Idiopathic subglottic stenosis recurrence during multiple pregnancies: A case report

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CLINICAL NARRATIVE

Introduction: Idiopathic subglottic stenosis (ISS) is a type of central airway obstruction primarily affecting adult Caucasian females. Its rarity and similar clinical presentation to asthma contribute to its frequent misdiagnosis. Timely treatment of ISS is crucial for pregnant patients who are at risk for emergent surgeries necessitating ventilation and intubation.

Case: A 29-year-old, previously healthy female reported new onset dyspnea on exertion and a dry, non-productive cough to her family physician. Clinical exam was unremarkable; however, pulmonary function testing revealed a moderate, fixed airway obstruction. Endoscopy visualized subglottic stenosis 1.7 cm distal to her vocal cords and surgical intervention using CO₂ laser dilatation was scheduled with Otolaryngology. However, the patient received a positive pregnancy test prior to her elective surgery date and after consideration of risks and benefits to mother and fetus, the surgery was postponed to her second trimester. Unfortunately, the patient miscarried her fetus and was scheduled for an emergent dilatation and curettage (D&C). She was successfully bag-mask ventilated during the D&C and subsequently underwent surgery for ISS with full resolution of her symptoms. In subsequent follow-up, the patient reported a return of dyspnea coinciding with another positive pregnancy test. ISS recurrence was diagnosed; however, as symptoms were mild, a repeat surgery was postponed until the patient safely delivered her child.

Discussion: This case illustrates the difficulties in the diagnosis and management of ISS in pregnant patients during their childbearing years as surgical intervention will impact both the mother and fetus.

INTRODUCTION

Subglottic stenosis is a type of central airway obstruction characterized by a circumferential fibrotic narrowing in the subglottic larynx and upper trachea and is associated with several conditions, including a history of intubation, sarcoidosis, gastroesophageal reflux, and Wegener granulomatosis (WG).^{1,2} In the absence of any identifiable cause, the condition is called idiopathic subglottic stenosis (ISS), a rare condition mostly affecting adult Caucasian females.³ Symptoms may include dyspnea on exertion, dry cough, and stridor. Due to the rarity of its presentation, patients are frequently misdiagnosed with asthma and/or COPD.¹⁻³ There is scarce literature on ISS encountered during pregnancy,^{1,2,4-6} however, its timely treatment is crucial in avoiding the dangerous “cannot ventilate, cannot intubate” scenario during anesthesia in emergent surgeries. We describe the management of a unique patient case who experienced ISS recurrences in the context of multiple pregnancies.

CASE REPORT

A 29-year-old G1P1 from Windsor, Ontario presented to her family physician with new shortness of breath on exertion and a dry, non-productive cough since the birth of her son 8 months ago. She reported decreased exercise tolerance affecting her work as a gym teacher and described the sensation of a “screen stopping her breathing”. She was previously healthy and has no history of asthma, environmental allergies, gastroesophageal reflux (GERD), or recent illness. Physical exam was benign and her lungs were clear to auscultation. She received a provisional diagnosis of asthma and was prescribed bronchodilators. In addition, chest x-ray and pulmonary function testing (PFT) were ordered. PFT indicated decreased predicted FEV1 value and decreased FEV1/FVC ratio (64% and 53.55% respectively) that did not respond to bronchodilator challenge. This was consistent with a clinical picture of a moderate, fixed airway obstruction. She was referred to Otolaryngology for further investigations and an endoscopy visualized a subglottic stenosis 1.7 cm distal to her vocal cords. The patient denied any history of intubation or surgery and serology for Wegener’s Granulomatosis (WG) was negative. At this time, her symptoms had worsened to the point of biphasic stridor at rest. The patient was diagnosed with idiopathic subglottic stenosis (ISS) and referred to a London ENT specialist for surgical intervention using CO₂ laser dilatation. During the wait for her booked surgery, the patient received a positive pregnancy test. Her clinical condition was reassessed by the ENT specialist and the elective surgery was deferred until her 2nd trimester. Unfortunately, the patient soon spontaneously miscarried her fetus and underwent dilatation and curettage (D&C). She was successfully bag-mask ventilated under general anesthesia during the procedure. The patient was subsequently treated for her ISS using CO₂ laser dilatation with microlaryngoscopy and her symptoms resolved (Fig. 1).

Figure 1. Endoscopic view of patient's larynx and upper airway during microlaryngoscopy and CO₂ laser dilation of her idiopathic subglottic stenosis.

At her 6 months follow-up, she reported another positive pregnancy test and a return of exertional dyspnea. Endoscopy revealed a recurrence of her ISS although the stricture was less severe in comparison to previous presentation. Once again, surgical intervention was deferred and the patient was closely monitored throughout her pregnancy for any worsening of symptoms. Her second CO₂ laser dilatation treatment was performed months after an uneventful delivery. Regular biannual ENT follow-up has revealed no further return of respiratory complaints or stenosis.

DISCUSSION

Idiopathic subglottic stenosis (ISS) is a rare condition, found in an estimated 5% of subglottic stenosis etiology.⁵ ISS during pregnancy has only been described in a few case reports, usually in emergent surgical scenarios without information on follow-up management.^{1,2,4-6} This case report presents the first description of ISS management in a childbearing patient who experienced recurrence in her subsequent pregnancy.

