

Importance of the Early Management of Omphalocele Minor

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Abstract

Background: This study was conducted to characterize the clinical features, associated anomalies, operative management, and operative findings in babies with omphalocele minor.

Material and Methods: This was a prospective study conducted over a period of 2 years, from 2011 to 2013. The clinical, radiological, and operative findings of all the babies diagnosed with omphalocele minor were recorded and analyzed.

Results: A total of 12 patients of omphalocele minor were admitted to our department. Ten babies required immediate exploration within first 5 days of life. The main indication of exploration was mechanical intestinal obstruction due to either associated atresia (five patients) or obstruction caused by mechanical compression of the gut by the small abdominal wall defect in remaining seven patients. Atretic segment was seen within the sac in two patients, distal ileal atresia in one, atresia at the neck of the abdominal wall defect in one, and ascending and transverse colonic atresia in one patient. Out of 10 patients in whom exploration was needed, 9 required resection anastomosis and in 1 patient intestine within the omphalocele sac had pregangrenous changes and did not require resection. There were three mortalities in our series; rest of the patients were doing well during the follow up.

Conclusion: Omphalocele minor although apparently seems to be a minor anomaly, but, as shown in our series, it has the potential of causing intestinal obstruction due to associated gut atresia or compression from tight abdominal defect. We conclude and recommend that all the patients with omphalocele minor should be admitted and observed for the possible development of intestinal obstruction and hence for early surgical intervention to prevent subsequent complications.

Keywords: Omphalocele minor; Intestinal atresia; Obstruction

Introduction

Omphalocele is a central defect of umbilical ring, resulting in persistent herniation of the abdominal contents [1]. The midgut, positioned in umbilical cord, rotates 90° counterclockwise around the superior mesenteric artery [2]. At 10 weeks of gestation, intestines return to the abdomen. The small intestines return first, followed by large intestines, which complete an additional 180° counterclockwise rotation. After the intestines return to the abdomen, they enlarge, lengthen, and fuse to the abdominal cavity. The abdominal wall then closes and the body stalk constricts to become umbilical cord [3]. Omphalocele results due to either the developmental arrest of the abdominal wall or failure of the abdominal viscera to return to the abdominal cavity [4]. Omphaloceles vary in size from 4 to 12 cm. Omphalocele major has a defect >5 cm and minor has a defect 5 cm in diameter [5]. Associated anomalies are reported in 30%–80% of cases of omphalocele among which cardiac defects are more common [6]. Chromosomal anomalies occur in 49% of cases of omphalocele [7]. Interestingly, multiple associated anomalies appear to be more common in omphalocele minor than omphalocele major (55% vs 36%) [8].

Our study focuses on the clinical features and radiological and operative characteristics of omphalocele minor. We discuss the association between the size of abdominal wall defect and formation of intestinal atresia, and etiopathogenesis of intestinal atresia and other gastrointestinal anomalies in omphalocele minor. We present a series of 10 patients who required early surgical intervention for mechanical intestinal obstruction, which was due to either the associated atresia or the mechanical compression by the abdominal wall defect.

Material and Methods

This study was a prospective and descriptive study conducted

between 2011 and 2013. All the babies with abdominal wall defect 5 cm reported during the study period were included. The data regarding the clinical features, associated anomalies, radiological findings or laboratory testing and operative details were recorded and analyzed statistically. The size of the defect was determined either by reported physical examination at initial assessment or by documentation of intraoperative findings. Patients were treated surgically, received standard perioperative and postoperative care including mechanical ventilation and TPN, if necessary, or nonoperatively with dry sterile dressing until fully epithelized in case obstruction was not present.

Results

During the study period, 17 cases of omphalocele were admitted. Out these, 12 babies that were included in this study had omphalocele minor. In seven babies, omphalocele was reducible, and in five babies it was irreducible at the first instance. Five patients, out of seven with reducible contents, developed intestinal obstruction. Four babies were preterm and rest eight babies were full term. Mean weight at presentation was 2.9 ± 1.3 kg (range 1.8–3.9 kg). Mean day of life at presentation was 39 ± 49 hours with a range of 3 hours to 5 days. All the babies had intact sac. Only two babies needed ventilatory and

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cardiovascular support for respiratory distress and congenital heart disease. All the babies were admitted to the hospital, and among 12 babies 8 babies developed symptoms of intestinal obstruction such as bilious vomiting, failure to pass meconium, and abdominal distension during the first admission. Most commonly associated anomalies observed were intestinal (in five patients), renal (in two patients), cardiac (in one patient), chromosomal (in one patient), cleft lip (in two patients), undescended testis (in one patient), and hydronephrosis (in 1 patient).

