

Case Report

Ompholoele in Adult Female: A Rare Case Report and a Review

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Abstract

Ompholoele is defined as congenital abdominal wall defect with herniated abdominal viscera due to weakness in abdominal wall. The incidence of ompholoele is more common in males compared to females. The survival rate depends on severity of complication. During routine dissection of formalin fixed female cadaver in Department of Anatomy, SDUMC, observed a mass in right, lower quadrant of lumbar region with mesentery, blood vessels bulged out through right side opening. The liver, cecum and appendix were in midline but not in anatomical positions in cadaver. This condition was named as isolated ompholoele. Isolated Ompholoele in female cadaver is a rare case report.

Keywords: Abdominal hernia, Giant Ompholoele, Abdominal wall defects, Gut rotation, Exomphalos.

Introduction

Ompholoele is also named as exomphalos. Ompholoele is defined as anterior abdominal wall defect with herniated viscera along with peritoneum. If it is in fetal life Wharton's jelly, amnion will also involve in the covering. ¹ In complicated cases like chromosomal aberrations, multiple organs rotation failure then survival itself is difficult. Only in isolated ompholoeles life span can expect till 90%. ² The incidence of ompholoele around the world ranged from 1 in every 5000 fetuses in births, 1-3 in 10000 live births. ³ The occurrence of ompholoele is more common in males compared to females by 5:1 ratio. ⁴

Classification of ompholoele: Firstly the anterior abdominal wall defects were classified as:

Based on location of defect and umbilical cord insertion:

- Epigastric – not reaching below the umbilicus –cranial
- Hypogastric - not reaching above the umbilicus- central, caudal
- Periumbilical- Near and around the umbilicus-complex ⁵

Based on the size:

- < 4 cms considered as small
- > 4 cms considered as giant ompholoele.

Giant ompholoele includes majority with liver defective and cardiac anomalies. ^{6,7}. Small size were complicated with gut

malformations. ⁷

Based on the associated anomalies:

- Isolated type
- Non-isolated type

Infants with isolated omphaloele may have minor intestinal problems usually recover from that and lead a completely normal life. ⁸ The frequency of isolated ompholoele is 1 in 5000 live births. ⁴

Case History:

During routine dissection of formalin fixed female cadaver of aged 80 years in Department of Anatomy, SDUMC, there was a mass present in right, lower quadrant of lumbar region. On further dissection of the anterior abdominal wall we found the abdominal contents (small intestines) bulged out along with mesentery through an opening on the right side. This opening is oval in shape with following measurements: Vertically, horizontally and obliquely was 3X4X4cms in figure 1. The opening is not strangulated as the intestinal contents were flexible through opening. The opening is covered by skin and deficient by abdominal wall muscles. The location of viscera was as follows:

Liver: shows incomplete rotation in figure 2. Spleen, stomach, pancreas were in normal position.

Intestines - small and large intestines, small intestine were protruded outside along with mesentery and mesenteric vessels. Whereas large intestine remaining portion was normal except for the caecum and appendix were in midline instead of in right iliac fossa. So this is due to midgut rotation failure.

Kidneys: The left kidney was larger than normal size with irregular shape. The ascent of the left kidney was incomplete and right side kidney was completely absent in figure 3. **Ureter:** The left Ureter is normal in its course and right side ureter abruptly ended in lateral pelvic wall in figure 4.

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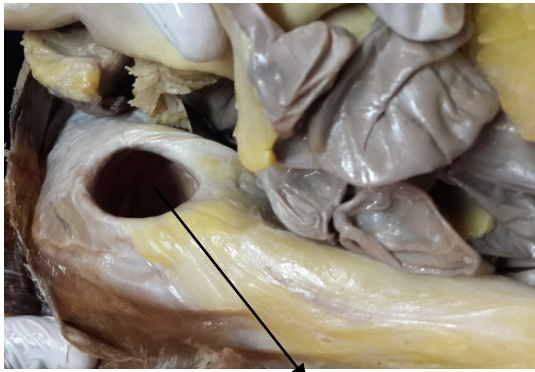
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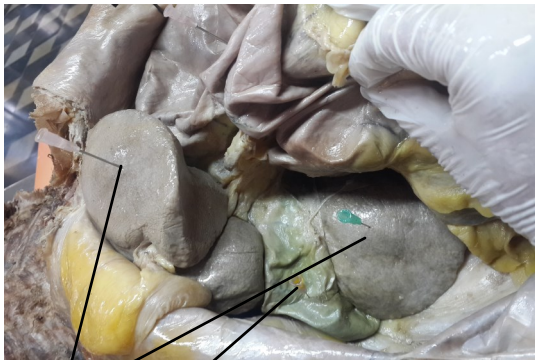
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Fig.1: Measurements of opening



Vertically, Horizontally and Obliquely: 3X4X4cms

Fig.2: Incomplete Rotation of liver



Liver with incomplete rotation lobes are not divided
Gall Bladder

Fig.3: Left Kidney with incomplete ascent



Left kidney was larger than normal size with irregular shape and incomplete ascent.

