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Omphalocele Diagnosis Problem in Rural Environment

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Abstract

Introduction: An omphalocele is a congenital malformation due to a failure of closure of the umbilical ring with exteriorization of the abdominal viscera covered by the translucent and avascular amniotic membrane. We report a case of giant omphalocele unrecognized at birth followed by a review of the literature. **Observation:** This was a 4-year-old girl admitted to the department for umbilical swelling that had been present since birth. Clinical and paraclinical examinations made it possible to objectivize a post-omphalocele hernia with hepatic content plus ectopic accession of the left ureter, the surgical procedure consisted of reintroducing the liver associated with a cure of the hernia + ureterovesical reimplantation. **Conclusion:** Omphalocele is a congenital malformation, antenatal diagnosis is possible during morphological ultrasound from the 12th week.

Keywords

Omphalocele, Ectopic Left Ureter

1. Introduction

An omphalocele is a congenital malformation due to a failure of closure of the umbilical ring with exteriorization of the abdominal viscera covered by the translucent and avascular amniotic membrane. It is a rare congenital malformation that falls within the framework of polymalformation syndrome. We report a case of omphalocele with hepatic content plus ectopic accession of the left ureter.

2. Observation

This is a 4-year-old girl, the third child of a family of four from a full-term preg-

nancy with a single ultrasound during prenatal consultations. The delivery took place in a regional community health center, after which a wound centered on the umbilicus with a false membrane was noted. A dressing was applied to this wound for a few weeks until complete healing was achieved. The evolution was marked by the addition of a reducible, non-painful, non-throbbing, reducible umbilical swelling which gradually increased in volume to the current size. Faced with this swelling, she was referred to us for treatment. On admission, the examination reveals an umbilical swelling on a non-expanding scar (Figure 1), on palpation, we note a firm, painless, irreducible, non-beating swelling. General signs WHO 1, colored conjunctiva, pulse 84 beats per minute, respiratory rate 26 cycles per minute. An abdominal and pelvic CT scan revealed the appearance of a polymalformation syndrome grouping together a hepatocele with L3 hemi-vertebra and ectopic abutment of the left ureter possibly on the urethra (Figure 2). The pre-operative assessment was requested, and the result is as follows: rhesus A positive grouping, hemoglobin level = 9.69 g/d, hematocrit 29.6%, blood sugar = 0.70 g/l TP = 74.5% TCK= 39.1 seconds.

Figure 1. Omphalocel before operation.

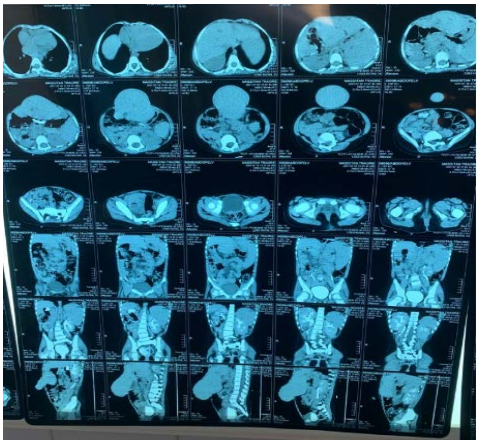
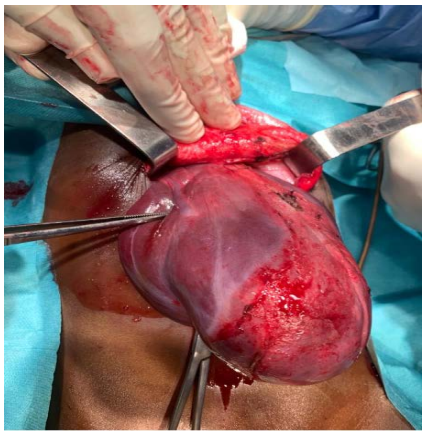
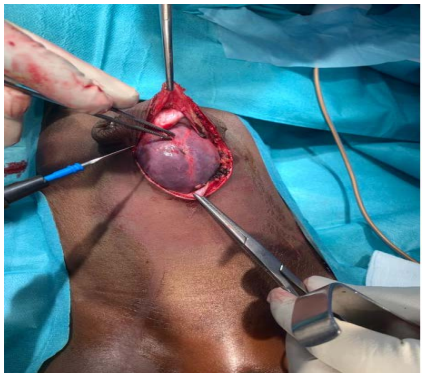


Figure 2. Radiologic of omphalocel.

