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Left Sided Gastroschisis With Limb Abnormalities

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Abstract: *Gastroschisis represents herniation of abdominal contents through a paramedian full-thickness abdominal wall fusion defect without involving the umbilical cord. Evisceration, usually, contains only intestinal loops and not covered by membrane unlike in Omphalocele. Gastroschisis with other serious birth defects is unusual. Neonates with Gastroschisis have better prognosis than those with an Omphalocele. Very rarely gastroschisis is associated with herniation of major viscus and their presence makes the prognosis worst. This is a rare case because of Left sided Gastroschisis with evisceration of major viscera and association of Left side upper limb abnormality and a duplicate limb.*

Key words: *Gastroschisis, Herniation, Left sided, limb abnormality, duplicate limb.*

I. Introduction

Omphalocele and Gastroschisis are the common forms of presentation of congenital abdominal wall defect. Omphalocele is more common, with a general incidence of 1:4,000 births. Omphalocele is a result of failure of formation and closing of the anterior abdominal wall and could therefore be associated with other forms of impaired organ formation[1,2]. Gastroschisis is a defect in the full thickness of anterior abdominal wall through which the abdominal content protrudes into the amniotic cavity. Incidence of gastroschisis is 1:10,000 births[1,2]. It typically presents to the right of the umbilicus, although the defect may occasionally be to the left of the umbilicus[3].

II. Case report

The detailed history of mother and the birth history are not available. The foetus is collected from a peripheral hospital. This is a premature female foetus aborted at the approximate gestational age of 24-28 weeks. There is herniation of gut along with major viscera. The herniation is to the left side of the umbilicus. There is herniation of stomach, small gut and large gut. The major viscera herniated are liver with gall bladder and spleen. The umbilicus is in the normal position. The right upper limb and both lower limbs are normal. The left upper limb has a constriction band in the mid arm and a short forearm and a single finger. There is non-development of wrist and palm in the left upper limb. There is a devitalized duplicate limb attached to the left lateral abdominal wall with a tag of skin which has the similar structure as that of the left upper limb. There is scoliosis towards the left side.

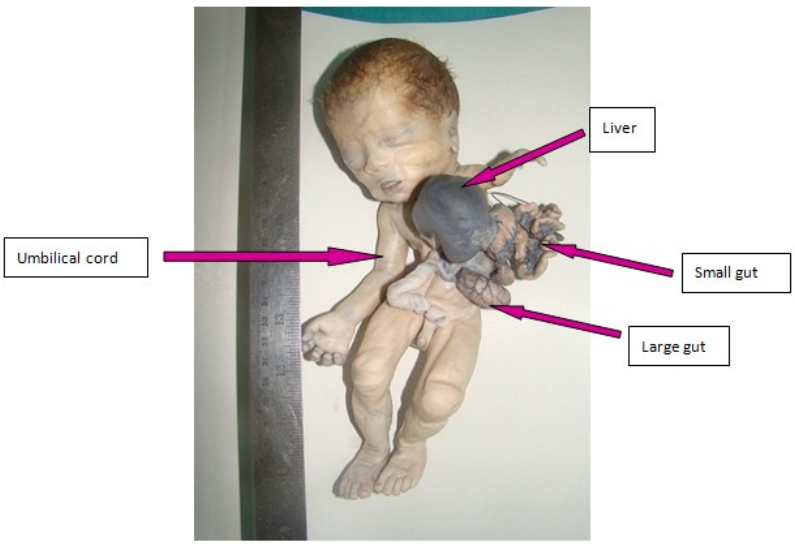

Figure 1: Showing umbilicus on the right side of the eviscerated bowels and viscera.

