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Case Report

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ANTENATAL DIAGNOSIS OF OMPHALOCELE: CASE REPORT

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ABSTRACT

An omphalocele is a congenital malformation due to a defect in the closure of the umbilical ring with externalization of the abdominal viscera covered by the translucent and avascular amniotic membrane. The diagnosis is feasible in 70% of cases between 11 and 13 SA. The herniated viscera are contained in a sac of amniotic origin on which the cord is inserted, the vascular elements of which can be clearly identified on ultrasound and possibly on Doppler. The prognosis of an isolated omphalocele, except for large forms, is generally good. It depends on the associated malformations, the earliness of treatment, the size of the neck and the Contents. The management of an omphalocele is urgent medico-surgical, it is multidisciplinary, is done by multiple and diverse therapeutic processes. Currently, the indication for surgery is systematic in the treatment of omphaloceles. We report a case of an omphalocele diagnosed later at 20 SA.

INTRODUCTION

The omphalocele is a congenital malformation of the abdominal wall. It is an embryopathy occurring during the first ten weeks of development and which would be linked to a defect in the closure of the umbilical ring leading to non-reintegration of the viscera covered with the amniotic membrane. The size of the parietal opening is variable (2cm to more than 10cm): from a simple hernia in the cord containing a few intestinal loops, to a giant omphalocele with a large part of the liver exteriorized. These differences in content make possible, on the one hand, to classify omphaloceles and, on the other hand, to try to better understand the chronology and pathogenesis of this malformation.

OBSERVATION

This is a 30-year-old patient, with no history, no notion of consanguinity, G2P1: the first is an uncuretted spontaneous abortion at 08 weeks, the second is a pregnancy estimated at 26 weeks, referred by a general practitioner for a morphological ultrasound; the patient did not benefit from a follow-up during the 1 first trimester.

The examination finds a patient out of work with TA at 12/06 afebrile, labstix negative, HU corresponds to the term.

The morphological ultrasound objectified an evolving single-fetal pregnancy whose biometry corresponds to the term, with a continuous addition image with the abdomen, characterized by regular contours and digestive contents(intestine and liver) with a Doppler vascularization at the center of the image making evoked an omphalocele with no other malformations detected (fig1, fig2).

The patient benefited from a follow-up in our department .the delivery was by cesarean, the newborn was female with apgar 10/10 and birth weight was 3200g (fig 3).

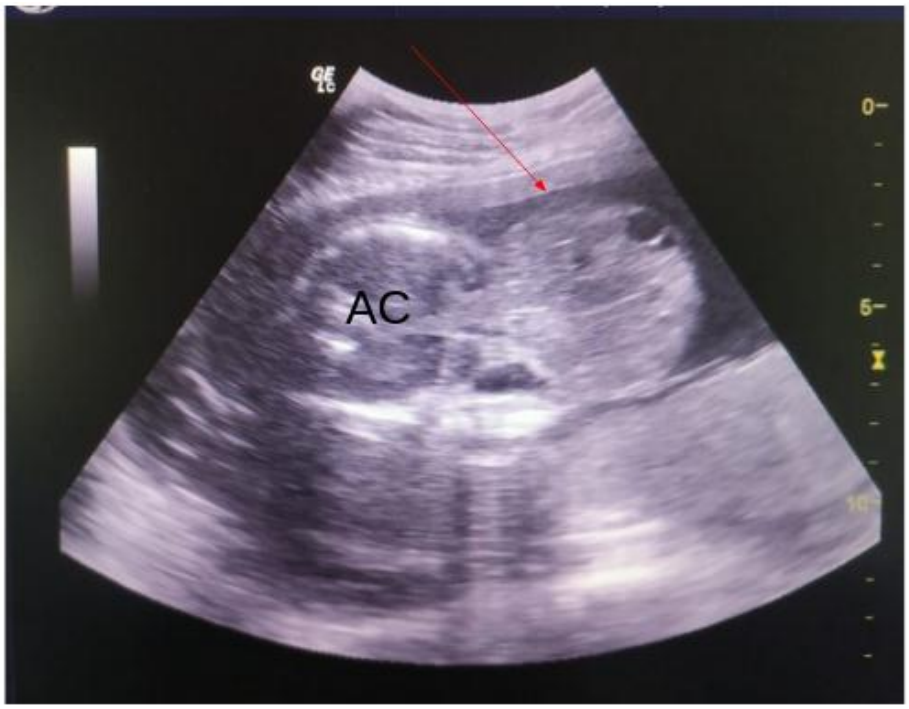

Figure 1: section of the abdominal perimeter showing an omphalocele.
AC: abdominal circumference ↘omphlocele

