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Huge Megacolon Complicating Chronic Colonic Volvulus in a Pregnant Lady with Partial Hirschsprung Disease: A Case Report and Review of the Literature

Sayed El-Akhras[1], Mohamed H Salama[1,2], Mohamed Abo-Elenen[3], Ali Elbadry[4], Diaa Abdelhalim[1,5], Panayiotis Tanos[6] and Vasilios Tanos[7,8]*

[1]Department of Gynecology and Obstetrics, Omam Hospital, Egypt

[2]Department of Obstetrics and Gynecology, Ain Shams University, Egypt

[3]Department of Internal Medicine, Omam Hospital, Egypt

[4]Department of Colorectal Surgery, Omam Hospital, Egypt

[5]Department of Obstetrics and Gynecology, Al-Galaa Teaching Hospital, Egypt

[6]Institute of Applied Health Sciences, University of Aberdeen and Aberdeen Royal Infirmary, UK

[7]University of Nicosia-Medical School, Aretaeio Hospital, Cyprus

[8]Department of Research and Development, Omam Hospital, Egypt

Abstract

Introduction: Megacolon is a rare, but potentially deadly complication of colonic inflammation. Colonic volvulus is the third most common cause of large bowel obstruction worldwide. Hirschsprung's Disease (HD), is a rare congenital colonic disorder. However, it can present in adulthood and can have particularly high risk of obstetric complications to both the mother and the fetus with main complication being refractory chronic constipation. Literature review detected 6 articles reporting the clinical manifestations and the management of patients with refractory chronic constipation in pregnancy of 9 cases.

Case: In this article, a complicated case of a 36 weeks parous lady, G3P2 with megacolon is presented. The consequences and delivery outcomes, of chronic constipation during pregnancy, due to chronic volvulus of an already dilated colon, on top of enteric atony due to partial HD, are discussed.

Conclusion: Preventive dietary measures, laxatives and frequent antenatal care may probably reduce the risk of complicated megacolon, caesarian section and premature delivery. Chronic constipation during reproductive age should merit further clinical attention during antenatal care, as well as multi-disciplinary approach in the management and treatment of these patients.

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Correspondence:
Panayiotis Tanos, Institute of Applied Health Sciences, University of Aberdeen and Aberdeen Royal Infirmary, NHS Grampian, UK, E-mail: p.tanos@outlook.com

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Introduction

Megacolon refers to huge colon dilatation due to fecal compaction as a result of bowel dysfunction without any mechanical obstruction [1]. Many congenital and acquired diseases characterized by innervation defect and/or transmural inflammation of the colon, lead to severe reduced peristalsis, chronic constipation and fecal impaction. Acquired conditions such as inflammatory disease, as well as neurological, systemic, metabolic diseases and medication may lead to megacolon [2]. Hirschsprung's Disease (HD) in pregnancy is frequently reported to also cause colon dilatation, negatively affecting maternal health and pregnancy outcomes.

HD is a rare disease with an estimated prevalence of 1:5000 live births and belongs in the family of intestinal innervation defects, also known as dysganglionosis [3,4]. Intestinal innervation defects can be divided into a) Genetic type (Hirschsprung's disease, Segmental aganglionosis of colon), b) Hypogenetic type (an isolated hypoganglionosis), c) Dysgenetic type (intestinal neuronal dysplasia causing a variable degree of intestinal dysfunction and colon fecal impaction) [5,6].

The diagnosis of HD is histological, whereas lack of ganglion cells in the area of the Auerbach plexus and Meissner plexus in the distal section of the large intestine is pathognomonic. The pathogenesis of HD is not fully understood. It is assumed that during embryonic development 4 to 12 week of gestation, due to migration failure of the primary neural cells results to aganglionic colon

[7]. Another hypothesis suggests that microenvironment changes cause abnormal differentiation, degeneration of ganglion cells and premature cell necrosis resulting to defective peristalsis partly or along the whole large intestine [8].

A very short segment has been reported isolated in the anal canal just above the pectinate line to the rarest and most severe clinical form of HD which is total colonic aganglionosis, observed in 5% of patients. The most common form occurs in the distal part of the sigmoid colon and rectum found in 75% to 80% of cases. In 10% of cases a more severe form of long aganglionic segment is observed extending from the rectum, sigmoid colon, and colon up to the splenic flexure [9,10]. Clinical symptoms appear in 70% to 90% during their first days of life and absence of meconium after birth raises the suspicion of HD [11]. Up to 80% of infants demonstrate defecation and dietary problems, delayed physical development, significant flatulence, and emesis. Other children are asymptomatic until late childhood. Chronic constipation, malnutrition, and physical development delay require colonoscopy and biopsy to rule out HD. Occasionally HD can cause acute enteritis and severe diarrheas with a 30% fatality rate [12,13].