The size of the defect ranged from 3 to 5 cm in diameter. Size of the defect did not corresponded with the incidence of obstruction or associated gut atresia. Four patients were discharged after proper counseling and educating parents about the any signs and symptoms of intestinal obstruction. Two babies reported back after 2 days with features of intestinal obstruction. All 10 babies had complete intestinal obstruction on X-ray with no gas in pelvis. Lateral X-ray also revealed air–fluid level in the omphalocele sac in the patients. All the babies were operated after stabilization. Upon exploration, two babies had atretic segment inside the sac and in one baby jejunum was ending into the omphalocele sac with distal ileal atresia (Figure 1). In another patient, atretic segment was seen at the neck of abdominal wall defect. Colonic atresia involving ascending and transverse colon was seen one baby. In four patients, obstruction had resulted complete gangrene of the segment within the omphalocele sac. Pregangrenous changes in gut were seen in one patient, which regained color on release of the tight neck of omphalocele. In six patients, resection and ileoileal anastomosis was performed. In two patients, jejunoleal anastomosis was carried out. One patient needed ileo-ocolic anastomosis, who later succumbed to short gut syndrome. Associated malrotation was observed in five patients. All the patients were explored through umbilicus, and after the end of definitive procedure sheath was closed with 3–0 vicryl suture and umbilicoplasty was performed in all the patients with good cosmetic results. Postoperatively, three babies needed ventilation and one baby succumbed to ventilator-associated pneumonia. One baby developed anastomotic leak, which was explored and anastomosis refashioned but postoperatively this baby succumbed to sepsis. Average hospital stay was 12 ± 19 days (range 7–28 days). In four babies with intestinal atresia, feeds were started on seventh postoperative day. All the seven babies who survived are doing well in follow-up with good cosmetic appearance of umbilicus (Table 1).

Discussion

Omphalocele occurs either due to the failure of anterior wall closure or because of ventral extension of body wall or body stalk.



Figure 1: Cross table X-Ray; gut loop trapped in omphalocele sac with multiple air fluid levels.

Omphalocele defects vary in size, with 5 cm diameter in minor to >5 cm in major. The size of the omphalocele and abdominal cavity influences the approach to the surgical management. Associated anomalies include congenital heart disease, chromosomal, renal genitourinary facial, skeletal, and gastrointestinal anomalies. The association of the intestinal atresia and omphalocele is rare, but our study showed that out of 12 babies G1 atresia was found in 4 (30%). In a series of 45 infants that had omphalocele 5 cm, 7 had malrotation and volvulus, 8 patients had Meckle's diverticulum, and one had ileal atresia [9]. Another case report by Salomon et al. [10] described an infant with 2 cm omphalocele on prenatal USG, which was not associated with sonographic abnormality. On surgical exploration, the infant was found to have ileal atresia. Our study shows that intestinal anomalies are distinctly associated with omphalocele minor. The etiopathogenesis of atresia associated with omphalocele minor remains elusive. Vascular occlusion has been shown to be a cause. This study shows that omphalocele minor developed intestinal obstruction in 10 of 12 cases. In five cases, intestinal atresia was the cause. We hypothesize that the intestinal atresia occurred because of mechanical compression of the intestines at the neck of omphalocele defect or because of the internal volvulus of the prolapsed segment during antenatal period. The small abdominal defect could divide the part of prolapsed gut, thereby causing the atresia. Atresia could also result in such cases because of ischemia subsequent to the interruption of blood supply at the reentry point of intestines to the abdominal cavity. In the remaining five patients, the intestinal obstruction was acquired either by the compression of the neck of the defect or by the adhesions around the omphalocele.

Upon reviewing the literature, it was found 19 cases of omphalocele with atresia have been reported [11]. In eight cases, intestinal compression by a tight defect has been described as the cause of atresia [12,13]. In three cases, authors concluded that omphalocele and intestinal atresia occurred independently of one another [14,15]. In eight cases, the origin of the atresia remained elusive [16,17]. Although these studies lack standardization regarding the measurement of the size of the defect, the literature supports that the development of intestinal atresia in omphalocele requires a small abdominal wall defect capable of intestinal and mesenteric compression. Pratap et al. [17] described a case of ileum entrapped within the omphalic ring and that entrapped segment of ileum was atretic. Patel et al. [18] have described an unusual case of ileal atresia resulting from antenatal strangulation of Meckle's diverticulum in omphalocele minor.

Out of 12 patients, 4 patients had obstructed omphalocele minor with gangrene of the entrapped ileum. The probable cause of gangrenous changes of the entrapped ileum was the mechanical compression of the gut and its mesentery by tight abdominal defect. In our study, the size of the defect did not correspond with the incidence of the obstruction or atresia.

Studies have shown that small omphalocele associated with noncardiac anomalies has overall good prognosis [19-21] (Figure 2). Patients with small omphalocele have fewer cardiac defects, which may explain their better prognosis as severe cardiac defects portend poorer prognosis. The size of the defect has also been reported to determine the outcome with mortality rate of 25% in infants with omphalocele major [22]. Prognosis is described favorable for infants with isolated small omphalocele and no associated structural and chromosomal anomalies [23]. This is consistent with our observation where three mortalities were due to the intestinal obstruction or its postoperative sequel and *per se* mortality was not related to the underlying associated cardiac or chromosomal anomalies. Furthermore, as described earlier, absence of cardiac defects is related to better prognosis, which is similar to that observed in our study (Figure 3).

