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Cloacal exstrophy associated with gastroschisis: Case report of a rare association with favorable outcome

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ABSTRACT

Abdominal wall defects are congenital malformations representing a challenge for pediatric surgeons. This paper presents a rare case of difficult surgical decision and clinical management, leading to a favorable outcome brought about by the multidisciplinary treatment of a patient with cloacal exstrophy associated with gastroschisis.

Abdominal wall defects are congenital malformations that represent a formidable challenge for neonatologists and pediatric surgeons [1]. This affection includes a wide range of defects in the closure of the anterior abdominal wall, such as ectopia cordis, gastroschisis, omphalocele, bladder exstrophy, and cloacal exstrophy [1,2].

Gastroschisis is a defect of the anterior abdominal wall, which is typically located to the right of the umbilical ring and commonly results in the herniation of the gut and occasionally of the stomach, urinary bladder, ovaries, and tubes without a surrounding membrane, reaching as far as the amniotic cavity [1]. The incidence of gastroschisis is estimated at 3.3/10,000 live births. Currently, this estimated global incidence of gastroschisis varies between 0.4 and 3/10,000 births and appears to be increasing [1,3].

Cloacal exstrophy, first described by Littre in 1709 [4], is one of the most serious and complex congenital anomalies that is compatible with life. It is characterized by the failure to closure the lower urinary tract and commonly results from an abnormality in the development of the mesoderm. This anomaly affects 0.76/100,000 births, with a higher incidence reported in females. It is usually associated with other anomalies, such as omphalocele, imperforate anus, spinal cord abnormalities, abnormalities of the lower limbs, genitourinary malformations, single umbilical arteries, and short bowel syndrome [4–8].

To the best of our knowledge, gastroschisis associated with cloacal exstrophy has not been reported in prior medical literature.

1. Case report

This case report analyzed a 36-week-old premature newborn (PN), with no apparent cause for his premature birth, from a young, healthy and non-consanguineous couple with no history of exposure to teratogens. The mother was 30 years old, who had had two prior successful pregnancies as well as eight doctor's visits for appropriate prenatal follow-up. Serological tests were performed, which proved to be negative for HIV and HBsAg, nonreactive to VDRL, immune to toxoplasmosis with no glycemic alterations, and with no signs of infection during pregnancy. Ultrasound was performed at 35 weeks, which showed gastroschisis and a single umbilical artery. The patient was referred to the Fetal and Maternal Medicine Center of the "Hospital das Clínicas" of the "Universidade Federal de Minas Gerais" (UFMG).

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The 36-week-old newborn was born by c-section, with a weight of 2.980 g (p75), length of 45 cm (p25), and a head circumference of 28 cm ($p < 10$), with meconium-stained amniotic fluid and an Apgar score of 9/9. Physical examination showed a large gastroschisis (defect on the anterior abdominal wall to the right of an intact umbilical cord, without traces of membrane) with an exteriorization of dilated bowel loops with swollen and thickened walls associated with cloacal exstrophy (a defect in the midline, with only vesicointestinal plaque, imperforate anus, ambiguous genitalia, and pubic symphysis diastasis). No evidence of omphalocele could be observed. Umbilical cord examination confirmed a single umbilical artery (Fig. 1).

The newborn was referred to the neonatal intensive care unit (NICU), where management and postnatal care was begun. All of the herniated contents were wrapped in moist and warm sterile dressings, umbilical venous catheterization was applied for fluid replacement, and antibiotic prophylaxis (ampicillin and gentamicin) was administered (Fig. 2).

Within 4 h of life, the newborn was taken to the operating room. Surgical procedures were performed under general anesthesia after tracheal intubation, adequate volume resuscitation, and prevention of hypothermia, and observed a visceroabdominal disproportion (massive eviscerated contents relative to the small size of the abdominal cavity), an abdominal wall defect to the right of the umbilical cord, no membrane, and a meconium exteriorization by intestinal loops interrupted in the midline at the vesicointestinal plate.

Due to the complexity and rarity of this defect of the abdominal wall (gastroschisis and cloacal exstrophy), as well as the absence of reports in the literature of this association, the surgical team decided to proceed to a staged surgical treatment of gastroschisis with the preservation of the vesicointestinal plate for subsequent surgical correction of the cloacal exstrophy. Thus, it was necessary to place a plastic bag, as the primary closure of the abdominal wall was not possible. The anatomical defect was expanded with a cranial longitudinal incision and the exteriorized loops were covered with a plastic bag used to transfer the blood products, according to that described by Miranda et al. [6]. The edges of the plastic bag were sutured bilaterally into the edges of the muscular aponeurosis with a nonabsorbable suture (Prolene 4.0) in order to create an extra-abdominal silo connected to the beginning point of the bladder plate covering the vesicointestinal plate (Fig. 3).

