

Management of Achalasia in Pregnancy in a Resource-Poor Setting

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Abstract

Achalasia is a rare motility disorder of the esophagus. Of more rarity is its occurrence in pregnancy. The disease is associated with varying degrees of malnutrition, with intrauterine growth restriction and even fetomaternal death reported in worse cases. Its diagnosis is usually confused for hyperemesis gravidarum in early pregnancy, and diagnosis is thus often delayed until the disease is advanced. Its evaluation and treatment pose a challenge due to associated fetomaternal risks of management. A case of achalasia diagnosed in the second trimester of pregnancy was presented. The patient was evaluated and subsequently had laparotomy and modified Heller's esophagocardiomyotomy with a good outcome. Based on a review of the literature and own experience, an algorithm is proposed for its evaluation and treatment in a resource-poor setting.

Keywords: Achalasia, pregnancy, resource-poor setting

INTRODUCTION

Achalasia is a motility disorder of the esophagus characterized by the absence of antegrade peristalsis and loss of reflex relaxation of the lower esophageal sphincter in response to a swallowed food bolus. This occurs due to the loss of ganglion cells within the esophageal wall.^[1,2] This disease is associated with varying degrees of malnutrition, with intrauterine growth restriction^[1] and even fetomaternal death reported in worse cases.^[3,4] Patients usually present with intermittent dysphagia to both solids and liquids, regurgitation, heartburn, and weight loss. Its presentation, diagnosis, and treatment pose a challenge because of its attendant fetomaternal risks. A proposal is made on the evaluation and treatment of pregnant women with achalasia presenting in a resource-poor setting.

CASE REPORT

The patient was a 35-year-old gravida 4 para 3 + 0, 3 alive, with an estimated gestational age of 23 weeks four days, who presented with intermittent dysphagia of one-year duration. She had lost 35 kg (38% of her body weight) since the onset of illness. She also had significant regurgitation and occasional aspiration at night. Barium swallow (done with an abdominal shield) revealed a dilated esophagus with a bird-beak appearance of the esophagogastric junction and

smooth appearance of the esophageal mucosa [Figure 1]. Upper gastrointestinal endoscopy revealed a roomy esophagus with narrowing of the lower esophageal sphincter. The scope however navigated into the stomach after some difficulty. No esophageal tumor was noted. Before presentation to our facility, she had undergone three sessions of esophageal dilatations with short-lived relief of her symptoms. She was counseled on modified Heller's esophagocardiomyotomy via laparotomy. Obstetric review was sought.

She was fasted for 24 h before the operation and had copious esophageal lavage. Under general anesthesia and cuffed endotracheal intubation, she had modified Heller's esophagocardiomyotomy. At laparotomy, the patient had a 24 week size gravid uterus [Figure 2]. Myotomy extended from the distal 5 cm of the thoracic esophagus anteriorly to 3 cm on the cardia of the stomach, ensuring that all muscle bands were divided, with a rim of muscle excised longitudinally along the length of the myotomy to prevent muscle re-apposition. Necessary precautions were taken during the institution and

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Figure 1: Barium swallow revealing a “bird-beak” appearance of the lower esophagus

reversal of anesthesia to prevent aspiration. The scrub nurse, surgeon, and assistant were ready to commence operation as soon as under anesthesia to reduce anesthesia time and its effects on the gravid uterus. Attempts were made to minimize the handling of the uterus during operation. The patient developed intermittent uterine contractions from immediate to 48 h postoperatively, necessitating the administration of tocolytics by the obstetricians. A single 5-mm esophageal perforation was noted 1 cm above the cardia, which was repaired using vicryl 2/0 and buttressed with an omental patch. She commenced oral intake on the fourth postoperative day and has been free of dysphagia since then. At 38 weeks of gestation, she was delivered a healthy baby via elective cesarean section.