Patient NO.	Sex / weight kgs.	Day of life	Presentation at first instance	Size of omphalocele(cms)	Associat-ed anomaly	Surgical exploration done at ---- D.O.L	Anomaly found on Surgical exploration	Operative proceedure done	Post-operative Period	Average hospital stay (Days)
1	Male/2.6 kgs	3 hrs	Reducible O.M Abdominal distention	3 cm	Radial limb defect	3th	Ileal atresia / Malrotation	Resection – Ileo-ileal Anastomosis and umblicoplasty	Uneventfull	8
2	Female/2.8 kgs	18 hrs	Irreducible O.M bilious vomiting	3.5 cm	ASD	3th	Colonic atresia	Resection – Jejunocolic anastomosis + Umblicoplasty	SBO---DIED	34
3	Male/2.2 kgs	20 hrs	Irreducible O.M	2.8 cm	-----	3th	Gangrenous entrapped ileal segment	Resection – Ileo –ileal Anastomosis with Umblicoplasty	uneventful	7
4	Male/3.5 kgs	24 hrs	Reducible O.M was discharged	3 cm	--	Delayed	----	Reduction of contents and umblicoplasty	Uneventful	9
5	Male/4 kgs	24 hrs	Irreducible O.M With bilious vomiting	3.4 cm	Renal – hydronephrosis	2 nd	Gangrenous entrapped ileal loops with malrotation	Resection – Ileo-ileal Anastomosis and umblicoplasty	Ventilated--VAP and died	30
6	Female/2.5 kgs	48 hrs	Reducible O.M was discharged	4 cm	Trisomy 13	Readmitted on 5 th D.O.L with intestinal obstruction	Gangrenous ileal loops	Resection – Ileo-ileal Anastomosis and umblicoplasty	uneventful	12
7	Male/3 kgs	36 hrs	Reducible O.M/failure to pass meconium	3.7 cm	-----	5 th	Ileal atresia with malrotation	Resection + jejunocolic Anastomosis + umblicoplasty	Anastomotic leak, sepsis, ventilated – death	27
8	Female/3.7 kgs	24 hrs	Irreducible O.M	3 cm	UDT	3th	Ileal atresia/ Malrotation	Resection – Ileo – colic Anastomosis+ umblicoplasty	Uneventful	7
9	Male/3.8 kgs	40 hrs	Reducible O.M – discharged home	4 cm	Cleft lip	5 th	Gangrenous entrapped segment of small intestines	Resection – Jejun-ileal Anastomosis + umblicoplasty	Put on Mechanical ventilator--recovered	7
10	Male /2.4 kgs	60 hrs	Reducible O.M	4.5 cm	Distal Hypospadias	4 th	Pregangrenous changes in entrapped gut	Reduction of contents and umblicoplasty	Uneventful	6
11	Male/3.0 kgs	25 hr	Irreducible O.M	3.9 cm	-----	3th	Volvulus of entrapped ileum with malrotation	Resection +ileo-ileal Anastomosis + umblicoplasty	Uneventful	9
12	Male/3 kgs	30 hrs	Reducible	2.8 cm	Cleft lip	Delayed	-----	Reduction of contents + umblicoplasty	Uneventful	5

OM—omphalocele minor, VAP—ventilator associated pneumonia, UDT—undescended testis, ASD—Atrial septal defect, MV—Mechanical ventilation

Table 1: Clinical, operative findings, associated anomalies in omphalocele minor.

Morbidity in omphalocele is also determined by the presence of associated anomalies and the size of the abdominal wall defect [24]. But in our study, morbidity was also related to the local gut problem within the omphalocele sac and by the compression of abdominal wall defect. Our observations are of relevance to the clinicians. The approach used to screen for concurrent anomalies may miss the gastrointestinal tract anomalies present within the omphalocele minor. These simple and minor looking omphalocele are associated with their own distinct variety of anomalies, among which intestinal atresia are common (Figure 4). As shown, morbidity and mortality in omphalocele minor are not due to cardiac defects and need of urgent laparotomy and the rates of morbidity and mortality are related to the intestinal anomalies either associated with the omphalocele minor or caused by the compression by abdominal defect. The surgeon must be aware of the potential undetected intestinal anomalies at the time of exploration and the chances of subsequent obstruction and strangulation of the gut by the abdominal wall defect. We also believe that reducibility of the contents at the first instance is not a sign that the baby will not develop the intestinal obstruction subsequently, as five out of seven babies with



Figure 2: Photograph showing ileal atresia with in the omphalocele sac.



Figure 3: Gut loops entering the omphalocele sac with color changes.



Figure 4: Pregangrenous changes in loop of gut in omphalocele sac.



Figure 5: Umblicoplasty completed.

reducible omphalocele minor needed early exploration (Figure 5).

Conclusion

We believe that omphalocele minor cases should be observed for development of any signs of intestinal obstruction because morbidity and mortality rates can be high in cases where diagnosis of the intestinal obstruction is delayed. Furthermore, the possibility of intestinal atresia with omphalocele minor cases needs close observation. We recommend that all the babies with omphalocele minor should be

admitted irrespective of size and reducibility, and be screened for associated intestinal and other anomalies. All the omphalocele minor cases should be operated early before discharge from nursery or at the time of referral from another hospital.

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