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Achalasia Cardia in Pregnancy Presenting as Hyperemesis Gravidarum in A Rural Kenyan Hospital: A Case Report and Literature Review

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ABSTRACT

Achalasia cardia is a motility disorder of the esophagus characterized by failure of relaxation of the lower esophageal sphincter (LES) during swallowing, resulting in dysphagia, regurgitation symptoms, vomiting, fluid losses, and malnutrition. Achalasia cardia is rare in pregnancy and is often misdiagnosed as hyperemesis gravidarum due to similar symptomatology, usually with significant morbidity and mortality. Definitive management is by surgical myotomy, which should be delayed until after delivery due to concerns about maternal and fetal safety. In this study, we present a case of achalasia cardia in a young primigravida in rural Kenya in the third trimester of pregnancy.

Keywords: *Achalasia cardia, Hyperemesis gravidarum, Dysphagia in Pregnancy, Barium esophagogram, Barium swallow, Heller myotomy, Kenya.*

INTRODUCTION

Achalasia cardia is a rare primary esophageal motility disorder in which there is progressive loss of ganglionic cells in the myenteric plexus of the walls of the distal esophagus and lower esophageal sphincter (Savarino et al., 2022). This results in loss of peristalsis in the distal esophagus and failure of relaxation of the LES during swallowing, thereby causing symptoms of dysphagia with solids and liquids, regurgitation of undigested food and saliva, heartburn, vomiting (which may cause some temporary relief), and retrosternal chest pain (Li et al., 2023). Achalasia cardia may be primary (idiopathic) or secondary. There are no known causes of idiopathic achalasia cardia, but secondary disease may be associated with any condition that causes loss of peristalsis in the lower esophageal sphincter region, including amyloidosis, gastric carcinoma, amyloidosis, Chaga's disease (caused by *Trypanosoma cruzi* and found in Central and South America (de Oliveira et al., 1995), eosinophilic esophagitis, etc. Achalasia cardia is even rarer in pregnancy, with less than 50 cases recorded in the literature since the first reported case in January 1964 by Karjalainen *et a*l (Barrett, 1964). Pregnancy itself may trigger achalasia cardia in women who are carriers of an insertion mutation in the HLA-DQβ1 region (Becker et al., 2016).

CASE SUMMARY

Presenting illness and physical examination

A 19-year-old, married lady from Narok, Kenya, was referred to us in mid-2021 with a history of refractory hyperemesis gravidarum in a primigravida. At the time, she was 29 weeks pregnant (gestation by date). She had had an insidious onset of nausea and postprandial vomiting since the 5th week of gestation, which had initially been managed with over-the-counter anti-emetics. By the 9th week of gestation, she developed worsening heartburn, regurgitation of eaten food, and vomiting that required repeated admissions for fluid resuscitation and intravenous antiemetics. The symptoms were recurrent and progressive, such that by week 26 of gestation, she was having associated dysphagia with both solids and semi-solids, with associated chest pains, progressive weight loss, and general lethargy.

Her clinical examination was significant for marked wasting, profound dehydration with hypotension; blood pressure 84/46 mmHg, heart rate 107 bpm, random blood sugar was 70 mg/dl, she was afebrile, and oxygen saturation by pulse oximetry was 94%. Her abdominal fundal height by palpation was 24/40 (discrepant from the gestational age of 29 weeks), she had marked epigastric tenderness but no peritonism, and the fetus had a normal regular heart rate of 136-148 bpm. Her chest had marked bilateral diffuse coarse crackles (aspiration events); she had normal heart sounds and a grossly normal neurological examination. She underwent urgent, comprehensive, and successful initial resuscitation.

Diagnostic work-up

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Her significant work-up included an elevated serum creatinine of 4.6 mg/dl, Na+ 141 mmol/L and K+ 3.1 mmol/L (features of acute kidney injury, which resolved in 48 hours with intravenous fluids), a normal complete blood count, and a normal antenatal care profile. Obstetric ultrasound showed a single live intrauterine pregnancy in cephalic presentation at 30 weeks and 2 days gestation with a heart rate of 144 bpm, an estimated fetal weight of 1.34 kilograms +/- 201grams, a reduced liquor volume with an amniotic fluid index of 7.13 cm and a biophysical profile score of 6/8. An upper gastrointestinal endoscopy showed a dilated esophagus with a tight lower esophageal sphincter (LES) (the scope was advanced with gentle force), gastric fundal erythema, and a normal duodenum. A subsequent barium swallow/esophagogram with serial imaging showed proximal esophageal dilatation tapering towards the esophagogastric junction in a typical "bird's-beak" appearance (see Images 1 and 2 of some of the barium esophagogram x-rays with the typical bird's-beak appearance). The barium study was done with a lead apron covering the abdomen to minimize fetal radiographic exposure.

