

Management of a Giant Omphalocele with Non-Cross-Linked Intact Porcine-Derived Acellular Dermal Matrix (Strattice) Combined with Vacuum Therapy

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Abstract

The management of giant omphaloceles at our department is primarily conservative. However, management can be challenging if the omphalocele is ruptured or the sac has to be removed. We report a case in which a giant omphalocele in a newborn female patient was managed by covering the abdominal defect with non-cross-linked intact porcine-derived acellular dermal matrix (Strattice reconstructive tissue matrix, LifeCell Corp., Branchburg, New Jersey, United States) sutured to the fascia combined with vacuum therapy.

Keywords

- ▶ omphalocele
- ▶ Strattice
- ▶ acellular dermal matrix

New Insights and the Importance for the Pediatric Surgeon

The management of giant omphaloceles is mostly conservative. However, management can be challenging if the omphalocele is ruptured or the sac has to be removed. We present PADM with vacuum therapy as a good alternative in the treatment of giant complicated omphaloceles.

Introduction

An otherwise healthy mother had a routine ultrasound at 20 weeks, which showed giant omphalocele with liver included in the omphalocele sac. Karyotyping of the amniotic fluid cells showed a normal female karyotype (46, XX), α -fetoprotein (AFP) in amniotic fluid was raised (38.7 mg/L; $3 \times \text{MoM} = 20.1 \text{ mg/L}$). These findings were discussed with the parents and it was decided to continue the pregnancy.

The baby was born at 38 weeks and 2 days of gestation by elective cesarean delivery. The birth weight was 3,500 g and Apgar scores were 8/9 at 1 and 5 minutes, respectively.

The omphalocele contained a big part of the liver, almost the whole bowel, and the spleen. The baby had no further anomalies or evident dysmorphisms. Immediately after birth, the omphalocele was covered with a sterile petrolatum

(Vaseline, Smith & Nephew, Kruisweg, Hoofddorp, Netherlands) gauze dressing, which was changed every other day. The baby expelled meconium shortly after birth and also upon arrival in the neonatal intensive care unit. Feeding was started via nasogastric tube and was well tolerated during the first days of life. On the 6th day after birth, the baby developed a bowel obstruction and underwent corrective surgery. During the operation, we resected the omphalocele sac (which was vulnerable by manipulation) and performed adhesiolysis to appropriately position the spleen and bowel in the abdominal cavity. The liver remained outside the abdominal cavity.

A Silo bag (SB06, 60-mm; Medicina Ltd., Bolton, United Kingdom) was used to temporarily cover the defect and central venous access was obtained in the left subclavian vein (**▶ Supplementary Fig. S1** in the online version of the

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Fig. 1 Completed suturing of the fascia to the porcine-derived acellular dermal matrix (14th day after birth).

article). The biggest Silo bag available (60-mm) was not big enough to accommodate the liver and was therefore enlarged by attaching a triangular patch made of silastic mesh. Because the use of a Silo is not optimal for treatment when the liver is situated outside the abdominal cavity, alternative treatment approaches had to be considered. On the 14th day after birth, we removed the Silo and placed non-cross-linked intact porcine-derived acellular dermal matrix (PADM; Strattice reconstructive tissue matrix, LifeCell Corp., Branchburg, New Jersey, United States) at the fascia and sutured it with Prolene 3-0 sutures (Ethicon, Somerville, New Jersey, United States) (→ Fig. 1). A vacuum dressing system was used to cover the PADM with a negative pressure of -75 mm Hg (→ Supplementary Fig. S2 in the online version of the article). The vacuum system was changed in the operation theater on postoperative days 3 (→ Supplementary Fig. S3 in the online version of the article), 9, and 12 and on the pediatric ward thereafter. Amoxicillin/potassium clavulanate and gentamicin were initiated for prophylaxis against infection and then changed to meropenem. Wound cultures were obtained periodically and remained negative. The PADM showed no signs of infection during this time (→ Supplementary Fig. S4 in the online version of the article).

