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An Interdisciplinary Approach to the Management of Idiopathic Subglottic Stenosis in Pregnancy

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Abstract

Background: Idiopathic subglottic stenosis (ISS) accounts for 18.5% of acquired causes of subglottic stenosis in adults. To date, there have been 11 cases reported of ISS in pregnancy. ISS in pregnancy poses a diagnostic challenge due to the presentation of symptoms similar to other airway conditions without a clear underlying cause.

Case: We report a case of pregnant women who presented with progressive respiratory distress due to severe subglottic stenosis causing significant airway obstruction in her 2nd trimester pregnancy. An awake tracheostomy was carried out at 28 weeks followed by an elective cesarean section at term. Beginning two months postpartum, successful serial balloon dilations led to resolution of the restenosis and subsequent decannulation of the tracheostomy and resolution of the problem.

Conclusion: This case highlights the importance of considering ISS in a pregnant patient with upper respiratory symptoms as well as the team collaboration needed to diagnose and manage the patient's airway.

Keywords: Idiopathic subglottic stenosis; Pregnancy; Multidisciplinary

Introduction

Subglottic stenosis in adults can be acquired from various sources, the most common of which is iatrogenic from either prolonged intubation or tracheostomy (54.7%). Less common causes include idiopathic (18.5%), autoimmune (18.5%) and traumatic (8%) with the last 0.3% not specified [1]. With idiopathic subglottic stenosis occurring predominantly in females, there have been 11 reported cases of ISS in pregnancy [2-12]. Idiopathic subglottic stenosis in pregnancy poses a diagnostic challenge due to the presentation of symptoms similar to other airway conditions without a clear underlying cause. The stridor and shortness of breath commonly present can be mistaken as asthma or bronchitis. A multi-disciplinary approach is needed to both diagnose and manage the patient's airway to assure safety for the mother and baby during pregnancy and delivery. We report a case of a pregnant female who presented with progressive respiratory distress due to severe subglottic stenosis in her 2nd trimester of pregnancy that was originally managed as asthma. Treatment required the joint decision making of a multi-disciplinary team to successfully diagnose and manage her condition throughout and after her pregnancy.

Case Presentation

A 34 year old Caucasian female at gestation of 23 weeks and 1 day was admitted by her primary care practitioner for worsening shortness of breath and wheezing for 2 days. Over the last few months she noticed worsening breathing to the point where she could no longer run and would get breathless with talking. One year prior she had similar symptoms, though much milder, for which she underwent pulmonary function tests which showed very mild obstruction and a flow-volume loop consistent with a fixed intrathoracic obstruction due to flattening of the inspiratory and expiratory limbs. No treatment was needed at that time. On admission she presented with stable vitals and pulse oximetry above 93%. She denied any vaginal bleeding, loss of fluid or contractions. She was a nonsmoker and did not have allergies. She had been gaining weight appropriately for the pregnancy. Her medications included omeprazole and a multivitamin. Physical exam showed labored breathing with inspiratory and expiratory stridor.

The lungs were otherwise clear but with decreased breath sounds. She had dyspnea with talking and was unable to finish sentences. A posteroanterior chest x-ray was normal. A fetal ultrasound was performed which showed a single live intrauterine gestation with estimated age consistent with 23 weeks.

Upon admission, the patient was treated with albuterol (0.5%) nebulizer treatment every 4 hours as needed , Ceftriaxone 1 gram daily and methylprednisolone 40 milligrams intravenous every 12 hours. Pulmonology was consulted for asthma management; however due to suspicion for upper airway obstruction they ordered a computed tomography scan (CT) of the chest and consulted otolaryngology (ENT). CT chest with contrast and CT neck showed circumferential narrowing of the subglottic airway with clear lungs. The patient was transferred to the medical intensive care unit for airway monitoring. The ENT service performed an awake bedside flexible laryngoscopy with a 3.5 millimeter scope. Significant subglottic stenosis was seen in the airway with circumferential narrowing and inflammation. The airway size was estimated to be about 5 millimeters. Her steroids were increased to nebulized Decadron 2 milligram every 8 hours and methylprednisolone 80 milligram every 6 hours. Three days later another flexible laryngoscopy showed no progression of narrowing. Because of her clinical improvement and stable airway, it was decided to transition her intravenous steroids to oral prednisone 40 milligram every morning and monitor her as an outpatient. She was discharged on oral prednisone. ENT follow up was to be done weekly in the office.