DISCUSSION

There are conflicting reports on the relationship between achalasia and pregnancy^[5] and may require further research to elucidate any relationship, if any. The symptoms of regurgitation, heartburn, and weight loss may mimic hyperemesis gravidarum and will require a high index of suspicion to consider achalasia.^[6] Thus, the disease is likely to be advanced by the time hyperemesis gravidarum is ruled out.^[5] However, a meticulous history may reveal the onset of symptoms dating back before pregnancy.

The classical esophageal manometric finding of a failure of the lower esophageal sphincter to relax in response to a swallow and absent peristalsis in the smooth muscle of the distal esophagus is the hallmark of its diagnosis.^[7] However, in our environment, due to the dearth of facilities for esophageal manometry, a barium swallow and esophagogastroduodenoscopy may suffice. Barium swallow reveals a dilated esophagus with smooth, tapered distal narrowing at the gastroesophageal junction (“bird-beak” appearance).^[7,8] According to the American College of Obstetricians and Gynecologists’ Committee on Obstetric Practice, with few exceptions and when necessary, radiation exposure through X-rays should be carried out at a dose much

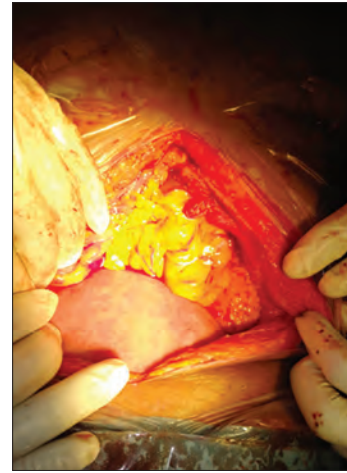


Figure 2: Gravid uterus visualized at laparotomy

lower than the exposure associated with fetal harm.^[9] In our environment, barium swallow remains pivotal in the diagnosis of achalasia and can be carried out with an abdominal shield with minimal fetal risks, as our patient had.

Definitive treatment requires esophagocardiomyotomy, which can be carried out via the thoracic, abdominal, or peroral endoscopic routes. In our environment, open thoracic and abdominal routes remain favored by many surgeons, with the laparoscopic route being on the increase.^[2,5] Reports of interventions for achalasia in pregnant women include medical treatment, Bougie dilatation, pneumatic dilatation, botulinum toxin injection, nasogastric and nasojejunal tube feeding, total parenteral nutrition, self-expanding metal stent, and laparoscopic Heller’s myotomy.^[1,5,10,11] These were all reported outside our subregion. Reports from our environment have not included pregnant patients.

From reports, it appears that many authors favor dilatation at any stage of pregnancy^[5] due to its minimal fetomaternal risks. Besides the need for repeated dilatations, a significant drawback of this modality is the potential risk of esophageal rupture, warranting the much-avoided surgery. Perforation rate is <1%–3%, with the initial dilatation posing the greatest risk compared to subsequent dilatations.^[12]

Nutritional rehabilitation can be offered via nasogastric/nasojunal tube feeding, percutaneous endoscopic gastrostomy/jejunostomy, or total parenteral nutrition. Nasogastric/nasojunal tube may be difficult to pass in pregnant women and may require endoscopy to achieve success.^[5,10] Nutritional rehabilitation may also be achieved via minilaparotomy under local anesthesia and insertion of a feeding gastrostomy/jejunostomy with minimal fetomaternal risks. Where severe malnutrition poses a great risk, nutritional rehabilitation appears logical. However, it suffices to note that nutritional rehabilitation may minimize but does not preclude the risk of regurgitation and aspiration associated with the disease.