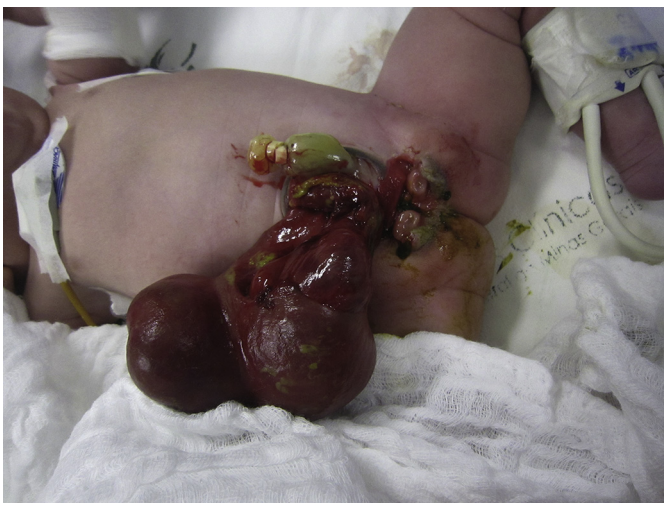

Fig. 1. Cloacal exstrophy associated with gastroschisis (see anatomical implantation of the umbilical cord).

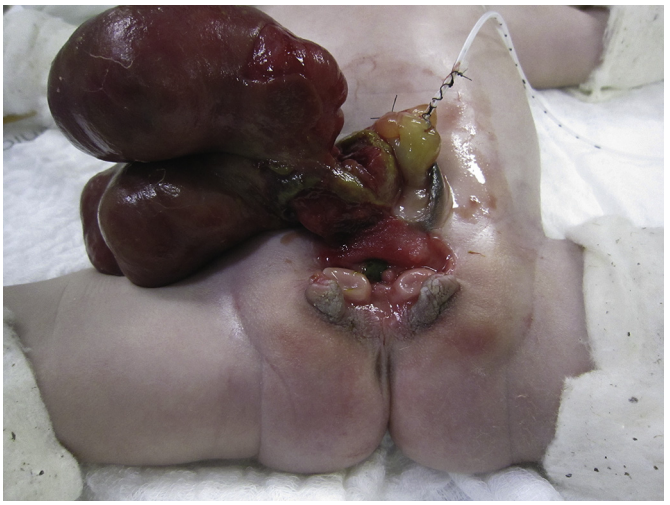

Fig. 2. During preoperative period with umbilical vein catheterization for fluid replacement and antibiotic prophylaxis.

After the first surgical procedure, the newborn was kept under continuous sedation and curarization in the NICU in order to facilitate the gradual accommodation of the herniated content. The silo was reduced by the surgical team every two days so that all of the viscera could be gradually introduced into the abdominal cavity allowing for the staged closure of the abdominal wall. On the eighth day of life, the second procedure was performed: removal of the silo and complete closure of the anterior abdominal wall, preserving the vesicointestinal plate. At this time it was impossible to measure intra-abdominal pressure by indwelling catheter due to the cloacal exstrophy (Figs. 4 and 5).

Antimicrobial prophylaxis was maintained until the closure of the abdominal wall had been completed; urinary prophylaxis was maintained due to the opening of the vesicointestinal plaque and the consequent risk of urinary tract infection. The patient was kept on mechanical ventilation for 13 days. Parenteral nutrition was maintained for 23 days. Enteral nutrition was started at 15 days of life and was progressively increased.

