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RESEARCH ARTICLE

GIANT OMPHALOCELE ABOUT A CASE

Khalid Lghamour, Amina Lakhdar, Najia Zraidi and Aziz Baidada
Gynaecology-Obstetrics and Endoscopy Department, Souissi Maternity Hospital, IBN SINA University Hospital, Mohamed V University, Rabat, Morocco.

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Abstract

We report the case of a giant omphalocele 10 cm in diameter diagnosed by ultrasound in the obstetric emergency department in a patient with a full-term pregnancy in labor, in whom an emergency cesarean section was indicated with fetal extraction, performance of emergency neonatal care, coverage of the omphalocele with non-adherent wet compresses and management by the pediatric surgeon with the result of uncomplicated operative sequelae without complications.

Introduction:

Omphalocele is a defect in the closure of the anterior abdominal wall, resulting in the externalization of part of the abdominal contents, covered by a membrane made up of peritoneum deep down and amnion superficially. It corresponds to a herniation of abdominal viscera from the abdominal cavity through a median orifice located at the base of the umbilicus.

Case Report:

30-year-old patient, no previous history, gravida 1, para 1, admitted to obstetric emergency with full-term pregnancy in labor. vaginal touch revealed a two-finger dilated cervix, 80% effaced. obstetric ultrasound revealed an evolving monofetal pregnancy, with cephalic presentation, full-term fetal biometry, sufficient amniotic fluid, estimated fetal weight of 3300 grams, giant omphalocele with stomach and liver present in the sac. a caesarean section was indicated with wide pfannential skin incision and hysterotomy, in the presence of the neonatologist and paediatric surgeon, allowing extraction of a live female neonate, apgar 6/8/10, birth weight 3260g. Examination of the newborn found a giant omphalocele 10 cm in diameter with intact sac. Asepsis of the sac and coverage with a dressing of non-adherent moist compresses, parenteral broad-spectrum antibiotic therapy and surgical management by the paediatric surgeon with intra-abdominal reintegration of the herniated viscera and parietal closure. The post-operative course was straightforward, with no complications.

Corresponding Author:Khalid Lghamour
Address**:**Gynaecology-Obstetrics and Endoscopy Department, Souissi Maternity Hospital, IBN SINA University Hospital, Mohamed V University, Rabat, Morocco.

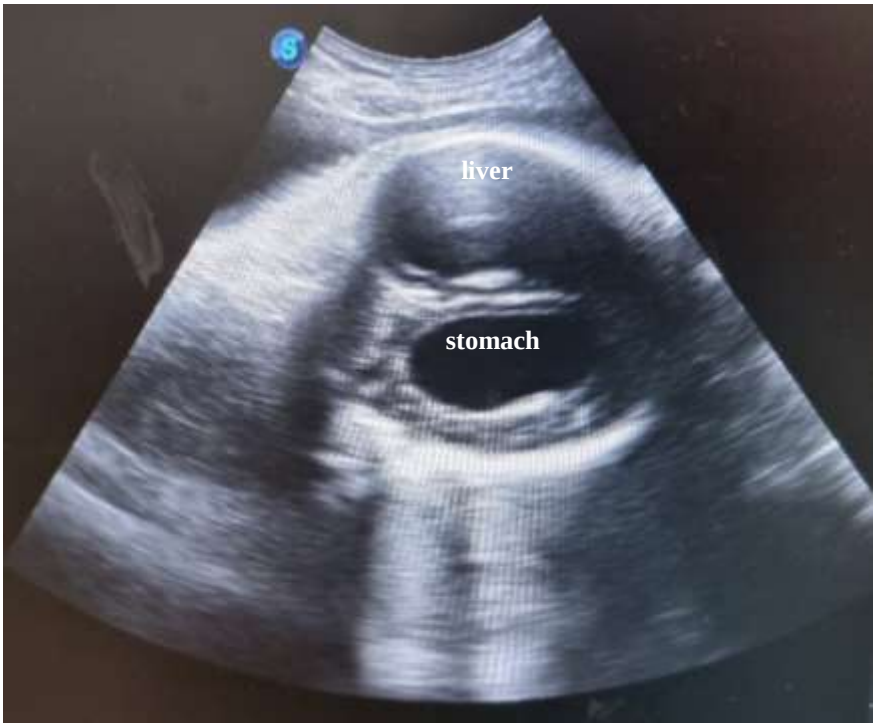

Figure 1: Obstetrical ultrasound showing giant omphalocele with herniation of stomach and liver.

