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Case Report

Total Hepatic Herniation Associated with Giant Omphalocele: Case Report

Daniel Manzano Moscoso[1]*, Laura González Gordón MD[2] and Ana Lucía Cevallos Rodríguez[3]

[1] *Pediatric Surgery Attending Physician, Armed Forces Specialty Hospital N°1 Quito- Ecuador.*

[2] *Pediatric Surgery Attending Physician, Hospital Institute of Social Security IESS Quito Sur. Quito- Ecuador.*

[3] *Resident Physician of Neonatology Service, Armed Forces Specialty Hospital N°1 Quito- Ecuador.*

***Corresponding Author:** Daniel Manzano Moscoso, Pediatric Surgery Attending Physician, Armed Forces Specialty Hospital N°1 Quito- Ecuador.*

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Abstract

Omphalocele is a congenital defect of the abdominal wall. The current knowledge of its etiology suggests that this defect is not due to the failure of closure or migration of the body wall. Rather, since the umbilical cord is inserted into the sac, it is considered that the omphalocele appears due to the lack of return of the viscera to the abdominal cavity. Consequently, the abdominal contents are herniated with an incidence of 1.5 to 3 per 10,000 births and are associated with multiple congenital and chromosomal malformations. We present an atypical and rare case of a 37-year-old mother; As a result of his fourth pregnancy, with an ultrasound diagnosis at 21 weeks of omphalocele with herniated fetal liver, with birth without complications by scheduled caesarean section, whose defect was considered a giant omphalocele (> 6cm), and which was treated with the placement of silo since the introduction of the herniated contents (liver) into the abdominal cavity was not achieved. The purpose of this paper is to try to explain the complexity of the management of this type of patient, and the complications related to its management, since, being of rare presentation, it becomes a complex challenge for the multidisciplinary team that must face this type of pathology.

***Keywords:** Omphalocele, Giant omphalocele, Abdominal wall defects*

Introduction

Omphalocele is defined as a congenital defect in the midline of the abdominal wall that affects the umbilical ring. It is characterized by having a 3-layer sac that covers the herniated abdominal contents (1). It has a prevalence of 1.5 to 3 per 10,000 births, with a predominance in males, and is associated with maternal age and the use of serotonin reuptake inhibitor drugs during pregnancy (2). Although the survival rate is high, there are cases of mortality associated with chromosomal and cardiac abnormalities (2).

Etiology and Pathophysiology

There have been multiple theories of the formation of this defect, among them is the failure to close or migrate the abdominal wall, but it is currently considered to be the lack of return of the viscera to the abdominal cavity. (3)

Unlike gastroschisis, the omphalocele is covered by a 3-layer sac that covers the abdominal contents and consists of an inner layer of peritoneum, an intermediate layer of Wharton's gelatin, and an outer layer of amnions. (4)

Within the variants of omphalocele, we have the giant omphalocele defined by : the following criteria: the diameter of the sac or defect greater than or equal to 5 cm, significant herniation of the liver, and a significant disproportion between the herniated viscera and those of the abdominal cavity. (4) They may be associated in 70% of cases with other malformations as described in Table 1.

Table 1: Malformations associated with omphalocele.

Anomaly	%	Frequency
Genitourinary	< 10	Bladder and cloacal exstrophy
Renal	< 10	Renal malrotations
Facial	< 10	Cleft lip and palate
Chromosomal	30 – 40	Trisomy 13 and 18, Beckwith-Wiedemann syndrome
Intestinal	40	Intestinal atresia, duplications, congenital diaphragmatic hernia, Cantrell's pentalogy.
Congenital Heart Disease	50	Tetralogy of Fallot atrial septal defects.