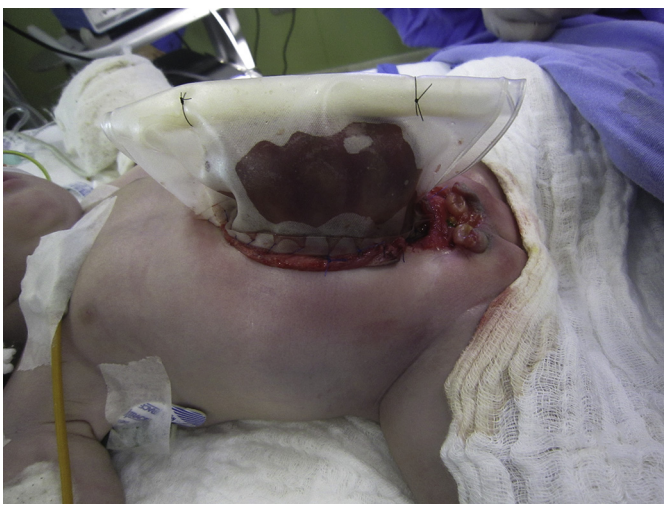

Fig. 3. First surgical procedure: placement of a plastic bag, as the primary closure of the abdominal wall was not possible. The anatomical defect was expanded with a cranial longitudinal incision, and the exteriorized viscera were covered with a plastic bag used to transfer the blood products.

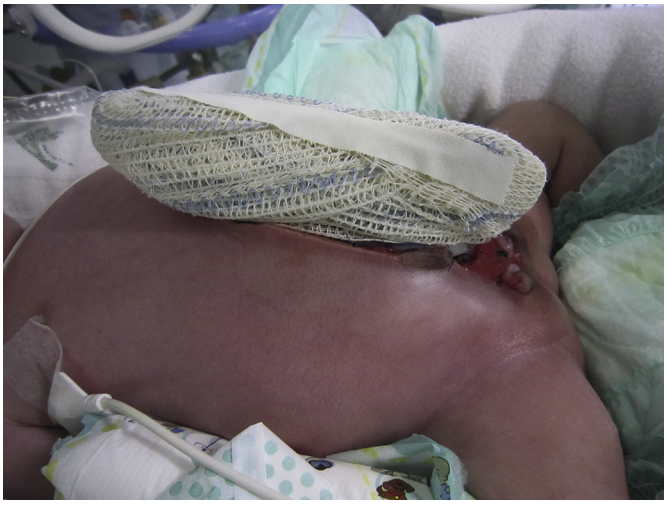

Fig. 4. Silo reduction by the surgical team every two days. Newborn was kept under continuous sedation and curarization.

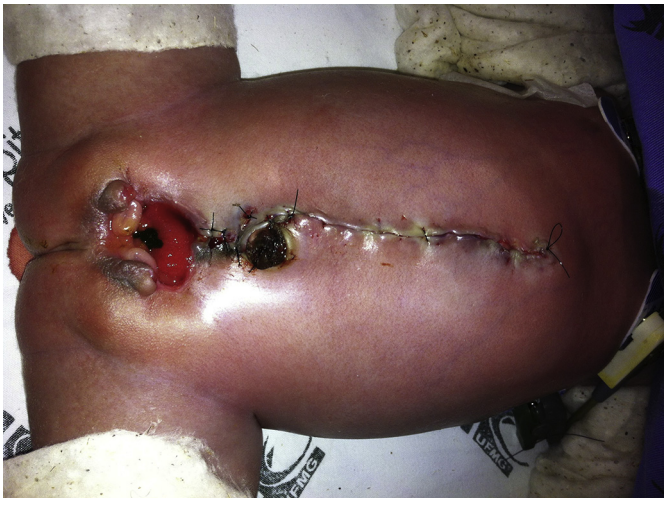

Fig. 6. Second procedure performed on the 8th day of life: removal of the silo and closure of the anterior abdominal wall with preservation of the vesicointestinal plaque.

The surgical correction of cloacal exstrophy was begun on the forty-fifth day of life, divided into two phases for better tolerance of the neonate to the procedure. The first approach consisted of bilateral anterior pelvic osteotomy performed by the orthopedic team. The next day, the newborn underwent dissection and closure of the urethral plate, ureteral catheterization, and cystostomy, end colostomy and any approximation of the pubic symphysis. To avoid the mobilization of the lower limbs, the patient was kept in Bryant's position, setting the lower limbs by using the same bandaging during a four-week period until bone callus formation and a satisfactory closure of the pubic symphysis had been achieved. This bandage was used in this service due to the unavailability of suitable weight and size of the child Sterna fasteners, hindering the process of patient immobilization (Figs. 6 and 7).

The cariotype was 46 XX. During prolonged hospitalization, no other malformations were observed, which was confirmed by ultrasound to rule out renal pathologies. Due to low doses of follicle-stimulating hormone (FSH), luteininzing hormone (LH), adrenocorticotropic hormone (ACTH), and cortisol, secondary adrenal insufficiency was diagnosed. Thus, a physiological dose of hydrocortisone was maintained. Transfontanelle examination showed sulci and gyri in the anterior region, with an enlarged interhemispheric fissure associated with a slight increase in the former vacuum lateral ventricles. During this time, blood transfusions and antibiotics were administered after a positive blood culture for *Streptococcus pneumoniae* (*S. pneumoniae*).