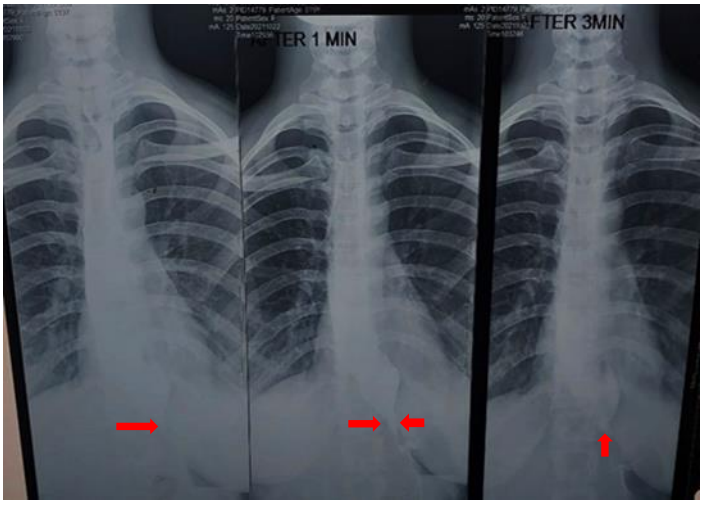

Image 1: Timed barium esophagogram at 0, 1 and 3 minutes. Note the classical bird's beak appearance at distal esophagus (red arrows) with proximal esophageal dilatation

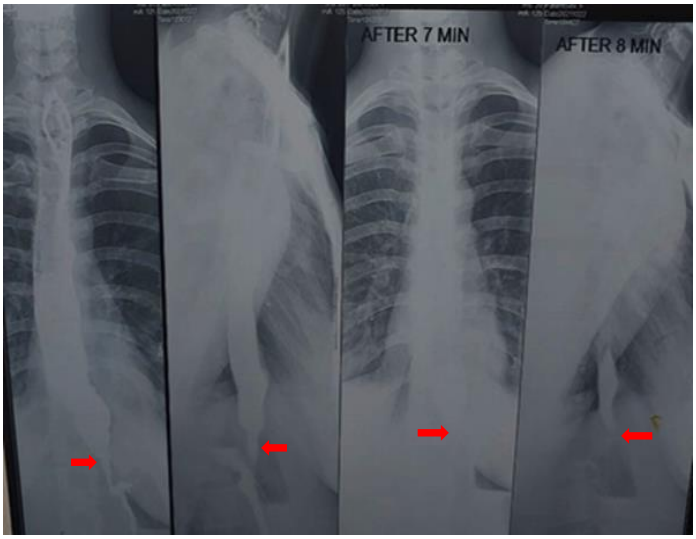

Image 2: Timed barium esophagogram at 5, 7 and 8 minutes showing persisting bird's beak appearance (red arrows) with slow trickling of small amount of barium distally.

Management and follow-up

The patient was diagnosed with achalasia cardia in pregnancy at 29 weeks gestation, complicated by fluid and electrolyte anomalies due to vomiting, maternal malnutrition, and moderate intrauterine growth restriction. Following a successful resuscitation, a multidisciplinary decision was made to offer conservative management and defer definitive surgical intervention until after delivery. A proposition for total parenteral nutrition was not feasible due to a multiplicity of factors. Therefore, she underwent serial endoscopic dilatation of the LES, which allowed for markedly improved enteral feeding (albeit with blended and semisolid foods), omeprazole and liquid antacids for acid suppression, and prokinesis with metoclopramide. In her subsequent antenatal clinic follow-ups, she progressively gained weight with improved nutritional status, and the baby's growth improved rapidly with normalization of the biophysical profile in follow-up ultrasound scans. She had an uneventful vaginal delivery at term of a female infant weighing 2.9kg. Two months after delivery, she underwent a successful, uncomplicated laparoscopic Heller myotomy and Dorr fundoplication (to prevent gastro-esophageal reflux after myotomy). She has remained stable since then and was discharged from active clinic follow-up. Two years later she's had no recurrent symptoms, and is a happily married mother of three.

DISCUSSION

The diagnosis of achalasia cardia in pregnancy is often difficult because the typical symptoms of heartburn, regurgitation, nausea, and vomiting are usually observed in normal early pregnancy and, when excessive, are accurately diagnosed as hyperemesis gravidarum (Austin et al., 2019). Consequently, achalasia cardia is diagnosed late in pregnancy, when patients may have significant fluid and electrolyte loss-related complications, malnutrition, and other life-threatening complications for both mother and fetus, including intrauterine fetal growth restriction and demise (Vosko et al., 2021). This was true for our patient, in whom the eventual diagnosis of achalasia cardia was made when maternal and fetal well-being was already jeopardized by severe maternal dehydration and malnutrition with resultant fetal intrauterine growth restriction. A clue for an alternate diagnosis of hyperemesis gravidarum in her case was the progressive worsening of the symptoms beyond the first trimester. The typical natural history of hyperemesis gravidarum is to progressively improve and resolve towards the end of the first trimester.

