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Case Report

Gastroschisis—A case report

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ABSTRACT

Gastroschisis is a congenital defect of the anterior abdominal wall in which the intestines and other abdominal organs, protrude from the abdomen through a small hole. In recent years rates of Gastroschisis have been increasing. Most of cases it is an isolated defect or associated with other anomalies. Ultrasonography can detect it within 14 weeks after physiological herniation. Gastroschisis and omphalocele are two defect that closely resemble each other. In this case 28 weeks still born fetus having paraumbilical defect of anterior abdominal wall through abdominal contents protrude. On autopsy there is no other gross abnormality.

1. Introduction

Gastroschisis is a full-thickness defect in the abdominal wall usually just to the right of a normal insertion of the umbilical cord into the body wall. A variable amount of intestine and parts of other abdominal organs are herniated outside the abdominal wall with no covering membrane or sac.[1] The incidence of gastroschisis ranges between 0.4 and 3 per 10,000 births but in the last decade its incidence in many countries appears to be increasing. Rates of gastroschisis are particularly high among younger mothers, including teen moms. [2] This increase may be multifactorial: low maternal age, smoking and the use of vasoactive drugs have been associated with gastroschisis. The incidence of associated anomalies is between 10% and 20%, and most of the significant anomalies are in the gastrointestinal tract.[3] It may be associated with increased obstetric complications, long durations of hospital stay and enteral feeding, repeated surgery for bowel complications and an approximate 10–15% incidence of long term developmental problems.[1]

2. Case presentation & finding

A 28 weeks intrauterine dead female fetus with gastroschisis was obtained from the Obstetrics & Gynecology Department of MIMS, Mandya after consent from parents and body donated to the anatomy Dept for autopsy.

On external examination, there is a hole in anterior abdominal wall 4 cm from right lateral margin of umbilical cord. It is not covered by a membrane. The body was fixed in 4% formalin for seventy two hours. Further during autopsy no other gross anomalies were found. No routine ultrasound had been done, the gestational age was based on the last menstrual Period (LMP) and biparietal diameter. The infant was delivered vaginally and the birth weight was below the normal range (Fig. 1).

On x ray examination, there is anteroposterior & lateral radiograph of stillborn show normal alingment and morphology of vertebra, ribs, and appendicular skeleton. Incidental notes of air pockets seen in lung bases as well as pertional cavity with overriding skull bone. No evidence of skeletal deformities or abnormal bone densities to suggest skeletal dysplasia (Fig. 2).

For histological examination of intestinal content after fixation, dehydration, clearing, block prepared then a thin section of intestinal content was prepared then stained with heamatoxyline & Eosin. On microscopic examination of slide, wall of intestine not well developed. Mucous lining of gut not continues. (Fig. 3)

3. Discussion

The abdominal wall is formed by infolding of the cranial, caudal, and two lateral embryonic folds. As the abdominal wall is formed, the rapid growth of the intestinal tract leads to its migration outside the abdominal cavity through the umbilical ring and into the umbilical cord during the sixth week of gestation. By the 10th to 12th week, the abdominal wall is well formed and the intestine returns to the abdominal cavity in a stereotypical pattern that results in normal intestinal rotation and later fixation.[1] The right paraumbilical area is an area at risk because it is supplied by the

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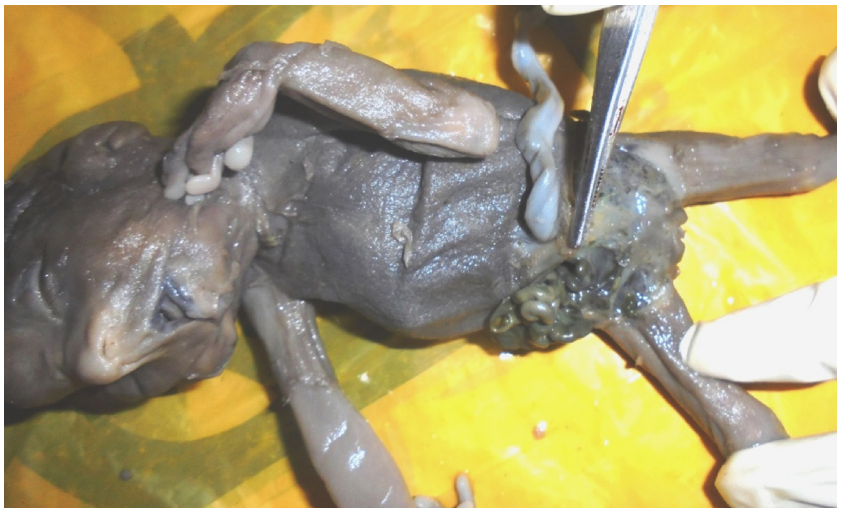

Fig. 1. Stillborn female fetus with Gastroschisis.