Having presented a favorable evolution of his medical condition, the patient was discharged after 80 days of life. At this time, the patient was able to properly tolerate enteral feeding, with excrements through the colostomy and epispadic urethra. The abdominal wall was properly closed, leaving no evidence of hernia.

The 18-month-old patient was followed monthly by a multidisciplinary team and remains asymptomatic, with normal laboratory tests and adequate growth and development.

2. Discussion

An abdominal wall defect in newborns represents a dramatic clinical picture and a challenge for the pediatric surgeon [5]. Despite the increased survival rates in recent decades, the results are still controversial, with a scarcity of knowledge regarding the etiology, anatomy, and embryology, as well as the role of prenatal diagnosis,

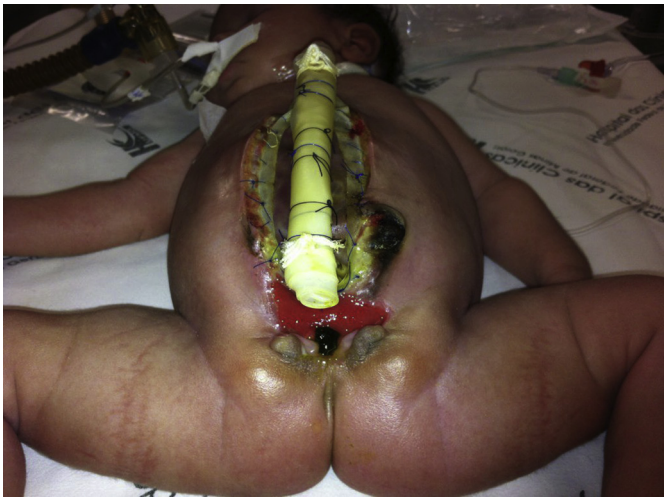

Fig. 5. 6th day of life with complete reduction of the extra-abdominal silo.

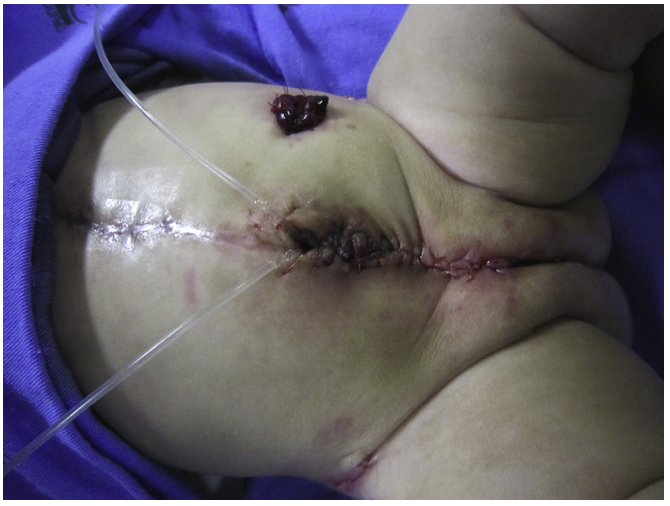

Fig. 7. Cloacal exstrophy correction: immediate postoperative period: dissection and closure of the vesical plate, ureteral catheterization, cystostomy, terminal colostomy, and approximation of the pubic symphysis.

childbirth, and postoperative management [3]. Recent studies proposed a theory that these defects of the abdominal wall occur due to vascular insufficiency, thus causing an imbalance between proliferation and apoptotic cell death during embryonic development [2]. Despite these studies, the exact etiology is still unknown.

The embryological origin of ectopia cordis, gastroschisis, and bladder and cloacal exstrophy is more commonly reported in the literature and differs from the origin of omphalocele. Early defects are most likely caused by an impaired closure of the ventral wall of the folds attached bilaterally to the mesoderm during the fourth week of gestation. In the case of omphalocele, a defect is attributed to the failure to return the bowel to the abdominal cavity at around the sixth to the tenth week of pregnancy, resulting in a physiological hernia in the umbilical cord. This anomaly differs from gastroschisis, given that a membrane covers and protects the viscera from the amniotic fluid, in turn preventing the process of intestinal serositis. In cases of exstrophy, despite the unsuccessful migration of the mesoderm's lateral folds, a rupture of the cloacal membrane (medial mesoderm) occurs before its fusion with the urorectal septum (cloaca – the confluence of the genitourinary and gastrointestinal tract), which most commonly happens during the eighth week of embryological development. The cloacal exstrophy shows a bladder divided in two halves by a strip of cecum associated with the prolapse of the terminal ileum, hemi falus, and pubic diastasis associated with different sizes of omphalocele [1,2].