Source: Tinoco NJ, Medicine and Surgery Repertory. 2021; 30(2)

Prenatal Diagnosis

The diagnosis is suspicious when there are abnormal findings on routine prenatal ultrasound and most defects are diagnosed by the middle of the second trimester. Ultrasound features after 12 weeks of gestation reveal a central herniation with the insertion of the umbilical cord at its apex, covered by a peritoneal membrane, where the contents may be intestinal, hepatic, or contain multiple organs. It is suggested to correlate the size of the defect by measuring the relationship between the transverse diameters of the omphalocele and the abdomen. When the liver is contained within the omphalocele, it will be considered large and, in its absence, small. (6) On the other hand, high levels of α fetoprotein allow the suspicion of chromosomal pathologies as well as defects of the abdominal wall. (6)

Clinical Case Presentation Prenatal Background

A 37-year-old mixed-race mother; the product of his fourth feat. With normal genetic studies. In prenatal control at 21 weeks, ultrasound was used to diagnose a giant omphalocele with herniated fetal liver. Figures 1 and 2.

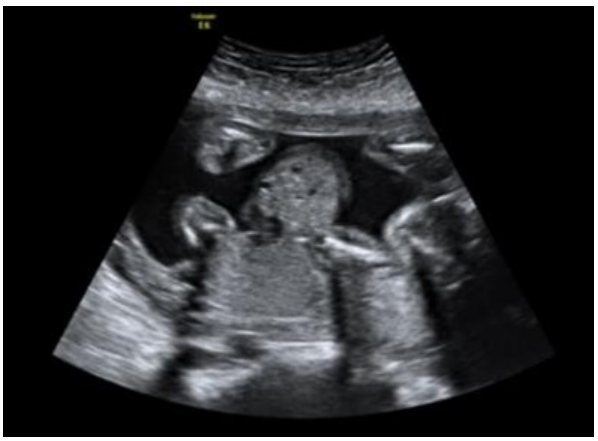

Figure 1: *Prenatal Echo 21 weeks.*

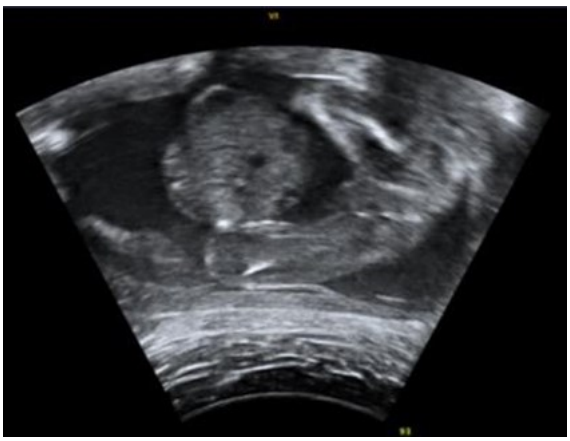

Figure 2: *Prenatal Echo 21 weeks.*

Source: Quito Military Hospital Imaging Service

Natural Background

Full-term newborn 38 weeks gestational age, male, born by elective cesarean section, with APGAR: 8-9. Capurro 38.3 weeks. Hassle-free. CLINICAL FINDINGS Newborn with vital signs and anthropometric data within normal parameters. Tables 2 and 3.

Table 2: Patient anthropometry.

Anthropometric data	
Weight	2.775 gr
Size	47 cm
Head circumference	35.5 cm
Thoracic circumference	29 cm
Abdominal circumference	26 cm

Source: Dr. Ana Cevallos. Quito Military Hospital

Table 3: Vital signs at birth.

Vitals	
Heart rate	154 ltm
Respiratory rate	57 rpm
Saturation	96% AA.
Temperature	36.5°C
Blood pressure	69/ 39

On physical examination: giant omphalocele approximately 12 cm in diameter, with intestinal loops and total liver herniation covered by the umbilical cord, which is adhered entirely to the hepatic organ. Figure 3.

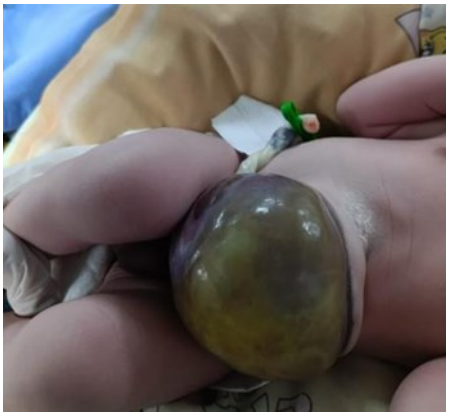

Figure 3: *Giant omphalocele.*

The patient underwent surgery at 15 hours of age to assess his congenital defect, which was a giant omphalocele (> 6 cm). However, due to complete exposure of the liver, and lack of continent abdominal cavity, it is not possible to introduce the liver, only intestinal loops, so it is decided to place a silo to continue progressive reduction. Figures 4 and 5.