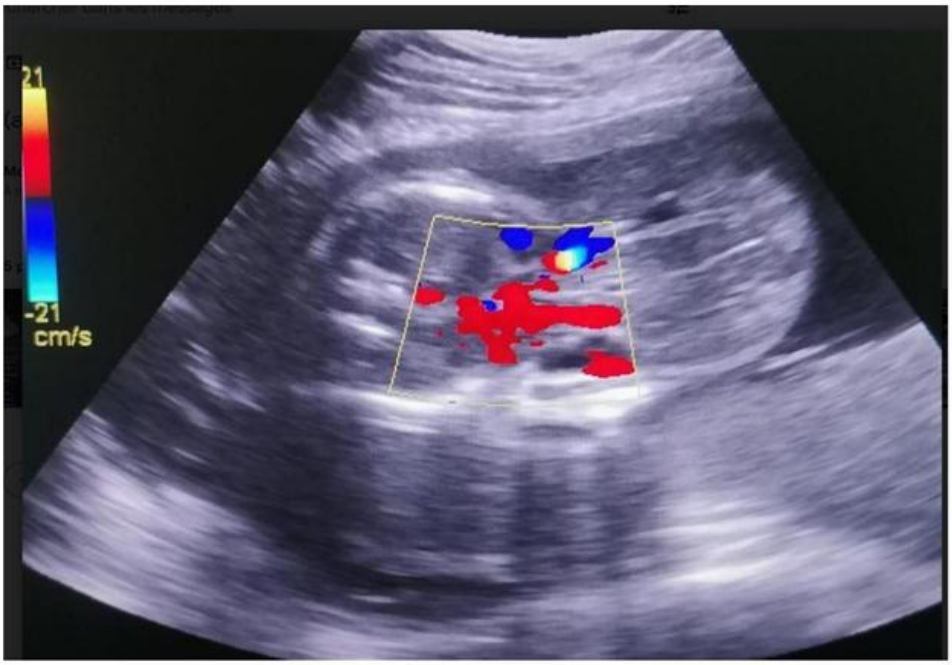

Figure 2: demonstration of a Doppler vascularization confirming the position of the cord.

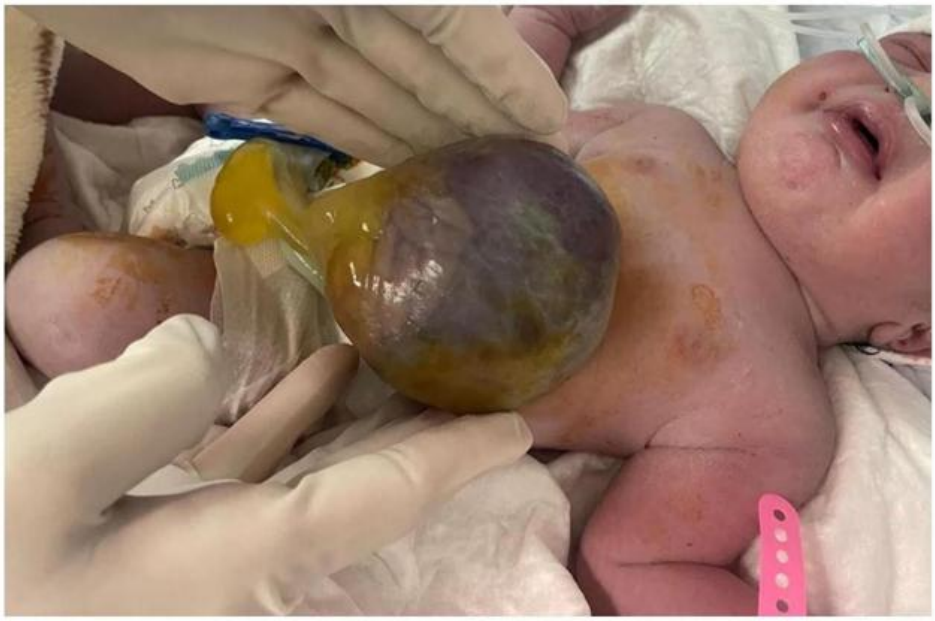

Figure 3: image of the newborn after birth confirming the diagnosis of omphalocele.
Omphalocèle content: intestine and liver

