

An unusual case of Foetal Gastroschisis with Limb aplasia and Extra-corporal liver

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Gastroschisis represents a herniation of abdominal contents through a paramedian full-thickness abdominal fusion defect without involving the umbilical cord. Evisceration usually only contains intestinal loops and has no surrounding membrane unlike in omphalocele. It is unusual for a newborn born with gastroschisis to have other serious birth defects and neonates 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 case is reported because of its rare association with extra-corporal liver and limb aplasia.

Introduction

Gastroschisis and omphalocele are congenital defects of the abdominal wall that occur in approximately 1.75 to 2.5 in 10,000 pregnancies.¹ Gastroschisis occurs when there is evisceration of abdominal contents through a paraumbilical defect in the abdominal wall and is less commonly associated with other abnormalities and has a better prognosis. On the other hand, omphalocele, which is a herniation of abdominal contents in to the base of the umbilical cord, is more common and carries an increased risk for concomitant abnormalities and associated poor prognosis. However presence of major organ herniation such as liver and spleen, makes the prognosis worst in both the cases.

Case report

A primi gravida at 30 weeks of gestation, reported to antenatal clinic at Dr. T.M.A. Pai Hospital (a peripheral hospital of Kasturba Medical College) with history of sense of heaviness in the abdomen and decreased foetal movement since two days. Patient had regular antenatal care outside. However she had not undergone any antenatal scans. General examination was unremarkable except for mild pallor. Abdomen was distended, tense and fluid thrill was present. Uterine height corresponded 34-36 weeks. Presentation was breech and foetal heart could not be localized. Ultrasound showed a single live foetus at 30W1D with gastroschisis and hydramnios. Liver and bowel loops

were freely floating in the amniotic cavity. There was no evidence of covering membrane. Right upper limb was not visualised.

The following investigations were obtained:

Hb% 9 gm%,

Blood group "O" positive,

HIV negative, HbsAg negative, VDRL negative,

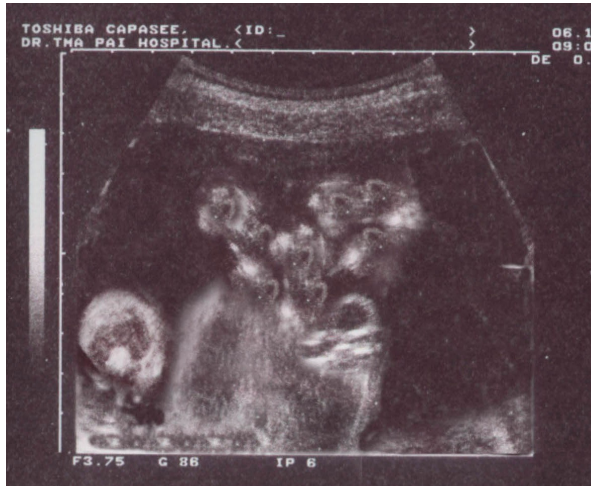
Urine examination -NAD

Serum AFP > 3500 ng/ml,

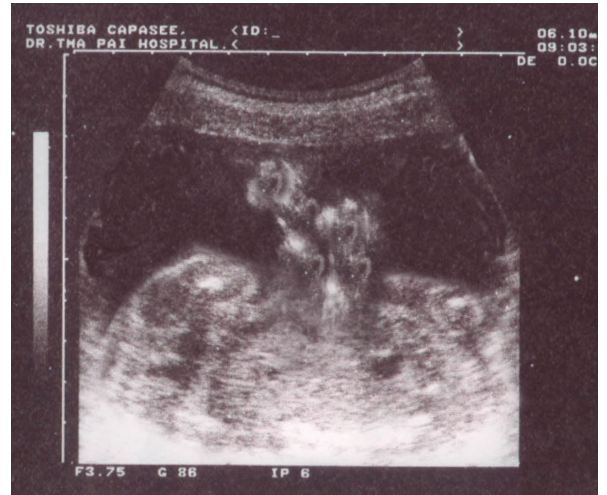
Serum Oestradiol > 3000 pg/ml

3 hr GTT: 84mg/ 196mg/ 163mg/ 89mg

With gastroschisis and right upper limb aplasia as the likely diagnosis and keeping in mind the presence of extra-corporal liver, she was explained about the magnitude of neonatal surgical problems and associated poor prognosis. As termination of pregnancy was opted by parents, she was admitted and cerviprim induction was done. She delivered a fresh stillborn female baby by breech. The foetus weighed 1.35 kg, 24.5 cm and in length and had multiple congenital anomalies with an abdominal wall defect through which most of the intestines and liver had eventrated. The defect was situated to the right of the insertion of the umbilical cord, which was normal, containing three vessels, and was separated by a tiny limb bud. There was aplasia of right upper limb. No other deformity was evident except for a common mesenterium. Autopsy showed hypoplastic right lung and an incidental cyst measuring 2.5 cm in the liver.