At 28 weeks gestation, the patient presented to the ENT department with increased work of breathing. A flexible endoscopy suggested an airway of 2 millimeters or less, 3 millimeters narrower than it had been upon presentation at 23 weeks gestation. It was decided after discussion between otolaryngology, maternal fetal medicine, obstetrics anesthesia and the family that the best option was an awake tracheostomy to secure the airway before it became critically small due to the progressive stenosis. Two days later, a successful awake tracheostomy was performed and the patient was hospitalized for 5 days. During the procedure, a backup intraoperative fetal monitoring was performed and the obstetric team was present to perform an emergent caesarean section if needed. The patient was stable throughout the tracheostomy procedure and no obstetric intervention was needed. Her vitals remained stable with a pulse oximetry above 93% throughout. The patient continued with uneventful weekly visits to her high risk obstetrician.

At 39 weeks and 3 days, the patient underwent a successful elective cesarean section. The physicians and patient determined that she could not tolerate a normal vaginal delivery. Spinal anesthetic was used. A healthy male infant was born weighing 3375 grams with apgars 9/9. The patient recuperated from her cesarean section uneventfully.

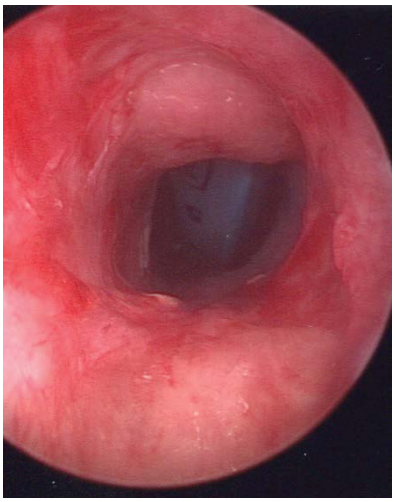

Figure 1: Endoscopic visualization of patient's subglottic larynx at 41 days post-partum before her 1st balloon dilation. The airway was measured to be 1 mm. The white illuminated structures on the far right and left of the image of the patient's vocal cords.

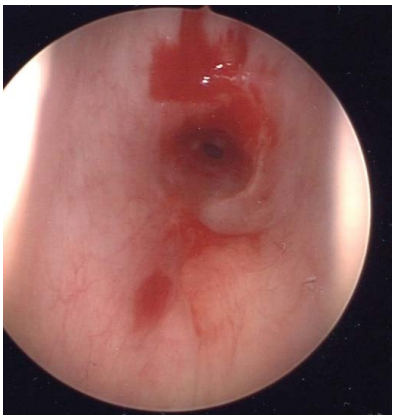

Figure 2: Endoscopic visualization of patient's subglottic larynx at 83 days post-partum, 4 weeks after her second balloon dilation. The airway is fully patent.

On postpartum day 41, the patient underwent an outpatient elective microlaryngoscopy, bronchoscopy and dilation. At the distal end of the subglottis there was circumferential narrowing of the airway to a pinhole of about 1 mm which was the narrowest it had been measured (Figure 1). The opening was dilated with 8.5 and 10 millimeters diameter high pressure balloons and 5 milliliters of methylprednisone was injected endoscopically into the area of stenosis. Endoscopy showed normal mid and distal trachea, carina and bronchi.

On postpartum day 52, the patient underwent the same procedure. The subglottic region was fairly patent and remucosalized and another balloon dilation was performed. At 83 days postpartum, the patient underwent a microlaryngoscopy and bronchoscopy showing a patent fully healed airway/ subglottis with no obvious stenosis (Figure 2). The tracheostomy tube was removed with office follow up schedule in one week. Two months after decannulation the old tracheostoma was healed and the patient returned to a normal breathing pattern and to running 3 miles per day for exercise.

Discussion

Idiopathic subglottic stenosis is a rare occurrence in pregnancy and may often be initially misdiagnosed as asthma, as was the instance in this case. Therefore, the need for specific diagnostic criteria of ISS is significant. Specifically, unique onset of wheezing or stridor and dyspnea during pregnancy is a good indicator of ISS inclusion as a differential diagnosis [11]. Due to its low prevalence, there is not a universally accepted treatment plan for ISS, during pregnancy and many options for management exist [13]. The lack of standardization of care of ISS is of particular concern in pregnant ISS patients, who undergo varying procedures resulting in differing outcomes. This report analyzes different treatment options in pregnant ISS patients; as each patient must be treated based on her presentation and degree of stenosis, with consideration of time frame of gestation and ultimately considering the safety of the mother and baby. These treatment options include observation, observation with systemic steroid treatment, laser therapy via bronchoscopy, endoscopic airway dilation, tracheostomy and/or open surgical resection of the stenosis. In particular, studies maintain that tracheostomy under local anesthesia is the safest option for airway maintenance prepartum [4,8,9]. In fact, Parsa et al explain that maintaining a prepartum tracheostomy with local anesthesia is the best option to not only maintain an airway should the need for surgery arise, but to also prevent complications like hypercarbia and hypoxemia of the fetus [9]. Another study confirms prepartum laser incision of the stenotic site followed by balloon dilation under local anesthesia or conscious sedation as a safe option [11].