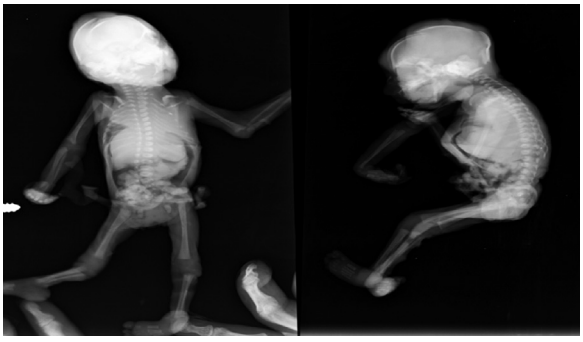

Fig. 2. X-ray of - fetus Antero posterior & Lateral view-showing overriding of skull bone & air pockets in abdomen and chest consistent with stillborn fetus. However no skeletal abnormality detected.

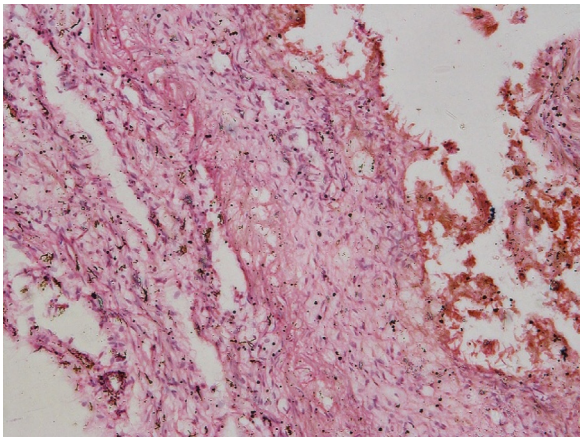

Fig. 3. Microscopic appearance of intestinal content of fetus stained with Hematoxyline & eosin stain.

right umbilical vein and right omphalomesenteric artery until they involute. If this ordered development and involution is disturbed in degree or timing, then a body wall defect could result from the resulting body wall ischemia. The damage of the herniated intestinal loops is quite variable from the normal looking intestine to extremely damaged due to prolonged contact with the amniotic fluid, rich of gastrointestinal enzymes and waste products.[7] Body mass index (BMI) and nutrient deficiencies in maternal dietary intake, low alpha-carotene, low total glutathione, and high nitrosamine intake during the trimester prior to conception have been associated with gastroschisis. This led to the hypothesis that younger age of mothers may lead to maternal fetal competition for nutrients with the result being maternal dietary inadequacy. [2]

Fetal growth failure in gastroschisis may be due to increased losses of protein from the exposed viscera, and inadequate supply of vital nutrients is an another factor. Oligohydramnios is being present in up to 25% of cases. It is usually of moderate severity and associated with IUGR, fetal distress, and birth asphyxia.[1]

About 10% of gastroschisis babies have intestinal stenosis or atresia that results from vascular insufficiency in the bowel at the time of gastroschisis development or from later volvulus or compression of the mesenteric vascular pedicle by a narrowed abdominal wall ring. Other less common associated anomalies include undescended testes, Meckel's diverticulum, and intestinal duplications.[1] The Interstitial Cells of Cajal (ICC) are responsible for rhythmic electrical activity. A paralytic ileus is present in gastroschisis (GS), a malformation due to a defective closure of the abdominal wall through which part of the intestine herniates during pregnancy Several tissues involved in normal ventral body wall closure are defective in the absence of AP-2a. It is involved in multiple developmental mechanisms directing the morphogenesis of the ventral body wall, including cell migration, differentiation, and death. There is a failure of migration and fusion of the body folds at the umbilical ring, as well as in the formation and migration of the abdominal bands and ventral musculature. Furthermore, the mechanism of cell deposition at the umbilical ring is disturbed. Consequently, the mesodermal compartment of the body wall is underdeveloped.[5]

The differential diagnosis includes omphalocele, pentalogy of Cantrell, amniotic band syndrome, and limb–body wall complex. Pentalogy of Cantrell, amniotic band syndrome, and limb–body wall complex are easily excluded on the basis of associated skeletal and vertebral anomalies. Gastroschisis and omphalocele are the two most common and important forms requiring differentiation.[6]

Gastroschisis	Omphalocele
Paramedian defect	Midline defect
Herniated contents are not covered by a membrane & located on the right side of the umbilical cord.	Herniation of membrane-covered abdominal contents into the base of the umbilical cord.

(*Continued*)

Gastroschisis	Omphalocele
Develops earlier in gestation, between weeks 4 and 8 week.	After 10 weeks
Most common complications as jejunoileal disruption and intestinal atresia	Associated with malformations & syndromes and chromosomalanomalies, particularly trisomies 13 and 18 and the Beckwith-Wiedemann syndrome, Neural tube defects, diaphragmatic hernia, and fetal heart defects

Gastroschisis is often diagnosed prenatally and can be seen on ultrasound as early as the 14th week of pregnancy. When maternal alpha-fetoprotein (AFP) levels are elevated nine times of average value, ultrasonography show loops of bowel (intestines) floating freely in the amniotic fluid.[6] Surgery for the gastroschisis is typically done as soon as the infant's condition is stabilized, usually Within in 12–24 h after delivery. More than 90% of infants born with gastroschisis survive and their long-term prognosis is excellent [2]

4. Conclusion

Gastroschisis is paraumbilical defect, in which content is not covered by a membrane. Radiograph of fetus allow us to evaluate the immature skeleton for detection of any bony deformities. Due to the prenatal diagnosis, advances within neonatal care units, and improved surgical techniques have increased the survival rates of Gastroschisis.

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