ISS is frequently misdiagnosed as asthma in patients for many years due to their similar clinical presentations.¹ However, undiagnosed ISS in patients undergoing surgery may lead to a disastrous scenario of failed intubation and failed ventilation during general anesthesia. Therefore, clinicians should raise their index of suspicion when patients present with worsening dyspnea or stridor at rest that is unresponsive to bronchodilator therapy. Physical exam findings of wheezing or stridor over the neck compared to peripheral regions of the lungs and a lack of hoarseness of voice, dysphagia, or odynophagia are suggestive of a supra- or subglottic stenosis.^{1,7} Additional investigations such as spirometry and endoscopy are helpful in its diagnosis and further management.⁷ It is important that common causes of subglottic stenosis such as previous intubation injury, GERD, or WG are ruled out prior to a diagnosis of ISS. In this case report, the period between initial complaints of symptoms to final diagnosis represented the longest interval prior to her surgery (5 of 10 months).

ISS case series report ISS diagnoses almost exclusively in young and middle-aged women, at rates between 80% to 100%.⁷⁻¹⁰ There is currently no reported data on whether the incidence of ISS is higher during pregnancy. Although the etiology is not definitively known, current literature attributes this higher incidence in females to hormonal factors as alteration in wound healing response has been noticed under estrogen effects.⁹ Clinicians should be sensitive to the competing needs of this particular patient demographic as surgical treatment for ISS may interfere with patient considerations for family planning and childbearing. For an elective upper airway surgery, Health Quality Ontario advises a “Time to First Surgical Appointment,” the length of time patients wait for an initial assessment by surgeon following referral, and a “Time from Decision to Surgery,” the length of time patients wait for surgery upon decision to proceed, both to be within 182 days each.^{11,12} However, these metrics do not capture the total length of patient wait times, such as wait time for diagnostic testing or specialist-to-specialist referrals. While our patient had a “Time to First Surgical Appointment” of 20 days and a planned “Time from Decision to Surgery” of approximately 90 days, both well

within the Ontario recommended target times, a total of 10 months had lapsed since her initial complaint at the time of her positive pregnancy test. Thus, timely referral and treatment should be emphasized to minimize the impact that elective surgeries can have on childbearing female patients. At the same time, physicians should discuss with their female patients the impact that pregnancy has on planned surgical management.

ISS during pregnancy presents a unique management challenge and physicians must carefully weigh the risks and benefits of surgical intervention for both the mother and baby. The American College of Obstetrics and Gynecology (ACOG) in their 2017 Committee Opinion provided 3 recommendations for non-obstetric surgery during pregnancy: (1) A pregnant woman should never be denied indicated surgery, regardless of trimester; (2) Elective surgery should be postponed until after delivery; (3) If possible, non-urgent surgery should be performed in the 2nd trimester when preterm contractions and spontaneous abortions are least likely.¹³ General anesthetic risks to consider include a potential reduction in uteroplacental perfusion due to maternal hypercapnia leading to fetal acidosis as well as insufficient clinical evidence on the teratogenicity of anesthetic drugs.¹⁴ Our patient presented with a mild-moderate subglottic stenosis that did not pose immediate danger of respiratory compromise, thus surgery was postponed until her second trimester. However, as illustrated in our case report, D&C following a miscarriage is a pregnancy complication with urgent surgical indications. Therefore, physicians must assess ISS patients for safe ventilation capabilities prior to delaying surgical treatment in the event of an urgent surgical indication during pregnancy.

Surgical treatments of ISS may include endoscopic procedures, open neck surgery, or tracheostomy, though no optimal management has been established.¹⁵ The endoscopic approach with CO₂-laser dilatation is a minimally invasive procedure with improved tolerability and shorter hospital stays and has been recommended as a first-line treatment modality in patients with first presentation of less severe and anatomically simple ISS.^{16,17} However, patients are more likely to experience recurrence at rates between 55-80%,²⁰ thus requiring multiple interventions.^{8,17} In comparison, open laryngotracheal resection has significantly lower recurrence rates at <10%.¹⁸ Patients reporting recurrent ISS, particularly for those considering future pregnancies or other surgical interventions, should be considered for definitive open surgical intervention.

CONCLUSION

Our case report illustrates some of the challenges in the diagnosis and management of idiopathic subglottic stenosis (ISS) in pregnancy. Undiagnosed ISS poses a high risk for morbidity and mortality in the mother and fetus. Physicians should raise their index of suspicion in patients presenting with dyspnea unresponsive to bronchodilator treatment. ISS is predominantly diagnosed in females and the timely referral and treatment of symptoms is important in minimizing delays or conflicts between treatment and childbearing. Physicians should assess patients for ventilation capabilities in the event of emergent surgery before making a decision to delay surgical intervention of ISS during

pregnancy. More research is still needed in the development of clinical guidelines for ISS treatment. At present, the management of ISS in pregnancy requires a multidisciplinary approach with input from Otolaryngology, Obstetrics, and Anesthesia.

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