During pregnancy, high and prolonged progesterone levels impair smooth muscle tone, causing in many women constipation, urine retention and varicose veins [14]. In addition, around 20 weeks of gestation, as uterus expands and compresses the bowel, constipation may become a significant antenatal treatment problem [15]. Additionally, laxative medications, and manual fecal disimpaction, are avoided in 2nd and 3rd trimester to avoid premature contractions being induced [16]. Consequently, the combinations of a pre-existing condition like HD, mechanical compression by the pregnant uterus as well as the hormonal effects on the intestinal tract, pose a high-risk to chronic constipation in pregnant women.

Case Presentation

A 25 years old lady, gravid 3, para 2, with 2 previous vaginal deliveries presented at our outpatient clinic at 34 weeks complaining of a progressive abdominal enlargement, discreet from the gravid uterus that has been growing since the third month of her pregnancy. Strangely, the patient reported, complete absence of bowel movements since early second trimester, with occasional flatus every few days and unintentional loss of 5 kg during the last 3 months of her pregnancy.

The patient reported similar progressive abdominal distention, along with constipation in her previous two pregnancies. However, in the current pregnancy the abdominal enlargement had been significantly more remarkable and with complete absence of bowel movements since, 18 weeks of gestation. Her bowel movements were compromised during both previous pregnancies, where she became constipated and had a single motion per week, despite using multiple laxatives. The patient's past medical history was insignificant except from a potential congenital bowel problem suspected in infancy for which investigations were refused. During adulthood the patient reports dependence on laxatives to maintain one or two bowel motions per week.

During her 3rd pregnancy, she rarely attended antenatal care where laxatives were recommended until fetal maturity. Finally, when her condition continued to deteriorate, she attended our hospital. Her vital signs were within normal limits, but she looked very weak and cachectic. Her weight was 47 Kg, and BMI was 17.6 kg/m^2. Abdominal examination revealed a large abdominal mass, measuring 45 cm craniocaudally and 18 cm across, rising on the right side of the abdomen from the symphysis pubis, up to the xiphisternum (Figure 1).

The gravid uterus was palpated at 36 weeks fundal level, displaced by the mass to the left abdominal side. The mass was rather mobile, mildly tender, of a semi-solid consistency, which was compressible on finger pressure. Abdominal examination was otherwise unremarkable. Vaginal examination revealed a hard, smooth, rounded mass filling the pelvis, but that was not the fetal head, which was high in the abdomen and pushed to the left side. The cervix was soft, closed, long, retracted behind the symphysis pubis and deviating to the left side. Rectal digital examination revealed an empty rectum, with no palpable masses or lesions. Pelvic-abdominal ultrasound scanning revealed a single viable fetus, with biometry corresponding to menstrual age. It also revealed marked distension of the bowel, with an oblong shaped hard lesion all over the abdominal and pelvic cavities, likely markedly augmented colonic distension, for further investigation. MRI of the abdomen and pelvis revealed marked dilatation of the large intestine till the sigmoid colon, where there is a large impacted fecal matter (Figure 2).

The patient was conservatively managed at our institution, hoping to push her closer to fetal maturity, as long as maternal and fetal wellbeing allowed. She was given steroids for fetal lung maturity at 36 weeks, and was scheduled for delivery at 37 weeks. Digital vaginal examination revealed she was not a candidate for vaginal delivery, as the fetal head was totally pushed aside out of the maternal pelvis by the distended colonic mass. The decision for caesarean delivery was made. During the c-section an extended abdominal midline incision revealed the huge colon was almost completely occupying the abdominal cavity and was remarkably bigger than the uterus, which was bent to the left side. Inevitably the gravid uterus was pulled extra-abdominally to gain good exposure of the lower segment and a healthy, 2,500 gm baby was delivered. Once the uterine wound was sutured and the uterus contracted, we could diagnose the hugely dilated and bizarrely elongated sigmoid colon, which was twisted ¾ turns around its mesosigmoid (Figure 3). The diagnosis of chronic sigmoid volvulus was then made.