Picture1. Ultrasound scan showing transverse section of the foetus with loops of intestine freely floating in amniotic fluid.



Picture2. Ultrasound scan showing sagittal section of the foetus.



Picture3. Photograph showing liver and intestine lying outside the abdomen and intact umbilical cord.



Picture4. Photograph showing absent right upper limb.

Discussion:

Gastroschisis is a right paraumbilical defect varying between 2.5 and 5 cm involving all layers of the abdominal wall. Synonyms used to describe gastroschisis include paraomphalocoele, laparoschisis, abdominoschisis, and embryonal ruptured omphalocoele.¹ The small bowel always eviscerates through the defect and is, by definition, nonrotated and lacking secondary fixation to the posterior abdominal wall. Skin is rarely interposed between the defect and the umbilical cord.² The loops of bowel are never covered by a membrane; hence, they are directly exposed to the amniotic fluid. Alpha-foetoprotein levels are markedly elevated. The loops usually develop a fibrous coating and are matted together. Other organs that may eviscerate are the large bowel (often), the stomach, portions of the genitourinary system (occasionally), and the liver (very rarely). The location on the left side has been reported, but is very rare. Congenital abnormalities of the other systems are seldom noted.

According to deVries, 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 embryonic life, where as Hoyme and associates³ suggest that intrauterine thrombosis of the omphalomesenteric artery is the primary cause.

Gastroschisis is found either accidentally during second trimester anomaly scan or because of elevated maternal serum alpha-foetoprotein levels.⁴ The diagnosis can be made with endovaginal sonography as early as 12 weeks.^{5,6} The striking feature in the foetus presenting with gastroschisis is the multiple loops of bowel floating freely in the amniotic fluid.

Since gastroschisis is usually not associated with other congenital or chromosomal anomalies, it carries a much better prognosis. Possible causes of death in this group would include sepsis, surgical complications, and low birth weight. These foetuses seem to get the most benefit from early prenatal diagnosis since they can be prospectively followed for intrauterine growth retardation and obstruction of gastrointestinal tract, as

well delivery in a tertiary care center with an available pediatric surgeon. Because of improvement in surgical technique and in parenteral nutrition in the past 2 decades, the survival rate is 80% to 90%. In uncomplicated cases delivered in a tertiary care center, the survival rate can approach 100%. 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 indicated.⁷ The thickening and distension of the bowel and extracorporeal liver are associated with a poor prognosis.⁸

References

1. de Vries PA: The pathogenesis of gastroschisis and omphalocoele. *J Pediatr Surg* 15:245, 1980
2. Tibboel D, Raine P, McNee M, et al: Developmental aspects of gastroschisis. *J Pediatr Surg* 21:865, 1986
3. Hoyme HE, Higginbottom MC, Jones KL: The vascular pathogenesis of gastroschisis. Intrauterine interruption of the omphalomesenteric artery. *J Pediatr* 98:228-231, 1981
4. Holmgren G, Sigurd J: Prenatal diagnosis of two cases of gastroschisis following AFP screening. *Acta Obstet Gynecol Scand* 63:325, 1984
5. Guzman ER; Early prenatal diagnosis of gastroschisis with transvaginal ultrasonography. *Am J Obstet Gynecol* 162:1253, 1990
6. Redford DH, McNay MB, Whittle MJ: gastroschisis and exomphalos: precise diagnosis by midtrimester ultrasound. *Br J Obstet Gynaecol* 92,54, 1985
7. Bair JH, Russ PD, Pretorius DH, et al: Foetal omphalocoele and gastroschisis: A review of 24 cases. *AJR* 147: 1047, 1986
8. Kirk EP, Wah RM: Obstetric management of foetuses with omphalocoele or gastroschisis. A review and report of 112 cases. *Am J Obstet Gynecol* 146: 512, 1983

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