Fig. 4 : Right side Ureter incomplete



Right side ureter abruptly ended in lateral pelvic wall

Discussion

Omphalocele derived from Greek word meaning 'outside the navel'.¹⁰ Poddar et al., in his study mentioned that in females omphalocele occurrence is rare.¹¹ The survival rate is close to 80%, and is related to severity of the associated anomalies. In 50%-70% of cases omphalocele is associated with anomalies.¹ In isolated omphalocele have survival rate of 90%.² According to Chen and Hwang et al., omphaloceles are accompanied with chromosomal aberrations, abnormalities and other birth defects.^{12,13} The exact etiology of omphalocele is still imprecise. In 6th week, due to improper development there was disorder in organogenesis so the gut contents fail to rotate and return to the abdominal wall cavity.⁹ The other fact is that the midline anterior abdominal wall develops from four ectomesodermal derived folds which fuse at 3rd to 4th weeks of embryonic development. If this abdominal wall formation fails to fuse results in omphalocele.¹⁴ Due to absence of midline anterior abdominal wall the abdominal contents will herniate.¹⁵ The abdominal contents like intestines with or without peritoneum will herniate and other organs like liver fails to rotate and it is the indication for large abdominal wall defects.^{1,2} In this present case, it is giant, isolated omphalocele in adult female cadaver of 80 years with right side kidney & ureter agenesis, with incomplete midgut rotation is a rare case report.

Liang et al., in his study mentioned that prenatal diagnosis help us to some extent to reduce fetal morbidity and mortality rate.⁴ Brantberg et al., in his study mentioned that omphalocele fetus with liver protruding, are not evident by karyotypes.⁵ Different studies piloted by different authors on omphalocele has been tabulated in table 1. According to Das that isolated omphalocele, will survive and can lead normal life.⁸ By Lamquami and Mack et al., in his study mentioned about that in gaint omphalocele involves liver in herniation.^{6,16} Rattan et al., commented on size of omphalocele, as it assists clinician.⁷ Present case study matches with the study piloted by following authors Brantberg, Das, Lamquami, Mack, Poddar, Rattan et al.,^{5,8,6,16,11,7}

Table 1: Representing different study opinions on omphalocele

S.No	Study Details	Opinions on omphalocele
1	Rattan 2018 Original Article, Haryana, India. ^[7]	Small sized omphalocele (< 5cms) related with gut malformations, and larger sizes (>5cms) related with cardiac disease.
2	Watanabe 2017, Original Article, Japan. ^[14]	Failure of the 4 mesodermal-derived folds is one of the reason.
3	Mack 2016, Review, Switzerland. ^[16]	Giant omphalocele herniated with intestinal organs, including liver and other associated congenital anomalies.
4	Das 2015, Case Report, Chennai, India. ^[8]	Isolated omphalocele have slight Gastro intestinal problems and leads completely normal life. Giant omphalocele is relatively uncommon
5	Gupta 2015, Case Report, Ranchi, India. ^[15]	Omphalocele are related with chromosomal abnormalities with trisomy 18.
6	Lamquami 2015, Case Report, Morocco. ^[6]	Slighter defects (< 4 cm) are associated with gastrointestinal defects, whereas bigger defects are more related with cardiac deformity. Gaint Omphalocele liver is confined in this deformity.
7	Pakhale 2015, Case Report, Maharashtra, India. ^[9]	Associated with anomalies and become worse with genetic abnormalities.
8	Shamima 2015, Case Report, Bangladesh ^[3]	Consistent intake of Folic acid decreases the prevalence. Preconceptional therapy.
9	Sankalecha 2014, Case Report, Nashik, India. ^[10]	Omphalocele inv50% related with major or minor malformation. The most common is cardiac anomalies. Pre-conceptional multivitamin use is associated with 60% reduction in risk of non syndromic omphalocele.
10	Present Study, 2019 ,Kolar, India	The findings of the study is similar with Brantberg, Das, Lamquami, Mack, Poddar, Rattan. Occurrence in females is rare and survival chances are less. As it is isolated omphalocele survival rate was there with 1 kidney & 1 ureter.

Conclusion

Prenatal diagnosis and genetic counselling for couples may reduce fetal morbidity and mortality. Using of folic acid at recommended dosages at preconception can reduce the incidence rate.

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