DISCUSSION

The omphalocele is a congenital malformation of the abdominal wall corresponding to a defect of closure of the umbilical ring with externalization of the abdominal viscera covered by the amniotic membrane occurring during the first ten weeks of development and which would be linked to a defect of closure of the umbilical ring leading to non-reintegration of the viscera covered with the amniotic membrane.[1]

The omphalocele or coelosomy media is the most frequent of these abnormalities of the development of the abdominal wall.[2] It is an anomaly of the morphogenesis which occurs during the embryonic delimitation in its lateral portions between the 3rd and the 4th week. For some authors, the protrusion observed is secondary to the failure of the closure of the anterior abdominal wall from the 4th week. For others, it is due to a lack of migration of the mesodermal somites.[3] Omphalocele is therefore a true embryopathy. The absence of muscular delimitation a explains the existence of a sac on which the umbilicus is inserted.

The omphalocele realizes a hernia of the viscera at the level of the umbilical region. The peculiarity of this hernia is that the viscera are covered not with skin, but with an amniotic membrane.

Usually the bag is formed by: the peritoneum on the inside, Wharton's jelly on the outside; It is transparent at the beginning allowing its contents to be seen. This bag can hold: the intestine, the liver, the omentum, the spleen, the ovaries or the associationof organs.

The development of digestive viscera outside the abdominal cavity has many consequences: a. abdominal wall hypoplasia. b. Intestinal malrotation and attachment defects the primitive loop intestine cannot its physiological rotation because it does not reintegrate the abdominal cavity, c. Hepatic adhesions, d. Adhesions with the intestine, e. Intestinal atresia. the malformations associated with omphalocele[5,6,7] are:a: chromosomal abnormalities: (40 to 60%) these include trisomies 13, 18, 21 and also Turner and Klinefelter syndromes and triploidy; b. cardiac abnormalities: (20-50%) include atrial and ventricular septal defects, tetralogy of Fallot, CIA, CIV, CAV. vs. genitourinary abnormalities: (40%), e: gastrointestinal anomalies: (40%) these are diaphragmatic hernias, malrotations, intestinal duplications, atresia and ascites.

Prenatal diagnosis

The use of endovaginal ultrasound allows the realization of an authentic fetal morphological examination at the end of the first trimester of pregnancy. Examination of the abdominal wall is possible from the 12th completed week of intrauterine life by ultrasound.[8] The presence of an umbilical hernia before the complete reintegration of the primitive intestinal loop is physiological before 12 weeks of amenorrhea[9], but its diameter is always less than 10 millimeters.[9] The diagnosis is easy by supra pelvic ultrasound. the endovaginal US can often provides more precision.

Diagnosis criteria for an omphalocele combines by ultrasound: it's hyperechoic mass, at the level of the base median umbilical cord, anterior, rounded with extra-abdominal development, connected to the abdominal wall by a collar. This mass is limited by a membrane wich the contours are clear. Its size and content are variable (mixture of liquid images and echogenic images depending on the organs present in the sac).[10] The viscera contained in the omphalocele

can be intestinal loops, the liver and the stomach. Identification of vascular elements can be aided by examination in color doppler mode: the umbilical vessels are located inside the herniated sac; in the middle, at the upper or the lower part of contents.

Ultrasound helps to determinate the antenatal prognosis of this malformation.[8] When the ultrasound diagnosis of omphalocele is made, it is necessary to look for the associated malformations and chromosomal abnormalities which are frequent and will intervene in the fetal prognosis.

The chromosomal abnormalities can be associated with the omphalocele: trisomies 13- 18-21.[13]

Delivery methods

The delivery must be in a structure where they are the neonatal resuscitation and pediatric surgery department for immediate care of newborn at birth.

The obstetric management of an isolated omphalocele is usually the vaginal delivery. The omphalocele is a soft swelling and does not present an obstacle to the progression of the fetus in the genital canal, a part from very large omphaloceles which sometimes require a cesarean section.[14]

PREOPERATIVE NEONATAL MANAGEMENT: From the birth of the child, the omphalocele must be covered by a sterile hail bag or by a sterile bandage ensuring protection against trauma, infection, necrosis and desiccation. Immediate management aims to prevent certain complications related to omphalocele such as: hypothermia, digestive distension, hydro-electrolyte imbalance and infection.

CONCLUSION

Omphalocoele is one of the most frequent malformations of the anterior abdominal wall where the associated malformations are frequent, in particular digestive, cardiac and urinary, which determines the prognosis.

Its expensive management requires a multidisciplinary technical platform: obstetricians pediatric surgeons, intensive care anesthetists, pediatricians, with the help of geneticists.

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