Diagnosis retained post-omphalocele hernia plus ectopic blockage of the left ureter. The treatment consists of a cure of the hernia + ureterovesical reimplantation. The postoperative course was simple and the excision was performed on day 5. The patient was seen again 1 month later with a normal clinical examination (Figures 3(a)-(c)).



(a)



(b)

(c)

Figure 3. ((a) (b)) In per-operation; (c) After operation.

3. Discussion

An omphalocele is a failure of closure of the anterior abdominal wall character-

ized by the absence of muscle, fascia and skin. The exteriorized viscera are only covered by the peritoneum, amnion and Wharton's jelly [1]. This is an embryopathy where associated malformations are common, particularly cardiac and urinary. It is sometimes part of a polymalformative syndrome [2] [3]. Omphalocele is a rare pathology, affecting 1/4000 births [4] [5]. Most authors report the predominance of the male sex [6].

The pathophysiology is poorly understood, and several theories have been put forward.

An arrest of the development of the abdominal cavity between the 8th and 12th week of gestation, a defect in reintegration of the primitive loop, an involvement of the AP-2α transcript involved in the morphogenesis of the anterior abdominal wall (mouse model), an early embryonic dysgenesis of the embryonic germinal disc, embryonic dysgenesis of the placodes ectodermal [7].

Omphalocele is favored by very young or, on the contrary, advanced maternal age, All causes of fetal macrosomia, primarily diabetes, and low socio-economic factors. In the present clinical case, the risk factor found was low socio-economic level [8].

Antenatal diagnosis of omphalocele is possible during a morphological ultrasound of the 2nd trimester [9]. In our case, the only ultrasound performed could not suggest the diagnosis of an omphalocele. This could be explained by the lack of means to carry out the prenatal assessment and the lack of qualified personnel in peripheral structures. The possibility of antenatal diagnosis by prenatal ultrasound makes it possible to schedule the birth in a specialized center.

The postnatal diagnosis of an omphalocele is clinical. It is established at birth by the discovery in the newborn of a pedunculated or sessile swelling of variable volume and content, located in the umbilical region. Additional examination makes it possible to look for associated malformations. Depending on the different types of associated malformations we can have Cantrell's pentalogy, body-stalk anomaly, Beckwith-Wiedmann syndrome [1]. In our patient, the CT scan showed a giant omphalocele with hepatic content associated with a genitourinary anomaly (ectopic connection of the left ureter to the urethra).

The management of an omphalocele depends on the size, the treatment of giant omphaloceles still remains a topical subject. The reintegration of exteriorized viscera into an insufficiently developed abdominal cavity has hemodynamic and respiratory consequences that must be considered for the surgical decision. Immediate wall closure by simple wall approximation is at risk. It can hamper the hemodynamic state of the newborn [10].

Other therapeutic alternatives exist to minimize the morbidity and mortality associated with this surgical technique. Immediate parietal closure by skin coverage using the Gross technique is simple. It reduces abdominal septic complications and abdominal hyperpressure. On the other hand, this technique requires subsequent correction of the hernia. The most used technique is progressive reintegration by silo (Schuster method) [11].

The principle of the non-operative technique is to epidermise the omphalocele

and treat the hernia remotely. This epidermization (tanning) makes it possible to avoid major surgery in the neonatal period [12]. Our therapeutic procedure consisted of a cure for the hernia + ureterovesical reimplantation) 4 years after epidermization obtained by bandaging the omphalocele. This technique is simple and gives very good results in the management of giant omphaloceles because it can be carried out in the absence of an adequate intensive care unit.

4. Conclusion

The management of giant omphaloceles varies according to the authors and depends on the existing technical platform. In our condition, where assisted ventilation is not available, the tanning epidermization technique gives good results and remains the best therapeutic alternative.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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