Due to the rare nature of ISS, especially in pregnancy, it is important to review the literature in an attempt to establish a basis for obstetric outcomes in patients with ISS. To our knowledge, ours is the first comprehensive study on ISS in pregnant patients to conduct such a review. There are eleven known case studies on ISS in pregnant patients. Including our case, prepartum tracheostomy was performed in 3 cases [4,8]. Postpartum tracheostomy was performed in 2 cases [3,6]. Laser excision or surgical resection was performed in 4 cases [2,3,9,11]. All patients delivered healthy babies and recovered fully from ISS. However, one study did not specify delivery method or outcome for the patient and fetus [2]. Due to this fact and the variety of treatment options, we suggest that an interprofessional clinical team is essential in order to address each patient's specific case of ISS in pregnancy. Further, the delivery method (vaginal or cesarean), was highly dependent on the presentation of ISS in the patient. Therefore, as suggested by our study, the decision for cesarean or vaginal delivery should be a multidisciplinary discussion with the patient, ENT, and obstetrics.

It is theorized that there is hormone involvement in the pathogenesis of the idiopathic subglottic stenosis [14-16]. This may account for the female predisposition for the condition with reports of the condition occurring in females ranging from 87.5% [16] to 98% [17] of the time. During pregnancy, hormonal changes alter many organ systems with the airway being one of them. In particular, due to high estrogen levels in pregnancy, there is an increase in total body water and mucosal edema of the vocal cords, larynx, and trachea [4,11]. Therefore, it is important to take into consideration that the airway is among the many systems in the woman's body that undergoes change during pregnancy. Our patient most likely had some element of existing idiopathic subglottic stenosis which was exacerbated by her pregnancy. Further, our patient's pulmonary function tests prior to pregnancy demonstrated flattening of the inspiratory and expiratory limbs of the flow-volume loop representing very mild obstruction and an intrathoracic obstruction.

When the airway does undergo pathological changes in pregnancy resulting in ISS, great care must be taken to assure safety for the mother and child. After diagnosis, the otolaryngologists worked closely with both obstetric and obstetric anesthesiology teams to determine the treatment plan. For instance, ENT initially considered subglottic dilation under general anesthesia. However, the anesthesiologist and obstetric teams proposed to avoid general anesthesia based upon the maternal airway and difficulties that may arise due to the subglottic stenosis. In particular, unsuccessful intubation attempts of a ISS patient under general anesthesia can lead to anoxia as well as tracheal or laryngeal cartilage damage [3]. Further, maternal respiration and airway undergo

significant changes during pregnancy that may contraindicate general anesthesia, especially due to the ISS. For instance, by the third trimester of pregnancy the basal metabolic rate is increased by 10 to 20% in the mother compared to non-pregnant patients, in order to respond to the increasing demands of the growing fetus and placenta [18]. Oxygen consumption increases approximately 20% during pregnancy as well [19]. These changes make the airway of vital importance and an acutely hypoxic episode could have detrimental effects on the growing fetus. In addition, if an airway emergency occurred during general anesthesia or at delivery, it would be particularly difficult to manage in pregnancy due to the physiological changes of a decreased functional residual capacity due to diaphragm elevation and decreased blood return to the right ventricle due to the weight of the fetus on the inferior vena cava [20]. Therefore, the interdisciplinary team played a vital role in discussing treatment options and managing the patient's condition.

Therefore, via a multidisciplinary discussion, it was decided to perform an elective awake tracheostomy under local anesthesia with minimal sedation prior to delivery and a balloon dilation afterwards. This solution avoided the complications of general anesthesia and any chance of a hypoxic episode or airway emergency. It also provided a solution during pregnancy to maintain a patent airway and allow for the safe uneventful delivery of a healthy baby [21].

Conclusion

This case highlights the importance of considering ISS in pregnant patients with upper respiratory symptoms as well as the multi-disciplinary team collaboration needed to diagnose and manage a complicated situation such as this. All team members were involved in the decision making process at every step of the way and were able to voice their opinion comfortably knowing that everyone's perspective was needed and valued.

Teaching Points

1) In pregnant patients with airway symptoms such as stridor, wheezing or dyspnea on exertion, idiopathic subglottic stenosis in pregnancy should be considered in the differential diagnosis due to the severe complications that may result to the mother and baby if unrecognized.

2) For best outcomes, a multidisciplinary team is needed to best manage the mother and baby during the pregnancy if the mother develops ISS. This includes maternal fetal medicine, otolaryngology and anesthesiology.

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