Surgical intervention is considered safest during the second trimester of pregnancy as the risk of spontaneous abortion or

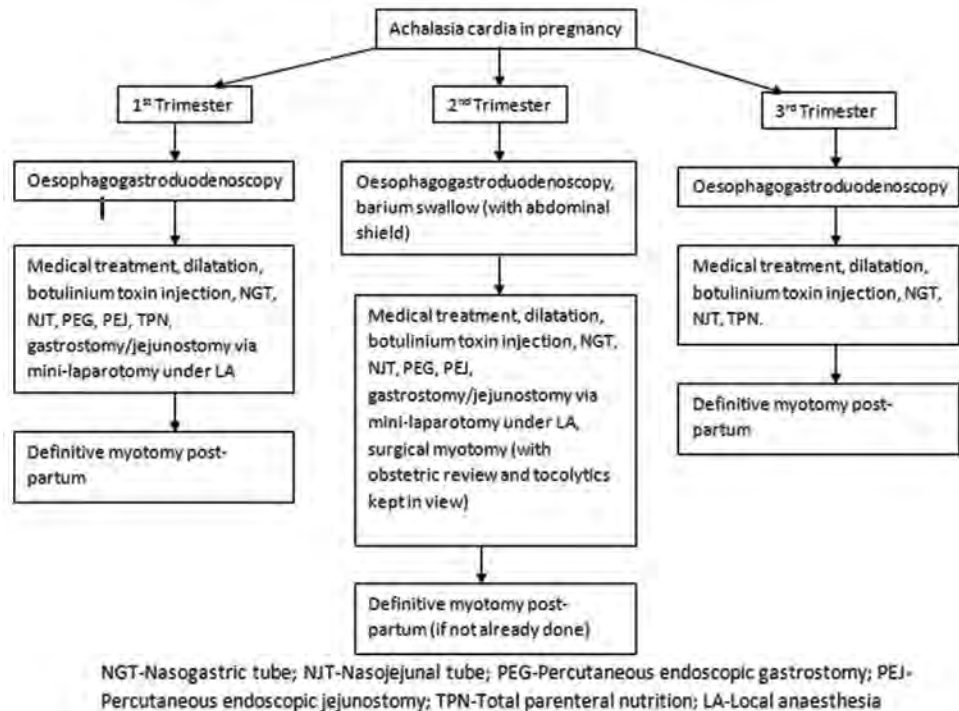


Figure 3: Proposed management algorithm for achalasia in pregnancy in a resource-poor setting

preterm delivery is less than 5% at this time.^[5] If carried out in the first trimester, abortion may occur as a result of the effects of anesthetic medication on fetal organogenesis. Likewise, surgery in the third trimester could precipitate labor in about 40% of patients.^[5] Before any surgical intervention is considered, an obstetric review should be done.^[13] The American College of Obstetricians and Gynecologists' Committee on Obstetric Practice recommends that corticosteroid administration for fetal lung maturation should be considered in patients with fetuses at viable premature gestational ages scheduled for surgery, and monitoring should be carried out postoperatively for symptoms and signs of preterm labor.^[13] Suffice to note that the risk of definitive myotomy lies with the exposure of the fetus to anesthetic medications. Being so, modes of regional anesthesia may theoretically be considered. Intercostal nerve blocks with local infiltration have been utilized for video-assisted thoracoscopic surgery, while epidural anesthesia or paravertebral block has been utilized for thoracotomy, all without general anesthesia.^[14] However, without cuffed endotracheal intubation, the risk of intraoperative regurgitation and aspiration persists. This risk can be minimized by copious preoperative esophageal lavage and suctioning, with consideration for the use of a supraglottic (laryngeal mask) airway in such awake patients in selected cases where necessary.^[15] Some authors have conducted planned delivery for very severe symptoms of achalasia when the fetus was considered viable.^[5]

In the light of the above, an algorithm for the management of achalasia cardia in a resource-poor setting is proposed [Figure 3].

CONCLUSION

Achalasia in pregnancy is a rare occurrence. The diagnosis, evaluation, and treatment pose a challenge due to the consideration of fetomaternal risks. The treatment option employed should be a shared decision, possess the lowest fetomaternal risk, as well as the availability of that modality in our subregion. Surgical intervention can be carried out with adequate multidisciplinary planning and acceptable risks in the second trimester. Obstetric consultation should be sought before any surgical intervention.

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Conflicts of interest

There are no conflicts of interest.

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