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GIANT OMPHALOCELE WITH SUCCESSFUL CONSERVATIVE MANAGEMENT IN A LOW INCOME SETTING: A CASE REPORT

Sylvia Tochukwu Echendu[1], Boniface Chukwuneme Okpala[1,2,3], Joseph I Ikechebelu[1,2,3], Louis A Nwajiaku[1], George Uchenna Eleje [1,2,], Cyril Emeka Nwachukwu[1,4], Chukwuemeka Jude Ofojebe[,2,3], Chijioke O Ezeigwe[2], Augusta Nkiruka Okpala[5], Ngozi Esther Umeadi[6]

Affiliations:

[1] Life international Hospital, Awka, Anambra State. Nigeria

[2] Department of Obstetrics and Gynecology, Nnamdi Azikiwe University, Awka. Anambra State.Nigeria

[3]Department of Obstetrics and Gynecology, Nnamdi Azikiwe University, Teaching Hospital

PMB 5025 .Nnewi, Anambra State.Nigeria

[4]Department of Anesthesia, Nnamdi Azikiwe University,Awka & Teaching Hospital PMB 5025 Nnewi, Anambra State.Nigeria

[5]Department of Family Medicine, Nnamdi Azikiwe University, Teaching Hospital

PMB 5025 .Nnewi, Anambra State. Nigeria

[6]Department of Pediatrics, Nnamdi Azikiwe University, Teaching Hospital

PMB 5025, Nnewi, Anambra State. Nigeria

***Corresponding Author:**

Boniface Chukwuneme Okpala

Abstract

An omphalocele is a rare midline anterior abdominal wall defect with unknown cause. It may be a small or a giant omphalocele. Management of giant omphaloceles is challenging, it can either be conservative then delayed surgical closure or immediate surgical closure. We report a case of male neonate with a giant omphalocele who was delivered by a 33 year old primigravida through an elective cesarean section. The baby was at a stable condition at delivery and throughout hospital admission. He subsequently had successful conservative management. Delaying the surgery expectedly will temporize for gradual return of the content of the omphalocele into the abdominal cavity. This prevented complications associated with immediate closure.

Keywords: Anterior abdominal wall defect, omphalocele, exomphalos, conservative management.

Introduction:

An omphalocele also known as exomphalos is a rare anterior abdominal wall defect at the umbilical base, with the herniation of the abdominal content into it. In contrast to developed countries where better pre-natal diagnostic tools, intrapartum/post-partum cares with improved surgical care are available, omphalocele still remains a major cause of neonatal morbidity and mortality in the developing countries like Nigeria [1].

It occurs in approximately 1 in 3000 to 10000 live births [2,3,4]. The etiology of omphalocele is not known, though many theories have been postulated, which include failure of the midgut to return to the abdominal cavity by 10 to 12 weeks gestational age, failure of lateral mesodermal folds to migrate centrally, and persistence of body stalk beyond 12 weeks of gestation [5].

The treatment of omphalocele depends on many factors, which includes the size, associated congenital abnormalities, and the gestational age at delivery. It can either be conservative or immediate/delayed surgical approach. The ultimate goal of therapy is to return herniated intestinal loops to the abdomen and to close up the skin and fascia without causing excessive intra-abdominal tension [6,7]. Immediate and sometimes delayed surgical management of large omphalocele predispose to hemodynamic and respiratory complications as a result of viscero-abdominal disproportion and associated cardio-respiratory anomalies [8]. Conservative management of such cases will avert such complications especially in low resource countries like Nigeria with poor neonatal intensive facility for management of post-surgical patients. There is need to wait for the bowels to completely return into the abdominal cavity before surgical repair should be carried out.

We present a case of giant omphalocele that was successfully managed conservatively in our hospital.

Case Report:

A term male neonate delivered on 23rd June, 2020 through elective caesarean section under subarachnoid block by a 33 year old primigravida on account of suspected anterior abdominal wall defect. He had good APGAR scores at delivery and weighed 3.1 kilograms. The mother had regular ante-natal care. She had several conflicting ultrasound reports with regards to diagnosis of omphalocele. Few of the ultrasounds done missed the diagnosis, while the scan done by a consultant radiologist made the diagnosis of anterior abdominal wall defect and decision to deliver by elective cesarean section was made on account of the result. There was no history of ingestion of herbal concoctions or non-prescribed medication drugs by the mother before or during the pregnancy.