Gastroschisis is an isolated and sporadic defect, though there are rare familial cases and some possibility of genetic determination [5]. The incidence of abnormalities associated with gastroschisis ranges from 10% to 20%, referent mainly to abnormalities of the gastrointestinal tract and cryptorchidism. The most common is intestinal stenosis or atresia, followed by Meckel's diverticulum and intestinal duplication [5]. Cloacal exstrophy has a varied incidence according to geographic location and is associated with several anomalies. Its incidence also increases when presented as a component of such conditions as imperforate anus, omphalocele, and spina bifida [4,7].

Through obstetric ultrasound, studies in animals have shown that these abdominal wall defects occur in the early stages of pregnancy and compromise the results of embryo formation and development [1,2,4,5].

The majority of defects of the abdominal wall can be diagnosed by ultrasound, allowing for prenatal diagnoses, counseling, and family education, as well as medical referrals to major fetal medicine centers. More recently, prenatal diagnoses have been made possible by means of magnetic resonance imaging (MRI). Despite the advances, however, no consensus has been reached regarding the best time to interrupt a pregnancy or the most recommended type of delivery (vaginal or cesarean) [9]. Prolonged exposure to the amniotic fluid can damage the intestinal wall. Intestinal injury is the greatest predictor of morbidity and mortality in these patients. Currently, it is impossible to determine this degree of injury during the prenatal period [1–3].

In the literature, some risk factors have already shown a direct relationship with gastroschisis, including young maternal age (under 20 years), smoking, use of illicit drugs, drugs with vasoconstrictor effects, and environmental toxins [1]. Fetuses can also present intrauterine growth restriction, prematurity, and stillbirths.

Immediately after birthcare and resuscitation, exposed viscera should be protected or enclosed in moistened sterile equipment. This coverage promotes protection against hypothermia, trauma, and infections. As soon as possible, newborns should be referred for surgical correction of the defect in the abdominal wall. This correction can be performed by means of primary or staged closure using transparent synthetic bags if any externalized content does not fit in the abdominal cavity [6]. This decision is made by the surgeon according to the level of tension in the abdominal wall after the reduction of the content and the surgeon's attempt to suture the wall by means of indirect measurement parameters for intra-abdominal pressure. A high tension closure of the abdominal wall can compromise the child's ventilation and hemodynamic status, which can develop into intra-abdominal compartment syndrome. In such cases, the placement of a silastic pouch allows for the protection of the viscera as well as the gradual reduction of herniated contents within the abdominal cavity according to accommodation and adaptation [5,6].

Surgical correction of cloacal exstrophy is normally performed in the first 48 h–72 h of life. The surgical procedure is defined according to the associated anomalies and the specificity of each case. Patients undergo the reconstruction of the genitourinary and gastrointestinal tract, closure of the defect in the abdominal wall, and a pubic symphysis approach, emphasizing the separation between the cecum and the two halves of the bladder, the closure of the defect in the abdominal wall (omphalocele, in most cases), and the preservation of the gut function. The success of this operation results in an adequate abdominal wall closure, the preservation of the renal function, the prevention of a short bowel syndrome, the functional and aesthetic construction of the genitalia, as well as an adequate urinary and fecal continence. If the defect in the abdominal wall is extensive, staged correction is recommended. In cases of giant omphalocele, conservative treatment with dressings that allow for the epithelialization and subsequent correction of incisional hernia is possible. Osteotomy is necessary to achieve an adequate approximation of the pubic symphysis [4,7–11]. Hence, the treatment of cloacal exstrophy is complex and requires several steps.

3. Conclusion

To the best of our knowledge, there are no reports in the literature on the simultaneous occurrence of huge gastroschisis and cloacal exstrophy. Despite the technological advances available today and improvements in patient survival, these two diseases also represent a challenge for pediatric surgeons and their staff. The present study reported on a rare case of difficult surgical decision-making and clinical management, which resulted in a favorable outcome due to the application of a multidisciplinary treatment.

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