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A DIAGNOSTIC CHALLENGE: NEWBORN WITH HYPOPLASTIC LEFT COLON SYNDROME AND HISTORY OF GESTATIONAL DIABETES CASE REPORT

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Abstract: Hypoplastic left colon syndrome is a very uncommon malformation and manifests itself with abdominal distension and vomiting in the first hours of life. It is associated with gestational diabetes and is diagnosed clinically and by imaging. OBarium enema, considered the standard for diagnosis, reveals an abrupt narrowing of the left colon. Its pathophysiology is not yet fully elucidated. Treatment is generally conservative in mild cases, and pediatric supervision is essential to avoid complications. The objective of the case presented is to highlight the importance of considering hypoplastic left colon syndrome in newborns with the symptoms mentioned in children of gestational diabetic mothers.
Keywords: Descending colon, gestational diabetes, neonate

INTRODUCTION

Hypoplastic left colon syndrome is one of the abdominal anomalies that affect newborns. Abdominal manifestations begin to occur in the first hours of life, such as abdominal distension and vomiting. Upon barium enema, which is the gold standard for diagnosing it, a sudden narrowing of the caliber of the left colon is noticed, compromising normal intestinal transit. (DAVIS et al, 1974) (STEWART et al, 1977) (VALERO et al, 2011).

This pathology has a direct relationship with maternal diabetes or gestational diabetes, with some studies stating that half, others highlight that more than half of hypoplastic left colon syndrome are caused in newborns with diabetic mothers, this very evident link between these two diseases has not yet been explained, the most accepted theories relate the influence of diabetes on the secretion of glucagon, which will influence sympathetic and parasympathetic fibers, which will interfere with intestinal motility, which will change the caliber of the colon. (DAVIS et al, 1974) (STEWART et al, 1977) (VALERO et al, 2011)

Most of the time, treatment is conservative, with the child's own growth and development, intestinal transit regains its normal function and rarely requires surgical intervention. (DAVIS et al, 1974) (STEWART et al, 1977), (VALERO et al, 2011).

In the following report we will present the clinical history, diagnostic examination and treatment of a patient diagnosed with hypoplastic left colon syndrome, in order to exemplify this pathology that presents little data in the literature.

CASE REPORT

Patient, female, daughter of a 43-year-old mother (G4P3) is a cocaine user (once/week) and a smoker (1 pack/day), who stopped when she discovered she was pregnant, also has asthma, congenital adrenal hyperplasia, depression, in addition to a suicide attempt in 2022 and gestational diabetes mellitus (GDM), which did not occur in previous pregnancies. She underwent prenatal care, totaling 27 consultations (11 in low-risk prenatal care and 16 in high-risk), carried out from the first month of pregnancy. Maternal serology and rapid tests before birth were negative. Caesarean section birth, due to broken water, amniotic fluid was clear and without lumps. There were no complications during the procedure, he was born on 11/06/2022 at 38 weeks of gestation, in good general condition, normal tone, spontaneous breathing, timely cord clamping, heart rate above 100 bpm, APGAR 8/9, weight of 3010g, 48cm in height, 35cm in head circumference and the newborn's temperature in the first five minutes was 36.2°C. After birth, the patient was transferred to the neonatal ICU for social hospitalization.

In the neonatal ICU, arterial blood gas analysis, infection control, coagulogram, blood culture, echocardiogram and brain

ultrasound were performed, showing results within normal parameters. During the hospitalization period: on 06/11 he received pasteurized human milk (LPH), evolving with abdominal distension, associated with vomiting, gastric stasis and meconium evacuation. On 11/07 and 11/08 he remained fasting, but still had abdominal distension. On 08/11, an abdominal X-ray was requested with significant distension of the loops of non-specific appearance. On 11/09, an abdominal US was performed which showed marked gaseous distension of the intestinal loops. Based on the clinic, the history of maternal gestational diabetes and the images previously reported, a barium enema was requested, due to suspicion of hypoplastic colon syndrome. The barium enema report found an apparent reduction in the caliber of the sigmoid, compared to the other segments. Thus, confirming the presence of Hypoplastic Left Colon Syndrome. (Figure 1)

After diagnosis, the patient was already tolerating formula in match adequately and without new episodes of vomiting or abdominal distension, which is why there was no intervention, as the patient was clinically stable. On 11/16, after discussion with social services, the patient was authorized to be discharged with her mother, following the guidance of the guardianship council. Follow-up after discharge was outpatient, along with high-risk childcare, pediatric surgery and pediatric gastronomy.

During outpatient follow-up, when the patient turned 6 months old, in a consultation the parents reported that they had started introducing food, but the child had constipation that improved only after guidance on nutrition and return for continued follow-up.