Examination revealed a midline protrusion at the umbilicus with an intact overlying membrane and the umbilical stump at the center of the swelling. The defect on the fascia was estimated to be about 7 centimeters in diameters (figure 1). There were no other identifiable associated gross congenital abnormalities.

The protrusion was covered with sofra-tulle (framycetin Sulphate BP 1%, Sanofil Aventis) and immediately transferred to Life International Hospital's Neonatal Intensive Care Unit (NICU) for admission and further management. At NICU, intravenous fluid 10% dextrose water at 80mls/kg over 24 hours, intravenous ceftazidine 115mg 12 hourly, intravenous metronidazole 25mg 8 hourly and intravenous gentamycin 5mg 12 hourly were commenced. Full blood count and blood urea, creatinine and electrolytes done were normal. Oxygen saturation (Spo2) was 98%.

The pediatric surgeon who was earlier invited due to antenatal ultrasound diagnosis of anterior abdominal wall defect reviewed and confirmed the diagnosis of giant omphalocele and instituted conservative management. Umbilical cord was ligated with silk suture and excess cord was excised. Daily dressing was done with dermazin cream (1% Silver Sulphadiazine U.S.P, SANDOZ) and sofra-tulle after cleaning with normal saline and was covered with gauze and crepe bandage. By the 2nd day of life, baby was commenced on breast milk substitute 5mls every 3 hours and was increased as tolerated. By the 3rd day, breastfeeding was fully initiated and breastmilk substitute was stopped. Intravenous antibiotics were converted to oral antibiotics after 48 hours.By the end of first week, evidence of

epithelisation became obvious and daily dressing continued. Vital signs and SpO_2 remained stable throughout admission. By the end of the second week, the baby was discharged from neonatal intensive care unit with a weight of 3.43kg. Daily dressing continued on outpatient basis. At about the 16th week, all the abdominal content has retracted and the defect covered by skin (figure 2). He is to be followed up for about 2 years when the abdominal cavity will be capacious enough to accommodate repair without much complication like compartment syndrome.

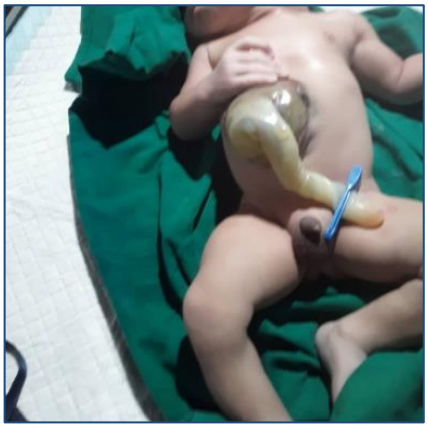

Figure 1 (Photograph of the index patient at birth)

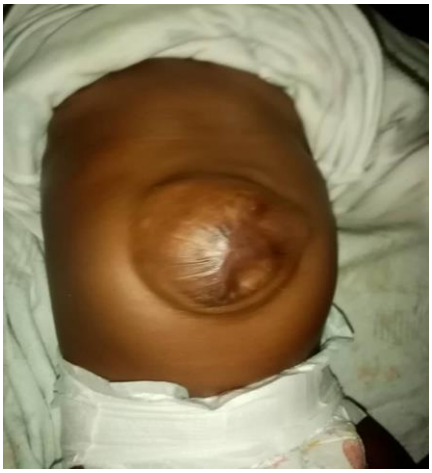

Figure 2 (Photograph of same patient at 16 weeks)

Discussion:

Omphalocele is a rare midline anterior abdominal wall birth defect, which results from failure of the midgut to return into the abdominal cavity by the 12th week of gestation [5]. It is classified as major when the defect is more than 5cm, like in the case under review in which the defect was about 7cm. Also, presence of liver or large loops of bowel classified it as major [9]. Diagnosis of omphalocele can be made during antenatal period with ultrasound, though it can be missed like in the index case reported where it was missed by some sonographers. Prenatal diagnosis will help the managing team to choose optimal mode of delivery and prepare for postnatal care. Despite the advances in neonatal surgical practice, management of giant omphalocele is usually challenging for the baby, the mother and health workers.

Management includes fluid and electrolyte maintenance, prevention of heat loss from the exposed abdominal content, prevention of sepsis, gastric distention, cardio-respiratory stability and repair of the midline abdominal defect [10]. The case under review received prophylactic antibiotics, the exposed site was dressed and covered with crepe bondage and he was closely monitored in NICU.