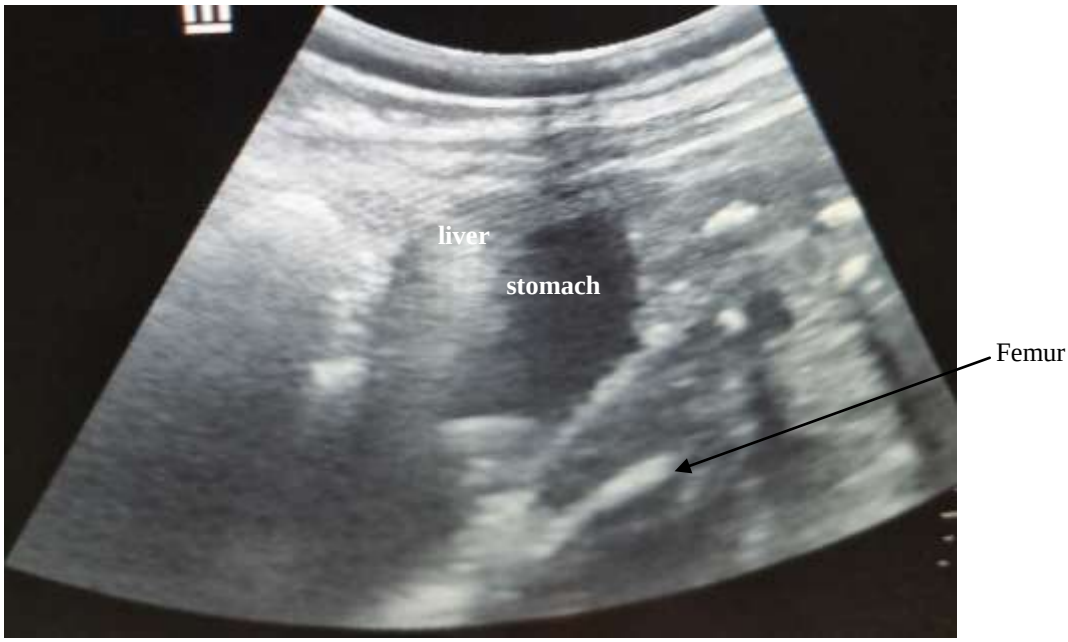

Figure2: Omphalocele on obstetrical ultrasound.

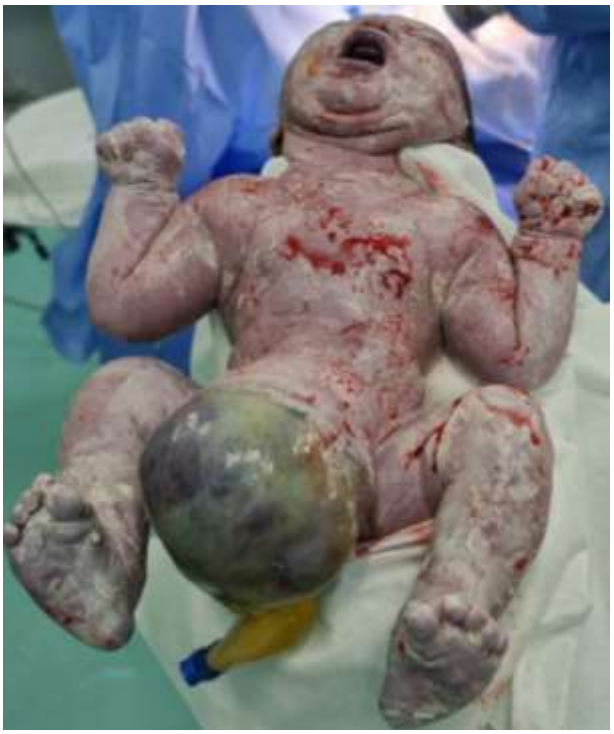

Figure 3: The newborn after Caesarean section extraction, giant omphalocele 10 cm in diameter.

Figure 4: Emergency neonatal care after birth, with non-adherent wet compresses.

Discussion:

Omphalocele is an embryopathy corresponding to an upper coelosomy contained in the cord, the parietal defect may be more or less extensive, involving all planes of the abdominal wall at the level of the umbilicus, the contents may be the small intestine, liver, gallbladder or stomach. The externalized viscera are protected by a sac composed of three layers: the outer amniotic layer, the intermediate Wharton's jelly layer and the inner peritoneal layer. Rupture of the membrane, leading to evisceration, can occur in utero, during delivery or after birth. The prevalence of omphalocele is estimated at 1/5000 births [1].

In 50% of cases, omphalocele may be associated with other anomalies: trisomy 15, 18, 21, heart disease, weidemannbeckwith syndrome or genitourinary malformations [2,3,4,5,6,7]. There are few or no associated digestive malformations.

Weidemann Beckwith syndrome occurs in 1/10000 births. In 50% of cases, it involves a methylation anomaly on chromosome 11, and in 20% of cases, paternal uniparental disomy. It includes omphalocele media, macroglossia, macrosomia and visceromegaly.

Pentalogy of Cantrell includes omphalocele, ectopia cordis, sternal cleft, anterior diaphragmatic hernia, and cardiac malformations such as tetralogy of fallot, ventricular septal defect and atrial septal defect.

OEIS syndrome accounts for some 1/400000 births and corresponds to an inferior coelosomy, omphalocele, cloacal exstrophy, anal imperforation and spinal anomaly.

Depending on the content of the herniated abdominal organs, the omphalocele is classified as containing or not containing liver. Small defects can usually be closed within the first 24 to 72 hours of life. Large defects usually require some type of silo within the first 24 hours and delayed closure.