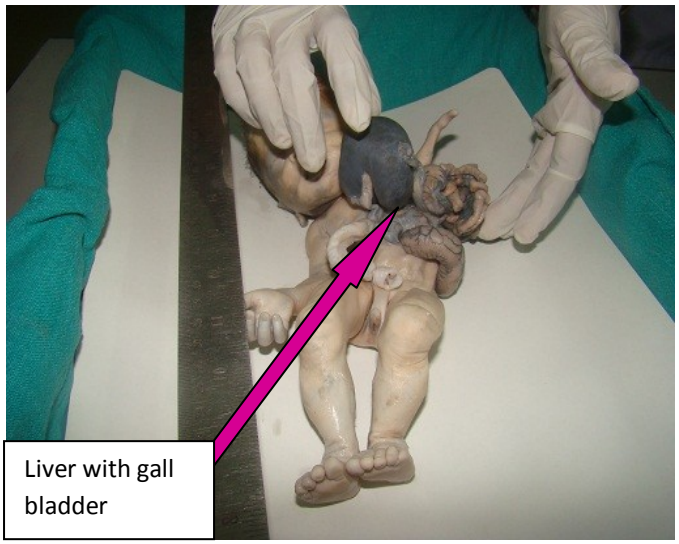

Figure 2: Showing eviscerated liver with gall bladder.

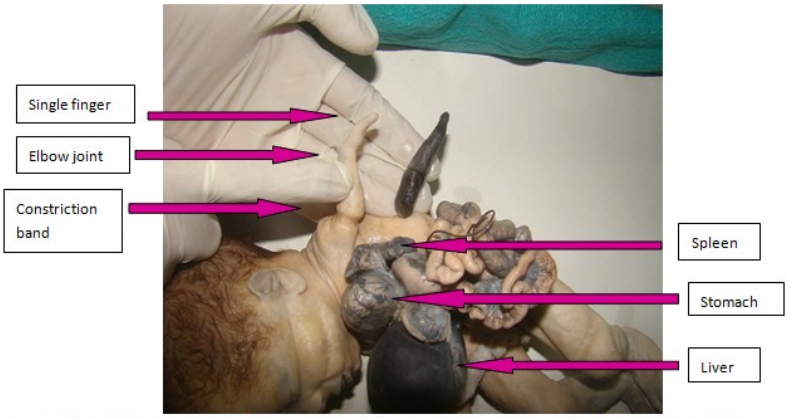

Figure 3: Showing evisceration of stomach and spleen. Abnormal left upper limb and duplicate limb attached to the lateral abdominal wall.

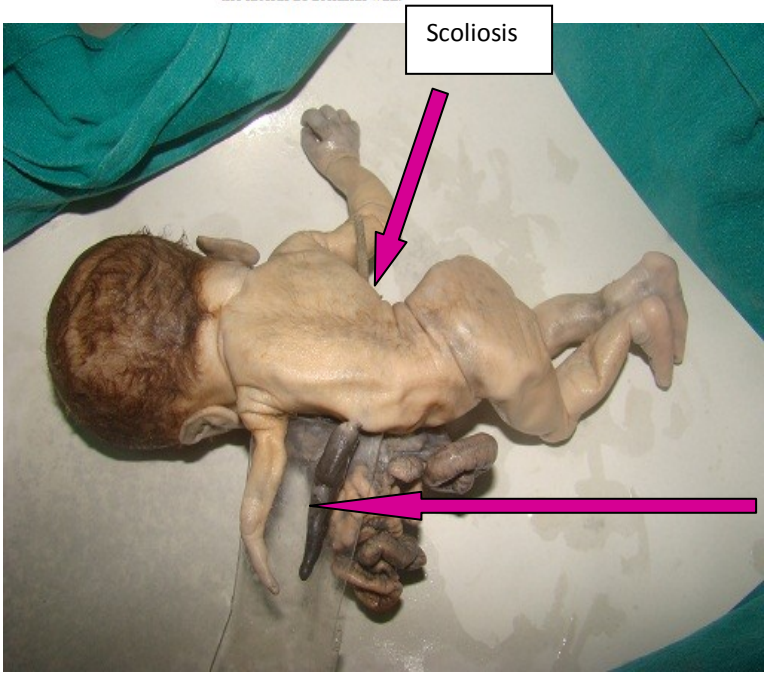

Figure 4: Showing scoliosis along with the abnormal left upper and the duplicate limb.

III. Discussion

Congenital abdominal wall defects can be defined as eviscerated organs not covered by normal skin. There are three distinct types: omphalocele, gastroschisis and umbilical cord hernia. Omphalocele and gastroschisis are by far the most severe[4].

Gastroschisis (gastro = stomach—the term generally used for abdomen; schisis = fissure, tear, or gap) is a defect in the full thickness of anterior abdominal wall through which the abdominal content protrudes into the amniotic cavity[1,2]. Until the 1950s, no clear distinction was made in most reports among omphalocele, gastroschisis and other abdominal wall defects[5]. In 1953, Moore and Stokes proposed a classification of ventral wall defects based on the appearance of the defect at birth, and the clinical distinction between gastroschisis and omphalocele was made[6].

