fever, productive cough, orthopnea, or chest pain. Prior to her pregnancy, she had symptoms compatible with gastroesophageal reflux for which she had been taking ranitidine with subjective relief. The patient denied any previous surgeries or anesthesia. She complained of some wheezing but only on exertion. Her symptoms did not improve with increased use of her rescue medication or with the addition of nebulized budesonide.

Physical examination revealed normal vital signs and pulse oximetry. Head and neck examination was normal with no evidence of tracheal deviation, adenopathy, or stridor. Auscultation of lungs revealed normal bilateral breath sounds with faint bilateral expiratory wheezes. Cardiac examination was unremarkable. Abdominal examination was compatible with her first trimester pregnancy.

Laboratory data included normal comprehensive metabolic panel and CBC. Spirometry revealed mild restrictive changes with a forced vital capacity (FVC) of 71% predicted and a forced expiratory volume/1 sec. (FEV1) of 76% of predicted. Fractional excretion of nitric oxide was normal. There was no arterial oxygen desaturation on ambulatory oximetry. Previous allergy testing did not reveal significant environmental atopy.

The patient was treated with a course of prednisone and nebulized albuterol and returned in follow up one week later stating that her wheezing had resolved but with no improvement in her dyspnea on exertion. Physical examination confirmed resolution of wheezing. Shielded one view CXR was normal. Repeat spirometry showed improvement in FVC and FEV1 to 82% and 83% of predicted respectively. Flow volume loop (Figure 1) showed findings suspicious for fixed large airway obstruction.

Consultation with perinatology was obtained with recommendation for CT imaging preferable to endoscopic procedures requiring conscious sedation. Computed tomography of the soft tissues of the neck revealed an area of subglottic stenosis with measured minimal luminal airway diameter of 6 mm, approximately 40% of normal. Length of the lesion was 7 mm. Sedimentation rate was normal. Antinuclear antibody and antineutrophil cytoplasmic antibodies were not detectable.

The patient was referred to Head and Neck Surgery. Perinatology recommendations favored intervention in the second trimester of pregnancy. Decision was made for intervention based on the patient's worsening symptoms as a temporizing measure until term delivery. In her 21st week of gestation, the patient underwent micro direct laryngoscopy with balloon dilatation and laser excision of subglottic stenosis with significant improvement to the stenotic area without complications.

The patient was significantly improved at discharge but slowly developed recurrent dyspnea with fiberoptic evaluation revealing recurrent stenosis. She underwent repeat balloon dilatation and laser excision of the stenotic area in the 35th week of gestation. Successful cesarean section delivery without complications was performed in the 38th week of pregnancy.

Discussion

Idiopathic subglottic stenosis (SGS) is a rare entity first described in 1972.[1] The disorder affects women (98%) mainly between the ages 30-50 years with 95% of Caucasian descent.[2] Idiopathic subglottic stenosis is felt to represent approximately 15% of all cases of subglottic stenosis.[2] The etiology of idiopathic SGS is uncertain. Given the almost exclusive incidence in women, hormonal factors such as the paucity of estrogen receptors from the areas of stenosis leading to increased release of fibroblast growth factor have been postulated.[3]

Trauma is the most common cause of SGS with 90% of all cases of acquired SGS due to tracheal intubation and tracheostomy. The reported rate of stenosis ranges from 1-10%.[4] Other etiologies described in the literature include tuberculosis, sarcoidosis, chemical or thermal burns, and Wegener's granulomatosis.[5,6] Gastroesphageal reflux has been postulated,

although several studies show lack of progression despite documentation of persistent reflux.[7,8]

The most common presentation is dyspnea. Patients are frequently misdiagnosed with asthma or chronic bronchitis, often with a history of poor response to bronchodilator and corticosteroid therapy.[9]

Symptoms are often exacerbated by pregnancy. This is attributed to increased minute ventilation of up to 150% of normal. Mucosal hyperemia with edema due to increased estrogen levels has also been suggested.[10]

Late recognition of this disorder in pregnancy is common with inability to intubate or adequately ventilate the patient during induction for cesarean section.[11]

Spirometry with flow volume loops give valuable information in assessing possible upper airway obstruction. Flattening of both inspiratory and expiratory curves suggest that further studies, including CT imaging and endoscopic evaluation, may be warranted.[12]

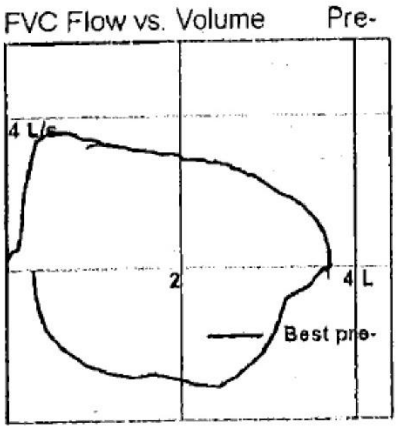
Management of SGS requires close coordination with Head and Neck surgery and anesthesiology, given the compromised airway. Available modalities include balloon dilation,[13] laser and corticosteroid injections. Surgical options include segmental resection with graft.[14]

Management of SGS during pregnancy poses significant risks to mother and baby often due to delayed diagnosis, which is usually made during difficult intubation for cesarean section. In our patient, diagnosis was made sufficiently early in the pregnancy to allow coordinated care between surgical, anesthesia, perinatology, and obstetrics services to facilitate delivery of a healthy infant.

In summary, subglottic stenosis should be considered in any patient with unexplained dyspnea or a diagnosis of asthma refractory to corticosteroids and bronchodilators. Symptoms may worsen during pregnancy secondary to increased minute ventilation and mucosal edema. Spirometry with special attention to flow volume loop is a useful screening tool to evaluate this disorder.

Figures

Figure 1. Flattened inspiratory and expiratory flow volume loop as seen in fixed large airway obstruction



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