This wall anomaly occurs early in fetal life, between 6 and 7 weeks' gestational age, and may show up on ultrasound between 9 and 10 weeks' gestational age [8]. An omphalocele that does not contain the liver can be reliably diagnosed antenatally after 12 weeks' gestation.

Before 12 weeks' gestational age, a small omphalocele can be difficult to differentiate from a physiological midgut hernia. Omphalocele is classified as [9]:
- “small”: <5 cm and containing a few bowel loops.
- “giant”: ≥5 cm or size relative to the fetal abdomen (cross-sectional area of the omphalocele >50% that of the fetal torso) and containing most of the liver (>50-75%).

Although there is no consensus on the definition of giant omphalocele (G.O.), the various criteria, according to the different definitions, often include: an omphalocele too large to be treated by primary closure, one whose neck is > 5 cm or one whose sac contains the liver [10, 11].

Aitken J. classifies omphalocele into types 1 and 2, type 2 being considered the definition of giant omphalocele [18]. Today, Aitken's classification remains the most widely used [13].

Type	Criteria
1 (all criteria must be met)	Collar diameter (fascia defect) < 4 cm
	Bag diameter< 8 cm
	Absence of liver in the bag
2 (only one criterion is required)	Collar diameter (fascia defect) > 4 cm
	Bag diameter> 8 cm
	Presence of liver in the bag

Table 1: Aitken's classification of omphaloceles.

Prenatal evaluation of omphalocele is based on measurement of volume, search for associated malformations, study of the thorax and assessment of progress in the third trimester.

Giant omphalocele can usually be diagnosed in the first trimester of pregnancy due to the high incidence of associated malformations. certain investigations such as genetic testing of amniotic fluid and additional imaging studies, including fetal echocardiography, are recommended [4,14]. These assessments can help identify associated anomalies, such as cardiac and pulmonary abnormalities, which have a significant impact on neonatal outcome and mortality [1,15]. Recent studies suggest magnetic resonance imaging to detect pulmonary assessment markers, as neonates with omphalocele and pulmonary malformations have a high mortality rate [16,17].the mode of delivery is planned according to clinical conditions, vaginal delivery is possible in the case of a small omphalocele, but in certain obstetrical conditions, a Caesarean section is recommended, as in our case of OG. Delivery is usually scheduled for around the 37th week of pregnancy. When the baby is born, immediate care is provided by the neonatology and pediatric surgery team. Pregnancy with an OG in the fetus requires a programmed caesarean section, with a large skin incision and hysterotomy to avoid rupture of the sac.

While the treatment of minor omphalocele can be achieved by primary closure, this is generally not feasible for the OG, at the risk of inducing intra-abdominal compartment syndrome, which would be life-threatening [18]. Several methods of treatment have been proposed, including surgical and conservative ones. Surgical means require either postoperative respiratory monitoring (Gross technique), or a silo and parenteral nutrition (Schuster technique), which are rare and costly in low-income countries [18,19]. Conservative treatment aims at the formation of an eschar, followed by its epithelialization, thus transforming the OG into an eventration. This treatment is associated with good results: better survival, shorter hospital stay, shorter time to full enteral feeding and elimination of complications associated with early closure [20,21]. For this reason, several substances have been applied to the OG sac, each with advantages and disadvantages, the most to be feared being intoxication with the tanning substance. The substance must have antimicrobial properties, to combat infection of the sac [20]. The use of aqueous eosin as an escarrificant was reported by Kouame BD et al. in a large series of 175 patients over 15 years. Results, in terms of duration of epithelialization, morbidity and mortality, were similar to those of studies using other topicals [21, 22]. Facial closure with repair of the ventricle is generally scheduled at 6 to 12 months of age, according to many authors [23].

Management of omphalocele in the delivery room is based on protection of the sac with sterile, non-adherent moist compresses of 2% aqueous eosin, 2 ml diluted in 500 ml isotonic saline, at a rate of 100 ml of the mixture per dressing, three times a day to maintain asepsis and avoid evaporation. The newborn should then receive IV fluids and broad-spectrum antibiotics while awaiting surgical intervention, which consists of reintegration of the herniated viscera into the abdomen and direct parietal closure or placement of a Silo (Shuster) prosthetic plate. When primary closure is not possible, various techniques can be used.

Post-operative management of omphalocele consists of directed wall healing and plaque care to avoid the risk of sepsis, prevention of gastro-oesophageal reflux by careful posture, milk thickening and anti-reflux treatment, and prevention of respiratory failure.

Despite advances in neonatal resuscitation, surgery, anesthesia and nutrition, giant omphalocele remains a challenge for the surgeon. In the absence of chromosomal abnormalities and severe associated malformations, the mortality of giant omphalocele is reduced [24].

Conclusion:

A rare, non-syndromic congenital malformation characterized by the absence of abdominal wall closure centered on the umbilical cord and extra-abdominal externalization of viscera protected by a sac. It may be associated with other anomalies, Fetal extraction is most often performed by Caesarean section, and protection of the hernia sac by moist dressings with 2% aqueous eosin is essential pending treatment, which may be either a consultative approach or, as in our case of giant omphalocele, a surgical procedure involving intra-abdominal reintegration of the viscera and parietal closure.

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