Combined milking of the sigmoid, abdominally and per rectal digital fecal disimpaction, failed. Colon damage and loss of function was evident due to prolonged ischemia after atony and extreme distention, accompanied by a very thin wall and dark color of the viscera (Figure 3). Sigmoidectomy and temporary colostomy followed and 2 intra-peritoneal drains were left in the pelvis. The abdominal wound was closed in layers of anatomical order.

Figure 1: Abdominal examination with a characteristic huge solenoid semisolid mass, measuring 45 cm craniocaudally, and 18 cm across.

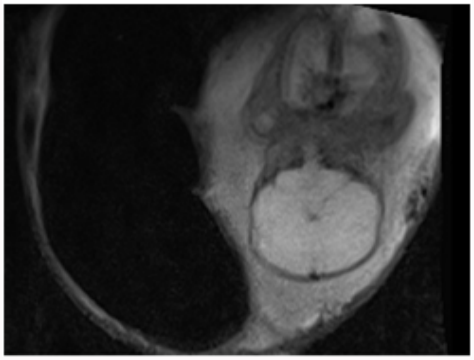

Figure 2: MRI of the abdomen and pelvis. Marked dilated colon with impacted fecal matter noted in the right abdominal side. Fetal cephalic presentation displaced in the left pelvic side.

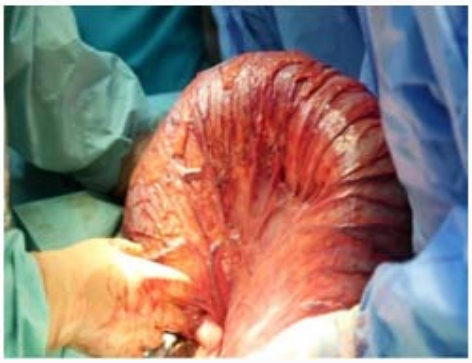

Figure 3: Hugely dilated and elongated sigmoid colon that was twisted ¾ turns around its mesosigmoid.

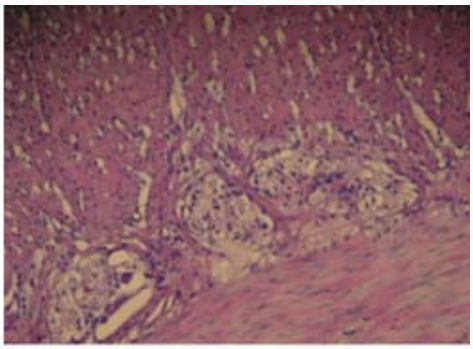

Figure 4: Excised huge sigmoid colon, measuring 107 cm in length and 27 cm in width.

The postoperative course of the patient was good, and she was kept Nil-Per-Os until the colostomy was functional within 2 days. Her vital signs were stable and the wound clean. The only remarkable finding was a moderate edema of the colostomy stoma, due to hypoalbuminemia (2.5 gm/dL), attributed to her malnourished preoperative state. This was corrected by human albumin infusion, with a resultant reabsorption of the edema. The patient and her newborn were discharged from the hospital 2 weeks later, in a good health. Reversal of the colostomy was performed 3 months post-operatively, without complications.

The removed huge sigmoid colon, measured 107 cm, in length, and 27 cm in width, and containing 6,800 gm of fecal matter (Figure 4). We are not aware of any such massive dimensions of a sigmoid colon being reported in literature.

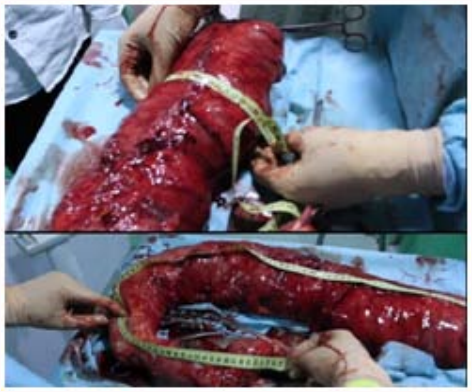

Figure 5: Histological biopsy demonstrating the lack of ganglion cells in the area of Auerbach plexus and Meissner plexus in the distal section of the large intestine, pathognomonic for Hirschsprung disease.