In gastroschisis the umbilicus is attached to the normal site in an intact anterior abdominal wall. The defect through which the bowel herniates is usually in the right side of the umbilicus, separated from it by a small bridge of skin. There is no membrane over the bowel. The eviscerate bowel segment is commonly loops of small intestine and colon, sometimes stomach. It is usually not associated with other anomalies. If the gastroschisis had occurred early in utero, the bowel segment is wholly covered by fibrinous material as a result of reaction to amniotic fluid[2]. Unlike omphalocele, gastroschisis is not associated with chromosome abnormalities, so the survival rate is excellent (85% to 97%). Volvulus (rotation of the bowel) resulting in a compromised blood supply may, however, kill large regions of the intestine and lead to fetal death[1,3]. Gastroschisis associated with herniation of major viscus makes the prognosis worst. The size of the defect or the length between the diagnosis and the delivery does not influence the prognosis; thus early delivery does not appear an indication[7].

It is more common in younger mothers. The incidence is 10 - 20 folds higher in women younger than 20 years than in women aged 25 to 29 years[3]. Other causes may be maternal use of vasoconstrictive drugs (ephedrine, pseudoephedrine or cocaine) and smoking[1].

The unusual associations of gastroschisis are limb- body wall defect syndrome (amniotic band syndrome), meningocoele, abnormal genitalia, umbilical cord abnormalities and gastrointestinal anomalies such as intestinal atresia, stenosis and malrotation[8,9]. In the literature, extra-intestinal anomalies are reported in 5% to 29% of infants with gastroschisis and include a variety of anomalies affecting the skeletal system, cardiovascular system and central nervous system[3,10,11]. In a study by Stoll C et al. (2012) for associated malformations with gastroschisis, between 1979 and 2003 in 3,34,262 consecutive births, out of the 60 cases of gastroschisis, had associated malformations in 10 (16.6%) [9].

Hoyme and associates (1983) suggest that intrauterine thrombosis of the Right omphalomesenteric artery is the primary cause leading to infarction and necrosis of the base of the cord and herniation of the gut through this infarcted area[12]. According to deVries (2002), gastroschisis results from an abnormal involution of the right umbilical vein that leads to a paraumbilical defect through which the small bowel prolapses at approximately 37 days of embryonic life[13].

The diagnosis can be made with endovaginal sonography as early as 12 weeks[2,14,15]. Sonographic visualization of freely floating loops of bowel within the amniotic fluid with an abdominal wall defect to the right of the insertion of the umbilical cord at any point after the normal embryonic return of the intestine to the abdominal cavity at 10 weeks of gestation confirms the diagnosis. Maternal serum α-fetoprotein (MS-AFP) is elevated with fetal gastroschisis. Since MS-AFP screening has become widespread, the prenatal diagnosis of gastroschisis has increased markedly, such that up to 95% of cases are diagnosed before birth[3].

The mesenchymal anlaga of the hand bones are formed between 33 and 41 days of gestation (4.7 to 5.8 weeks). This also correlates closely with the development of an omphalocele during the fourth week. Congenital anatomic deformities could be related to a decreased intrauterine space. Decreased intrauterine space from oligohydramnios, as seen in the Prune-belly syndrome and Potter's syndrome, is felt to be the cause of severe deformities (arthrogryposis and associated clubfeet, dislocated hips, lower limb hemimelias and others)[4].

Limb differentiation occurs roughly between the fourth and fifth weeks of embryonic development. Position of a cell on the limb bud during early development determines the fate of that cell[16]. Limb duplication is a rare disorder. The extra limb is most commonly shrunken and/or deformed. Sometimes an embryo start as conjoined twins, but one twin degenerate completely except for one or more limbs, which end up attached to the other twin. Duplication of the limb derives from the influence of the Apical ectodermal ridge (AER). Abnormal splitting of AER creates 2 sets of limbs[17]. Limb duplication is commonly associated with vertebral and/or neural tube anomalies.

IV. Conclusion

Although Gastroschisis has rare associated malformations, this case of left sided gastroschisis highlight the associations of both spinal and upper limbs anomalies along with duplicate limb, which is a rare case.

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