

Cloacal Exstrophy: Documenting a Particular Case in Yaoundé, Cameroon

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Abstract

Cloacal exstrophy in its most complex form with OEIS syndrome is characterized by the existence of an Omphalocele, Exstrophied bladder, Imperforate anus, and Spina bifida. We report a particular case of Cloacal exstrophy with OEIS complex and further birth defects. The diagnostic and therapeutic approaches, as well as difficulties encountered in a limited resource setting, are highlighted. This was a 1-day old neonate referred for the management of multiple congenital malformations, including antenatal diagnosis of malformative uropathy. On admission, the clinical findings included: a type 1 omphalocele, an anorectal malformation with a recto-urinary fistula and a covered lumbosacral dysraphism. Paraclinical examinations with cardiac and trans fontanelle ultrasound revealed associated cardiac anomaly and findings in favor of lombo-sacral dysraphism. Supportive care was given and surgical reconstruction of birth defects on day 20 was done. In conclusion, cloacal exstrophy is a rare morbid congenital polymalformative syndrome in neonates, especially when presenting with OEIS complex or syndrome. They require prompt diagnosis and immediate postnatal multidisciplinary management, with long-term follow-up for a favorable outcome.

Keywords: Cloacal exstrophy, OEIS Syndrome, epispadias-exstrophy

Introduction

Cloacal exstrophy in its most complex form with OEIS syndrome is characterized by the existence of an Omphalocele, Exstrophied bladder, Imperforate anus and Spina bifida. These are grouped under the term OEIS complex or syndrome [1, 2]. It is often associated with other malformations, involving the gastrointestinal and skeletal spheres [1]. It was first described in 1709 by LITTRE, and for a long time was considered a devastating pathology due to infectious, metabolic or nutritional complications associated with short bowel syndrome and intestinal obstructions [3, 4]. In effect, the prognosis, although dependent of associated anomalies, remains uncertain in context with low technical platform.

Case Presentation

This is a newborn baby on its first day of life referred for the management of a congenital malformation noted at birth. The mother was primiparous, aged 22 years, there wasn't a notion of consanguinity. Four antenatal consultations had been carried out in a remote rural area with an undocumented infectious workup. We did

not find any exposition of teratogenic substances. There was no history of chronic or pregnancy-related pathology in the mother. She had received iron and folic acid throughout the pregnancy in adequate doses. A 33 weeks gestation obstetrical ultrasound had shown pyelocal dilatation of the right kidney in favor of fetal uropathy. Delivery was per vaginal at 35 weeks' gestation + 3 days, with amniotic fluid described as meconium-stained and fetid. The birth weight was 2000 g with an Apgar score of 8 and 10 at the 1st and 5th minute respectively.

Physical examination on admission revealed a newborn with normal vital and anthropometric parameters for gestational age. Signs of prematurity with an estimated gestational age of 36 weeks according to the Ballard score. We had a type 1 omphalocele with an umbilical hernia (fig1), cloacal exstrophy with two exstrophied hemi bladders, separated with everted cecum (fig1). There was a sexual differentiation anomaly, as well as anorectal malformation with recto-urinary fistula (fig2) and a covered lumbosacral dysraphism (fig3).



Figure 1 and 2: omphalocele type 1, cloacal exstrophy, anal imperforation, external genitalia ambiguity



Figure 3: spina bifida occulta meningocele

Cardiac ultrasound revealed an ostium secundum type inter-atrial communication of approximately 2.5 cm, the interventricular septum being intact (fig4). Abdominal and pelvic ultrasound concluded there was bladder exstrophy, with the integrity of the intra-abdominal organs. The transfontanellar ultrasound showed a posterior superficial formation of pure anechoic content measuring 38 x 17 x 33 mm, that is 11 cc in volume, with an opening into the medullary canal, suggesting a meningocele. We requested karyotype, which could not be done due to lack of financial means.

The newborn was put on triple antibiotic therapy (cefotaxim, ampicillin, and gentamycin) in view of the meconium-stained and

fetid coloration of the amniotic fluid for 10 days according to the department's protocol. Enteral feeding with breast milk was started on the second day of hospitalization due to the absence of secretions coming back through the nasogastric tube. Twice daily wet dressings of the abdomen were performed until the surgery.

The child underwent surgery on the 20th day of life. This consisted of a two-step surgical intervention, of which the first comprised an exploratory laparotomy with a peri omphalocele incision and the reconstruction of digestive and urogenital defects. The second step consisted of lumbosacral dysraphism meningocele repair.



Figure 4: Ostium secundum type atrial septal defect of about 2.5 cm

Discussion

Cloacal exstrophy is an extremely rare polymalformative syndrome with prevalence estimation as low as to 1/400000 live births. It is believed to be predominant in male neonates when the gonads and external genitalia can be well appreciated. This was not the case with our patient due to sexual differentiation anomalies. As the diagnosis of this syndrome is difficult to pose, the very low global incidence reported is thought to be an underestimation by some authors. However, to the best of our knowledge, no such cases had earlier been documented in our context [5-7].

Although a karyotype could not be done in our situation, young maternal age, just as fetid and meconium-stained amniotic fluid as infection criteria, were nonnegligible risk factors for congenital malformations [8-11]. On the other hand, the antenatal ultrasound-based diagnosis of malformative uropathy was a pertinent finding of pregnancy follow-up, which was in favor of Cloacal exstrophy. Early referral to specialized structures for better management was preconized, but not honored. This was probably because of parental psychological unpreparedness to accept the antenatal diagnosis [12-19]. As a matter of fact, antenatal diagnosis when accepted by parents offers the advantage of a better understanding of the condition, buys time for financial and moral preparation for treatment, makes in utero transfer possible, and enables preparation for maternity [5, 19-22]. The clinical diagnosis of cloacal exstrophy, in this case, was not obvious at birth, as extra malformations were confusing, with a necessity for literature review.

The management of the pathology was multidisciplinary including neonatal resuscitation, supportive care, antibiotherapy, and finally surgery. This was planned by taking into consideration the urgencies presented by the patient, given visceral exposure with the risk of hydro-electrolytic imbalances, superinfection, necrosis, and multi-visceral failure. Surgery was only practiced once the neonate was safe enough, contrary to 72 hours recommendations in neonatal surgical emergencies [23-28].

The surgical interventions consisted of a patient-adapted protocol

with the successful reconstruction of the genitourinary and anorectal tracts, followed by omphalocele repair in the first stage. While the second stage of the intervention consisted of mentoplasty. The interventions were successful and no immediate postoperative complications were noted.

Conclusion

Cloacal Exstrophy is a rare morbid congenital polymalformative syndrome, especially when presenting with the OEIS complex. We reported a particular case in our setting associated with further defects which may render diagnosis difficult, with a need for literature review. Regular and effective antenatal follow-up with obstetrical ultrasound had contributed to early antenatal diagnosis. However, the psychological unpreparedness of parents was an obstacle to referral and prompt intervention. The management of the neonate at birth was multidisciplinary and interventions were adapted to the immediate patient's medical needs. This stabilized and made the patient operable before surgical reconstruction could be practiced, and the outcome was favorable.

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