Histopathological examination of the resected colon revealed full thickness colonic wall tissue with few scattered lymphoid follicles in mucosa and submucosa along with prominent nerve trunks between circular and longitudinal layers of the muscularis propria. Ganglion cells could not be identified in submucosa or muscularis propria. Ganglion cells could not be identified in the resection margin of the narrow end. However, few ganglion cells were seen in the resection margin of the wide end. This was consistent with the diagnosis of hypoganglionosis with a-ganglionic distal end, picture of Hirschsprung's disease (Figure 5).

Discussion

In this article, a complicated case of pregnancy with megacolon is presented. The consequences of chronic constipation due to chronic volvulus of an already dilated colon, on top of enteric atony due to partial HD, during pregnancy and delivery outcomes are discussed.

The management options of the case were thoroughly discussed among the multidisciplinary team prior to surgery. Midline laparotomy was decided to expose both the uterine lower segment and the hugely dilated colon. Once the fetus was delivered by cesarean section and the uterine wound was closed, the sigmoid in torsion was untwisted. Trial of closed drainage of the sigmoid failed. Very hard, pasty fecal matter was delivered, at a slow rate because of its hard consistency. The evidence of prolonged severe ischemia was prominent. The decision of sigmoidectomy was based on the clinical diagnosis of the complete loss of function of the colon based on the following: a) the very thin wall and dark color of the colon b) the extreme compaction of the mass due to long-standing extreme dilatation and, c) doubt that the colonic tissue can withstand the trauma of squeezing. The distended colon was resected and exteriorized for colostomy which was previously reported in literature to be successful [17,18].

On reviewing the literature, most of the cases were pregnant women, in whom warning symptoms of severe and refractory constipation appeared between 16 and 28 weeks of gestation. Among the 9 pregnant women who had megacolon after chronic constipation, 2 had a history of ulcerative colitis and the other 7 had history of colonic innervation defect. Most of the patients (6/9) had vaginal delivery by forceps or vacuum mainly due to dystocia, and the

rest had a cesarean section. Colectomy and temporal ileostomy were performed in 3 out of the 9 women. Most of the patients had been using unsuccessfully, oral volume expanders and laxatives to facilitate defecation [14-16,19-21].

Chronic constipation resulting in megacolon has been described in the literature in patients with inflammatory bowel disease, and Hirschsprung's disease. The peak incidence of most of these diseases coincides with childbearing age. When patients have been managed by colectomy, then infertility can be a problem mainly due to abdominal adhesions [22,23].

In case of Hirschsprung's Disease (HD), the pathological condition leading to megacolon is different. The lack of parasympathetic ganglion cells in submucosal and mesenteric plexus of part of colon causes functional obstruction and colonic dilation proximal to affected segment. The colon innervations defect and decreased peristalsis deteriorate by the pregnancy high progesterone levels and facilitates fecal compaction. Chronic and refractory to medication constipation seems to lead to chronic colon inflammation and extreme distention, increasing the risk of toxic megacolon. The obstetrical complications such as intra uterine growth retardation and preterm labor are anticipated once the colon is over distended and nutrients absorption is compromised.

The timing and type of delivery depends upon the gestational age, fetus weight, maternal health condition, degree of feces impaction and birth canal obstruction. Once gestation is close to term earlier delivery due to maternal discomfort and malnutrition, inducing vaginal delivery due to imminent complete fecal compaction might be indicated. Vaginal delivery should be the preferential delivery option for maternal and neonatal advantages. Elective cesarean section due to severe or complete colon impaction is obligatory once obstructed labor or malnourished parturient are diagnosed. Manual disimpaction is an alternative, however with a risk of failure and fetal bradycardia during the process due to induced persistent uterine contraction [16,24].

Conclusion

To conclude, unavoidably, clinicians as well as specialist gynecologists and general surgeons will at some point have to deal with one of these conditions and not unlikely a combination of them simultaneously. Due to the anatomical locations involved, specialist knowledge and cross discipline collaboration is vital. The management of HD during pregnancy should include more frequent multi-disciplinary prenatal care with special attention to bowel motions and development of complications. Early laxative treatment in combination with special nutrition might prevent impaction of feces and prevent megacolon, prolonging pregnancy to term, and avoiding and dystocia. Frequent ultrasound and estimated fetal weight can reassure fetal condition and pregnancy outcome. High-water intake, enemas and laxatives are the major conservative preventive measures. Delivery planning should be based on inputs by obstetricians, colorectal surgeons, anesthesiologists and neonatologists to ensure the best outcomes for both the mother and baby.

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