CASE REPORT
Omphalocele Containing Bowel, Liver and Pancreas

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ABSTRACT

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Omphalocele is a midline defect of the anterior abdominal wall, characterised by herniation of abdominal viscera into the base of umbilical cord. The incidence is 1 in 4000 to 1 in 5000 live births. (1). We present a 31 year old female, primigravida, who reported to the hospital at 22 weeks of gestation diagnosed as omphalocele? gastrochisis and admitted for termination of pregnancy.

1. Introduction

The patient was a 31 year old primigravida who came to the hospital for the first time with 6 months of amenorrhoea and an outside USG diagnosing omphalocele? gastrochisis in the fetus.

Her last menstrual period was on 6th December 2012. The patient was a primigravida with a married life of 1 year. There was no history of any Diabetes Mellitus, Hypertension, or any prolonged illness. There was no history of any similar congenital anomaly in the family.

A repeat USG was done at our centre which showed SLUUF with GA 21 weeks+ 2 days with ? Omphalocele ? Gastrochisis with Polyhydramnios (AFI= 20). The patient was counselled for fetal echocardiography and amniocentesis which she refused. Pediatric surgeon reference was done along with the USG report where the attendants were explained the prognosis and they opted for termination of pregnancy.

At the time of admission, she had completed 21 weeks+6 days of gestation. On Examination- the vitals were stable. P/A - Fundal Height- 28 weeks, uterus relaxed, liquor seemed to be increased. P/V - cervix - anterior,soft,closed,uneffaced. Routine antenatal investigations were done. Hb=10.6 gm%, ABOrh- O+ve, RBS= 77 mg/dl. After taking informed written consent, Induction of labour was done by Prostaglandin gel followed by Misoprostol pessary. The induction- Abortion interval was 25 hours. The patient expelled a male fetus weighing 435 grams with omphalocele seen. The abdominal organs were covered by a membranous covering with umbilical cord attached to the centre.

The patient stayed in the hospital till 24 hours post partum and was discharged in satisfactory condition.

The fetal blood sample was sent for karyotyping and the fetus was sent for an autopsy. The fetal karyotype was XY. An autopsy was done which showed a trilobed liver with normal bowel, gall bladder pancreas within the sac. The covering membrane was peritoneal covering with Wharton’s jelly. Another associated anomaly was Polycystic Kidney (FIG 1).

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Omphalocoele is a group of birth effects that is related to anterior abdominal wall. It is characterised by herniation of abdominal viscera into the base of umbilical cord. Omphalocoele is a defect of the umbilical ring, which results in the failure of two lateral abdominal wall folds to migrate and fuse normally in the midline around 3rd or 4th week of intrauterine life(2).

The incidence is approximately 1 in 4000 to 1 in 5000 live births. It occurs more frequently in males than in females(1).

The etiology of omphalocele is not known. Various theories have been postulated; these include failure of the bowel to return into the abdomen by 10-12 weeks, failure of lateral mesodermal body folds to migrate centrally, and persistence of the body stalk beyond 12 weeks’ gestation.(3)

The herniated viscera are included in a sac, which is internally formed by peritoneum and externally by Wharton’s jelly and the amnion. The protruding organs are typically covered by an amnio-peritoneal membrane. Some investigators give physiopathologic significance to the presence or absence of hepatic tissue in the sac (4).

There are two generic syndromes which have been classically associated with omphalocele. They are Pentalogy of Cantrell—which includes midline supraumbilical abdominal defects, a defect of lower sternum, anomalies of diaphragmatic pericardium, and anterior diaphragm, and cardiac anomalies. The other syndrome is Beckwith- Wiedmann Syndrome, which is characterised by macrosomia, macroglossia, visceromegaly, Pancreatic hyperplasia, diaphragmatic hernia and several degrees of omphalocele (5).

Chromosomal abnormalities have been reported in as many as 43% foetuses with omphalocele, the most common being trisomies 13,18, 21 and Turner’s syndrome. An abnormal karyotype has been found to be associated to the presence of polyhydramnios or oligohydramnios and to the absence of liver in the herniated sac (5).

The anomaly is usually detected during routine ultrasonographic surveillance, during an investigation of a disparity of uterine size with time from conception or other obstetric indications, or during an evaluation of an increased maternal serum alpha-fetoprotein (AFP) level. Omphalocoeles and gastroschisis are two open, ventral wall defects that are detected by means of AFP measurement. Acetylcholinesterase levels may also be increased.(6)

Results of ultrasonographic studies suggest that in differentiating between normal physiologic herniation and a concern for omphalocele, one should note that physiologic midgut herniation should not exceed 7 mm in diameter and that physiologic herniation should not be apparent in fetuses with a crown-rump length greater than 44 mm.

Diagnostic amniocentesis is indicated when an omphalocele is suspected on antenatal sonograms. The finding of an omphalocele should prompt a targeted ultrasonographic examination to search for associated anomalies. Fetal echocardiography and karyotyping should also be performed.(7)

Omphalocele is to be differentiated from a similar anterior abdominal defect which is Gastroschisis.

Gastroschisis is a paraumbilical defect of the anterior abdominal wall usually located on the right side of the cord insertion, whereas omphalocele is a midline defect with attachment of the cord in the centre (FIG 2). There is absence of membrane covering the defect with herniated organs floating in amniotic fluid and the presence of thickened bowel loops matted together in case of Gastroschisis where as the membrane covering the omphalocele contains the peritoneum internally and Wharton’s jelly and amnion externally.

CONCLUSION
Termination of pregnancy should be discussed with the parents when the diagnosis is made before fetal viability. If the fetal karyotype is normal and no life threatening associated abnormalities are seen, expectant management is indicated.

The mode of delivery in these foetuses should not be altered by presence of defect. Delivery should be in a tertiary care centre and pediatric surgeon and neonatologist should be notified.

Several studies have demonstrated that vaginal delivery does not alter the prognosis. Cesarean section should only be performed for obstetrical reasons or in foetuses with herniated liver that may be damaged during vaginal delivery.(1)
REFERENCES