Immediate surgical repair of large omphalocele poses a lot of risk to the babies, which include compartment syndrome, reduced venous return from compressed inferior vena cava leading to shock and renal failure [9, 10]. To prevent the above complications, several surgical methods like the use of mesh, silos and staged surgeries with use of flaps were developed [11]. These methods may also be associated with wound infection, dehiscence, recurrent ventral hernia, intestinal obstruction and perforation [9].

Conservative management of omphalocele with daily dressing will allow for epithelisation of the wound with escar and wound contracture, this will allow for small anterior wall hernia that will be repaired later in live. Repair at later stage will reduce risks associated with early closure and will also give time for the baby to be stable enough to withstand surgery. Unlike umbilical hernia that spontaneously closes before 5 years of age, large omphalocele will require secondary closure. Oumama et al [12], reported 5 case series of giant omphalocle that were conservatively managed in Benin republic in West Africa and later were transferred to Centre Hospitalier

Universitaire Vaudois, Lausanne, Switzerland for repair at a median gestational age of 22 weeks. This shows that surgery can be delayed long enough before repair can be carried out.In this index case reported, conservative management

involved daily wound dressing with dermazin cream and sofra-tulle. Both have antibiotic effect that prevented secondary infection during healing. The synergistic effect of both agents might have helped in faster healing and wound contracture and subsequent reduction in the size of the defect.

Conclusion:

Although management of omphalocele is multidisciplinary, conservative management of omphalocele is challenging for Obstetricians and Gynecologist, pediatricians, pediatric surgeon, nurses and the family. It poses fear, anxiety and financial burden on the family. The key points in the management of this case include prevention of infections, temperature regulation and electrolytes maintenance. Support and encouragement of the family of affected child with omphalocele is paramount. Time should be given for the intestinal organs to gradually retract into the abdomen before surgery to avert complications that may follow immediate surgery. Also need for more training on prenatal diagnosis with ultrasound is important; this will prevent missing an important prenatal diagnosis like it was missed by some radiographers in the index case.

Conflict of interest: There was no conflict of interest

Author's contribution: All authors contributed to the report's design, discussion and conclusion

Consent for publication : Written informed consent was obtained from the patient for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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References:

[1.] Abdur-Rahman L, Abdulrasheed N, Adeniran J. Challenges and outcomes of management of anterior abdominal wall defects in a Nigerian tertiary hospital. Afr J Paediatr Surg 2011;8:159–63.

[2.] Ledbetter DJ. Gastroschisis and omphalocele. Surg Clin N Am. 2006; 86:249–60.

[3.] Stoll C, Alembik Y, Dott B, Roth MP. Omphalocele and Gastroschisis and associated malformations. Am J Med Genet A. 2008; 146 : 1280-5.

[4.] Barisic I, Clementi M, Häusler , Gjergja R, Kern J, Stoll C. Evaluation of prenatal ultrasound diagnosis of fetal abdominal wall defects by 19 European registries . Ultrasound Obstet Gynecol 2001; 18: 309 – 16.

[5.] Kurkchubasche AG; The fetus with an abdominal wall defect. Med Health R I. 2001; 84(5): 159-61.

[6.] Verla MA, Style CC, Olutoye OO. Prenatal diagnosis and management of omphalocele. Semin Pediatr Surg 2019;28:84-8.

[7.] Anyanwu LJ, Ade-Ajayi N, Rolle U. Major abdominal wall defects in low- and medium-income setting: Current status and priorities. Pediatr Surg Int 2020. doi.org/10.1007/s00383-020-04638-8.

[8.] Osifo O, Efobi A. Challenges of giant ventral hernia repair in children in an African tertiary care centre with limited resources. Hernia 2009;13:143–7.

[9.] Ekot EA, Emordi VC, Osifo DO. Does omphalocele major undergo spontaneous closure? Journal of Surgical Case Reports, 2017;8, 1–3 doi: 10.1093/jscr/rjx156

[10.] McNair C, Hawes J, Urquhart H. Caring for the newborn with an omphalocele. Neonatal Netw. 2006; 25:319–27

[11.] Tarca E, Aprodu S. Past and present in omphalocele treatment in Romania. Chirurgia (Bucur) 2014;109:507–13.

[12.] Oumama EE, Raymond B, Olivier R, Sabine VM, Anthony BR. Delayed Closure of Giant Omphaloceles in West Africa: Report of Five Cases. Eur J Pediatr Surg Rep 2017;5:4–8.