Figure 1. Bare Enema which showed a reduction in the caliber of the sigmoid in comparison to the other segments. Possibility of "Hypoplastic Left Colon Syndrome"

DISCUSSION

The incidence of the syndrome is still difficult to measure, as many cases are asymptomatic. Furthermore, the information in the literature about the disease comes from case reports or series of isolated cases, compromising statistics. A rough estimate made in the study by Valero and collaborators indicates that the incidence of symptomatic disease is 1/22,500 live newborns. (VALERO et al, 2011)

The relationship between hypoplastic left colon syndrome and maternal diabetes was first established by Davis and colleagues in 1974, when the disease was first described, although this relationship is well established, as approximately 40 to 50% of this pathology is related to maternal or gestational diabetes, the pathophysiology is not yet known, in addition, other factors may also influence the incidence of hypoplastic left colon syndrome, such as: metabolic disorders (hypothyroidism or hypermagnesemia), prematurity, maternal intake of psychotropic drugs and stress neonatal. (DAVIS et al, 1974) (DE OLIVEIRA et al, 2015) (KANG et al, 2015) (MIRABELLI et al, 2021) (STEWART et al, 1977) (VALERO et al, 2011)

GDM has a prevalence of approximately 14.3 million in Brazil, according to the Unified Health System (SUS). The risk factors for developing this pathology are:

age over 25 years, obesity or excessive weight gain during pregnancy, family history of diabetes, short stature.(DE OLIVEIRA et al, 2015) (MIRABELLI et al, 2021)This maternal disease can have consequences for pregnancy, as high hyperglycemia increases the availability of glucose to the fetus, altering metabolism, which can generate macrosomia, hypoglycemia, prematurity, congenital malformations, in addition to hypoplastic left colon syndrome, as reported here. study. (DE OLIVEIRA et al, 2015) (ELLIS et al, 2009) (MIRABELLI et al, 2021)

The onset of the disease tends to appear within the first 24 to 48 hours. The most common signs are mild to moderate abdominal distension and vomiting, which may contain bile contents. In the present report, it was noted that the patient presented with abdominal distension, vomiting and gastric stasis, similar to reports in the literature. (STEWART et al, 1977)

The exam of choice for diagnosing the pathology is the barium enema, from this exam it is possible to notice the reduction in the caliber of the affected colic element in comparison with the other preserved ones, in addition, according to Davis and collaborators, in their first report on the disease, highlighted that the loops at the level of the splenic flexure are always dilated, with the presence of meconium, despite this, in the minority of cases there is obstruction in the region, that is, the meconium is evacuated. Although, in some cases the evacuation of meconium may be delayed, lasting from the usual 24h to 72h. In the case described, the patient had a normal meconium evacuation. The barium enema, in addition to being diagnostic, is therapeutic, as it promotes the evacuation of contrast and meconium, facilitating intestinal clearance. (DAVIS et al, 1974) (ELLIS et al, 2009) (STEWART et al, 1977) (VALERO et al, 2011)

Many authors in the literature recommend, in addition to this exam, a rectal biopsy to rule out a ganglionosis. Despite this, Valero and his collaborators state that this practice is not necessary, since if there is an anal inhibitory reflex present and a barium enema suggestive of hypoplastic left colon syndrome, there is no appropriate indication for a biopsy. (DAVIS et al, 1974) (VALERO et al, 2011)

In another study, Ellis and collaborators analyzed the clinical evolution of 105 newborns, children of diabetic mothers, in an Australian hospital. Of the total, 5 children started with a condition similar to the patient reported in the first three days of life. In newborns, enemas for cleaning, helping to unblock intestinal transit, none required surgical treatment and all children completed the treatment without any difficulty in evacuating, the average follow-up was 2 years. (ELLIS et al, 2009)

Ellis et al. highlight that they even looked for cases of hypoplastic left colon syndrome in non-diabetic mothers, but did not find any cases. Furthermore, one of these cases analyzed in the study was twins, reaffirming the influence of maternal diabetes, as they both presented the same pathology. (ELLIS et al, 2009)

Although the disease does not have a very clear pathophysiology and etiology, some hypotheses relate neurohumoral instability between the autonomic nervous system and glucagon, in which gestational diabetes or maternal diabetes influences the hormone glucagon, which interferes with the sympathetic and parasympathetic autonomic system, generating interference in the motility and development of the left colon, this would partly explain the direct relationship between hypoplastic left colon syndrome and maternal diabetes or gestational diabetes. The differential diagnosis of this pathology in newborns is meconium plug syndrome,

low colon stenosis or atresia, Hirschsprung (which is rarer in diabetic mothers) and other dysganglinoses. Diagnosing hypoplastic left colon syndrome is often difficult because these other diseases are more prevalent, reducing the chance that doctors will think about hypoplastic left colon. (DAVIS et al, 1974) (ELLIS et al, 2009) (STEWART et al, 1977) (VALERO et al, 2011).

The evolution of the disease is benign in the vast majority of cases, resolving spontaneously, there are rare complications such as intestinal perforation mainly in the cecum and peritonitis. (KANG et al, 2015) (VALERO et al, 2011) The treatment of hypoplastic left colon syndrome is conservative, but cleansing enemas may be prescribed to facilitate intestinal transit. (STEWART et al, 1977) (VALERO et al, 2011).

CONCLUSION

Hypoplastic left colon syndrome, an uncommon pathology. Newborns with constipation, abdominal distension and vomiting, whose mothers are diabetic, must be considered as possible cases and abarium enema to confirm the diagnosis and, in some cases, as treatment. In most cases, intestinal transit recovers over time, but pediatric supervision is necessary to avoid complications.

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