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CASE REPORT

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Repair of a giant omphalocele by a modified technique

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Abstract Large omphaloceles that contain centrally herniated liver pose challenges to surgical closure, the most significant being the space limitation of the abdominal cavity. In addition, the "pedicled" nature of the liver on the inferior vena cava creates a predisposition to acute hepatic vascular outflow obstruction as the liver is reduced into the abdominal cavity. In such cases, the alternatives include conservative treatment or staged silo reduction. The worst complication of silastic silo (SS) placement is tension and infection of the fascia with disruption of the suture line. Once infection or premature disruption occurs, closure of the defect is difficult or impossible.

This case report details a different management technique for a newborn with a giant omphalocele and presents an interesting variation of the usual SS technique that may be helpful in the management of some cases, especially in an emergency. The thick silk sutures applied in the present case absorbed the tension and the silastic sheet prevented the risks of infection and adhesions.

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Introduction

Failure of the lateral ventral folds to close by the 3rd month of embryonic life results in an omphalocele, the most common congenital abdominal wall defect (AWD). Its reported incidence is 1 in 4,000 to 5,000 live births. Moore [1] suggested classification into types 1, 2, and 3 with defect diameters of less than 2.5 cm, 2.5 to 5 cm, and over 5 cm, respectively. With larger defects, the liver is a median organ and lies within the sac. With small defects, only bowel or stomach is found outside the abdominal cavity. For larger defects, the degree of viscero-abdominal disproportion (large viscera with a small, underdeveloped peritoneal cavity) often makes it difficult to reduce the viscera in one stage without causing hemodynamic or respiratory compromise.

Successful management of infants with this defect is very challenging. Surgical integration in such cases is only possible provided technical devices are employed in progressive stages. We present a case of a giant omphalocele initially managed with standard operative silo reduction, but because of tearing of the silo, our modification was carried out to close the defect successfully.

Case report

In a 29-year-old multigravida undergoing routine antenatal care at 36 weeks of gestation, the fetus was sonographically diagnosed as having a giant omphalocele containing liver, stomach, and intestine. The mother was referred for further management. An elective cesarean section was performed. Examination revealed a giant omphalocele of 9.7×9.1 cm with intact membranes. The umbilical cord was attached to the right lower part of the sac. There was no other associated congenital anomaly. The baby was managed in the neonatal intensive care unit, optimally stabilized, and on the 3rd day of life the sac was excised, revealing contents including a central liver, a portion of stomach, and the entire gut (Fig. 1). The cecum and appendix were lying in the left iliac fossa. In addition, there was a small, broad-based Meckel's diverticulum. The abdominal wall was stretched digitally to allow the return of as much of the herniated viscera as possible. During attempted reduction of the liver, the patient developed bradycardia and hepatic venous congestion. Transient episodes of decreased oxygen saturation (SPO_2) during reduction of the liver improved on reversing the procedure. At this stage, it was decided that the abdominal cavity would not be able to accommodate the reduced contents of the omphalocele and a staged procedure was undertaken.

An opened-out tissue expander was used as a silastic silo (SS) and sutured around the full thickness of the margins of the defect with a continuous prolene suture. The silo was suspended from its apex. At regular intervals of 24 to 36 h, it was reduced in size by twisting the apex and occluding it at a lower level. After three such reducing maneuvers it was noticed that a part of the SS had torn and had detached from the margin of the defect. The patient was therefore reoperated for reapplication of the silo. At this stage, visceral integration was possible up to one-third of the volume

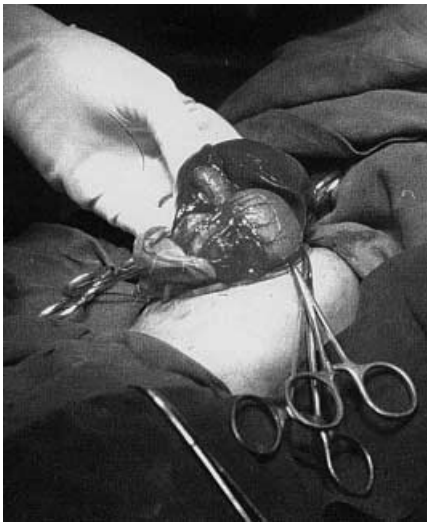

Fig. 1 Pedicled liver with gallbladder, stomach, and coils of intestine in omphalocele sac

of the omphalocele. The entire liver was still lying outside the abdomen. The fibrinous adhesions and pseudomembrane covering the viscera were gently separated. The intestines were deflated by continuous nasogastric suction and digital milking of the contents distally. The liver was partially repositioned in the right paracolic gutter and the intestine toward the left side of the abdomen. This time, the SS was applied to cover the defect as an onlay patch and sutured around the margins of the fascial defect. Additional black silk sutures with long tails were applied to the full thickness of the AWD. The tails of the corresponding sutures on opposite sides of the defect were tied to each other (Fig. 2).