Confirmatory tests for achalasia cardia are available. High-resolution manometry (the gold standard) is safe in pregnancy but has no standardized values specific to pregnant women. It allows for therapeutic and prognostic classification of achalasia cardia (Patel et al., 2022; Vaezi et al., 2020). Barium swallow/esophagogram with serial timed x-ray images will reveal the pathognomonic "bird's beak appearance", in which proximal esophageal dilatation is accompanied by tapering at the esophagogastric junction (Vaezi et al., 2020). Plain chest x-rays and a CT scan of the chest may show a dilated tortuous esophagus with food residue (Ishii et al., 2019). According to the American College of Obstetricians and Gynecologists' Committee on Obstetric Practice, the

risk of radiation exposure to the fetus with these x-ray modalities is much lower than that associated with fetal harm. Therefore, they should not be withheld from a deserving pregnant woman ("Committee Opinion No. 656 Summary: Guidelines for Diagnostic Imaging During Pregnancy and Lactation," 2016). Endoscopy helps to exclude mechanical obstruction and may show a dilated, tortuous esophagus with food residue and secondary esophageal ulceration or infection. It is relatively safe for the fetus and allows for interval therapeutic LES dilatation (Friedel et al., 2014; Vosko et al., 2021). In our rural set-up here, we were able to safely make a definitive diagnosis by means of an initial endoscopy followed by a complementary barium swallow or esophagogram. Out of abundance of caution, a lead-shield apron was used to cover the abdomen of the patient and thus limit fetal radiation exposure during barium fluoroscopic studies.

Management of achalasia cardia in pregnancy includes adequate fluid and electrolyte resuscitation, nutritional support (using a nasogastric tube, parenteral nutrition, or gastrostomy feeding tubes), and medications to reduce the LES pressure, e.g., calcium channel blockers and long-acting nitrates (Vosko et al., 2021). Endoscopic treatments include injection of botulinum toxin (Botox) into the LES (Li & Tang, 2020), pneumatic dilatation, and peroral endoscopic myotomy [POEM] (Vosko et al., 2021; Youn et al., 2016). A proper risk-versus-benefit analysis must be individualized for each considered modality. Our patient successfully underwent serial endoscopic dilatation, which allowed for progressively normal alimentation, thus allowing for successful maternal and fetal nutritional recovery and normalization of growth. The definitive treatment is a surgical myotomy, either done laparoscopically or by the open method. The choice and timing of surgery are determined by fetal maturity and safety, maternal safety, and the availability of expertise and resources. Generally, surgery is safe in the second trimester due to very low risks (<5%) of spontaneous abortion or preterm labor (Vosko et al., 2021). There are several reported successful second trimester laparoscopic procedures, including one laparoscopic Heller myotomy (Palanivelu et al., 2007). Nonetheless, significant risks of perinatal complications, including fetal demise, remain, and as such, it is advisable to postpone surgery as much as possible until after delivery. This is what informed our decision to offer laparoscopic Heller myotomy after delivery. Due to the rarity of achalasia cardia in pregnancy, no consensus guidelines are available so far on its management, though there has been a recent proposal based on an existing literature review (Vosko et al., 2021). To the best of our knowledge, this is the first published case of achalasia cardia in pregnancy in Kenya.

CONCLUSION

Hyperemesis gravidarum is the usual diagnosis in pregnant women with significant vomiting, especially in the first trimester of pregnancy. Where vomiting is persistent and progressive into the second and third trimesters and associated with regurgitation of undigested food, dysphagia, severe acid reflux, and ensuing dehydration and malnutrition, surgical causes (e.g., achalasia cardia) must be ruled out.

RECOMMENDATIONS

Achalasia cardia in pregnancy can be diagnosed using a simple barium esophagogram and upper gastrointestinal endoscopy, with the latter also useful for interval dilatation of the LES to allow for adequate alimentation. These can be safely done without any significant risk to the fetus, as demonstrated in this report. Definitive management is surgical, i.e., Heller myotomy, which can be done laparoscopically or by the open method. Whereas case report data suggests that this can be done during the second trimester of gestation, there is no strong, robust scientific evidence so far that assures the safety of such a procedure in pregnancy. More collaborative data is needed. Therefore, surgery should be delayed until after delivery to assure maternal and fetal well-being.

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