On the 26th day of life, the patient developed sepsis from the central venous catheter infection. Central venous system blood cultures were positive and the patient experienced nephritis and renal insufficiency that subsided spontaneously. We started minimal enteral feeding 2 days after implantation of the PADM. The feeding was well tolerated and defecation was facilitated by daily rectal washout. Enteral feeding was suspended during the sepsis; however, the patient was tolerating full enteral nutrition by 45 days post-natal age.

On the 41st day of life the result of the postnatal array-CGH (Agilent Technologies, Santa Clara, California, United States) was returned, it showed a maternally inherited 780 kb deletion on chromosome band 3q28 containing part of the *TP63* gene, and four more genes that cause autosomal recessive syndromes.



Fig. 2 Cosmetic result after 6 months of the abdominal wall reconstruction.

The patient was discharged home at age 2 months and 10 days with the vacuum therapy, which was discontinued 1 week later. After removing the vacuum therapy, the patient was bathed daily by her parents and had a sterile petrolatum gauze dressing in place until the wound closed completely (→ Supplementary Fig. S5 in the online version of the article).

Ventral hernia repair was performed when the patient was 2 years and 3 months old (→ Supplementary Fig. S6 in the online version of the article). The PADM had become integrated into the surrounding fascia and was used to close the abdominal wall (→ Supplementary Fig. S7 in the online version of the article). A sample of this PADM taken during ventral hernia repair was sent for histological examination.

The patient did well postoperatively (→ Supplementary Fig. S8 in the online version of the article) and experienced no complications through 6 months of follow-up (→ Fig. 2).

Discussion

Omphaloceles are often associated with other malformations and chromosomal anomalies.^{1,2} The small chromosome 3 deletion in this case (1) is to our best knowledge (based on queries in PubMed database and Decipher database) not known to be associated with omphalocele, (2) contains no genes explicitly associated with omphalocele,³ and (3) was inherited from a healthy mother.⁴ Therefore, this deletion was considered to be most likely of neutral effect. The raised amniotic fluid AFP was associated with the omphalocele.⁵

Giant omphaloceles are not common, which makes it difficult for a single pediatric surgery department to gain extensive treatment experience. Furthermore, most of the very large omphaloceles can be treated conservatively with sterile petrolatum dressing until the epithelialization of the defect results in a large ventral hernia that can be subsequently closed when there is adequate space in the abdominal cavity. In cases where the omphalocele sac has to be removed

or when a rupture occurs, the pediatric surgeon can face surgical challenges.

The use of a combination of a biological matrix graft and vacuum therapy for treating abdominal wall defects has been previously reported in the medical literature⁶⁻⁸; however, the use of PADM for this purpose has not yet been described in our knowledge.

Strattice is a non-cross-linked porcine, acellular dermal matrix for use in xenograft transplantation, serving as a biological scaffold to support tissue regeneration.^{9,10} It is derived from porcine dermis and processed in a manner to remove the cells and porcine antigens which would provoke an immune response in humans, while retaining all its extracellular matrix (ECM) components, its three-dimensional structure and its biomechanical strength. As such when implanted, it acts as a scaffold to support tissue ingrowth. Its ECM components attract in cells and vascularity from the patients adjacent tissue, and over time it is replaced by the patients' own tissue with all components of the porcine matrix removed, in a regenerative tissue healing process.^{11,12}

In this patient, the use of PADM was successful and showed good long-term biological compatibility, as the histological examination of the collagen showed no difference with the human collagen 2 years after suturing the patch to the fascia (→ **Supplementary Fig. S9** in the online version of the article). The microscopic examination showed dissections of tissue fragments, irregular collagen bundles containing numerous small vessels, and signs of very mild chronic infection. Based on the histology examination, it was not possible to determine if the sample consisted of porcine collagen tissue or human collagen, even after applying Sirius red and Verhoeff-Van Gieson staining.

The PADM became integrated with the fascia and was strong enough to be partially used to strengthen closure of the abdominal wall defect.

Conclusion

PADM with vacuum therapy is a good alternative in the treatment of giant omphaloceles.

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