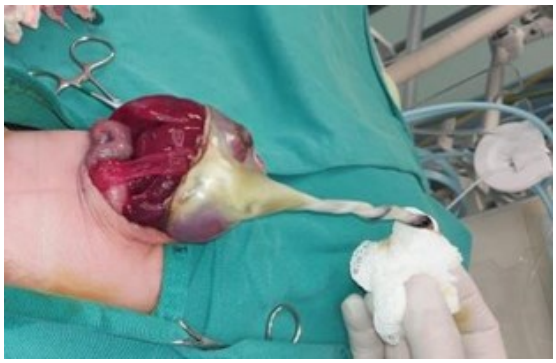

Figure 4: *Trans surgical exploration.*

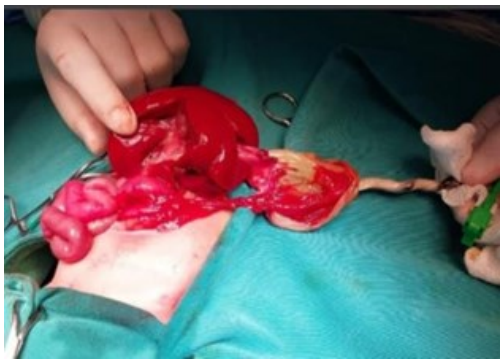

Figure 5: *Exposed liver.*

Source: Dr. Daniel Manzano. Pediatric Surgeon. Quito Military Hospital.

During his hospitalization, he remained under mechanical ventilation in moderate parameters, double sedation analgesia (fentanyl and midazolan) with episodes of reactivity, on the sixth day of life muscle relaxant was added (rocuronium 0.4 mg/kg/h) and continued with progressive introduction of the liver, monitoring intra- abdominal pressure whose measurement varied between 16 mmH2O to 20 mmH2O, but with limited progress. Figures 6 and 7

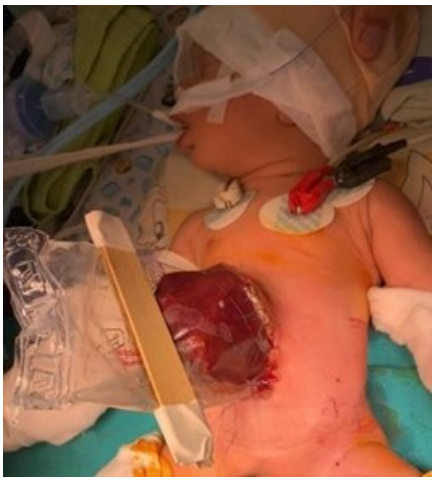

Figure 6: *Silo Management in Neonatology.*

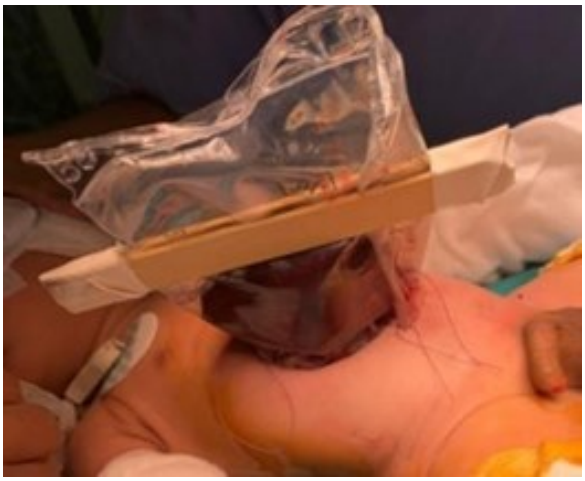

Figure 7: *Silo reduction.*

Source: Dr. Daniel Manzano. Pediatric Surgeon. Quito Military Hospital.