At regular intervals these sutures were tightened to bring the margins of the defect closer. With this maneuver, the silastic sheet under the sutures was folded effectively inside along with its contents, preventing any leakage, adhesions, or infection. On the 14th day of life, the SS was removed and definitive closure of the defect was possible in two layers. Postoperative recovery was uneventful. There was no need to mechanically ventilate the infant at any time, however, parenteral nutrition was given until definitive closure of the defect was carried out. The abdominal wound healed well.

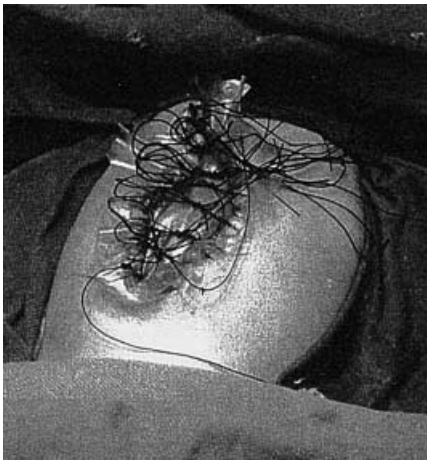

Fig. 2 Modified technique of closure of large omphalocele: silastic sheet used as onlay patch, silk sutures applied to full thickness of abdominal wall used to approximate defect

Discussion

Even in the absence of major associated anomalies treatment of giant omphaloceles is difficult, primarily because of the disproportion between the large volume of the omphalocele and the small volume of the intra-abdominal cavity. Advances in surgical techniques, neonatal intensive care, and ventilatory support have made primary fascial closure a superior approach without jeopardizing the chance for survival, but in a number of patients this is not possible. In these cases, a staged approach is mandatory to achieve uncomplicated reduction without causing hemodynamic or respiratory compromise.

The standard treatment of large hepato-omphaloceles has been SS placement followed by delayed primary closure. Still, in many centers nonoperative management using painting is considered the most reliable therapeutic approach for such cases [2]. The aim of surgical management is to provide complete fascial and skin closure without causing further injury due to excessive intra-abdominal pressure or abdominal-wall tension. DeLuca et al. [3] have suggested application of external compression by Ace bandage and Velcro abdominal binder to reduce the omphalocele for several days before reoperation is undertaken. Similarly, Brown and Wright [4] also recommended delayed external compression reduction as a safe and effective alternative to SS placement in giant omphaloceles. Klein et al. [5] reported the use of dura mater to close the AWD. Pampaloni et al. [6] observed a successful primary repair using a Gore-Tex patch. Hong et al. [7] suggested sequential reduction of the sac to reduce the contents of the omphalocele into the peritoneal cavity. Bax et al. [8] used the intra-abdominal tissue expander to enlarge the abdominal cavity for successful closure of the defect. Verlende and Zoltie [9] also used tissue expanders, but they were placed subcutaneously for later reconstruction.

The survival of omphalocele patients depends on the size of the defect, degree of viscero-abdominal disproportion, and the presence of associated anomalies. In our case, the defect measured approximately 10 cm and was the sole congenital anomaly. When the liver and small bowel have lost the right of domain to the abdomen, reduction of these structures is associated with angulation of the hepatic vein, compression of the inferior vena cava, ischemia of the intestine and liver, and respiratory embarrassment. In our case, reduction of the liver led to acute congestion associated with a severe reduction in cardiac output, bradycardia, and a fall in SPO_2.

There are a number of techniques available to close giant omphaloceles, depending on the availability of prosthetic material, facilities, and the experience of the surgeon. An innovation on the operating table to close the defect without causing respiratory, metabolic, or hemodynamic compromise should be the goal in each case. In the absence of facilities for measurement of intra-abdominal pressure or Doppler measurement of visceral blood flow to monitor the progress, reliance on the clinical condition with particular attention to the physiological status of the patient is a good indicator, as in our case. When primary or staged repair is not possible for any reason, nonoperative initial management may be the only alternative.

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