At 15 days of age, a new surgical exploration was performed, where the silo was removed and progress was made in the introduction of the hepatic organ into the abdominal cavity through a bilateral transverse lateral incision of the abdominal wall, managing to bring the introduction of the liver to the skin. Figure 8.

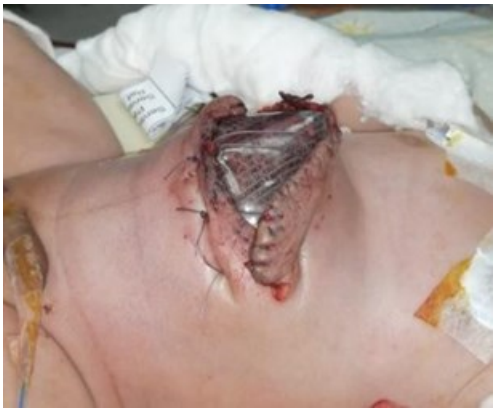

Figure 8: *Reduction of the liver to the abdominal cavity.*

At 20 days of age, the abdominal defect was closed by maintaining intra-abdominal pressure of 18 mmH2O, the skin of the abdomen remained shiny, the distal capillary filling was preserved, and iliac pulses and media were present.

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24 hours after his last intervention, the patient remained intubated on invasive mechanical ventilation but with high ventilatory parameters, presented desaturation that reached 84%, and blood pressures between the 5th and 50th percentile. Prolonged heart rate between 97-162, with 3-4" capillary filling. Subsequently, he presented sudden bradycardia up to 54 and saturation of 60%,advanced resuscitation was given, he received a dose of 0.5 ml of adrenaline, a bolus of saline solution with transient improvement, due to the persistence of bradycardia and desaturation, a second dose of 0.6 ml of adrenaline was administered without definitive recovery ofheart rate, and the patient finally died.

Discussion

Omphalocele is a rare abdominal wall defect, with a prevalence of 1.5 to 3 per 10,000 live births; Mortality was found to be related to the presence of associated malformations in 50% to 60%. Among them, the most associated chromosomal diseases are trisomies 13, 18, and 21; Turner's syndromes, Klinefelter's syndromes, and tri- ploidies, and 50% of cases are associated with cardiac abnormalities. (7,8) Over time, its diagnosis and management have been optimized, directly benefiting the patient. Patient survival in the last two decades has increased from 70% to 90%, thanks to improvements in surgical technique, mechanical ventilation, and parenteral nutrition. (9), but going through a series of surgeries that also carry great risks, which, however, once overcome, the affected children can enjoy a state of health and quality of life similar to that of the general population. (10) . In a study carried out by Borbón in 1995, in a 5-year review in Costa Rica, they found 10 patients with herniated livers of which 30% died, and this is due to the difficulty that exists to face the edges of the defect, which causes closure to be deferred and this in turn postpones the start of the oral route and increases the time of hospital stay(11). Giant defects of the abdominal wall considered larger than 6 cm.

(12) with hepatic herniation relative to giant omphaloceles without total hepatic herniation have the worst prognosis (11). This is also because there is not enough space in the abdominal cavity to be able to introduce the liver easily since respiratory distress can occur secondary to the mass effect exerted by the liver inside the abdomen on the thorax. which would trigger the death of the patient due to the pulmonary hypertension that is generated. The complications that occurred in our patient in whom silo placement was performed, delayed closure of the defect that was reached after 21 days, but with poor tolerance by the patient due to the complications previously exposed, which led to his death since the degree of visceral- abdominal disproportion made it impossible to introduce the organ into the cavity without being able to avoid hemodynamic and respiratory compromise.

Conclusions

Patients with giant omphalocele in whom there is no total evisceration of the liver or only the presence of intestinal loops, it is possible to achieve either primary or secondary closure after silo placement or using minimally invasive maneuvers such as the use of synthetic prosthetic material (Duoderm), but in patients who have several exposed organs or the liver in its entirety, Its treatment becomes a real challenge for the surgeon and his team since the complications that arise when using any type of maneuver for its reduction can be associated with complications that would lead to high mortality.

Conflict of Interest

The authors declare no conflict of interest.

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