

Left sided Gastrochisis with spinal and limb deformities: a case report

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Abstract

Gastrochisis represents a herniation of abdominal contents through a paramedian full-thickness abdominal fusion defect. The abdominal herniation is usually to the right of the umbilical cord. Very few cases of left sided Gastrochisis have been reported, Here we report a case of left sided Gastrochisis along with multiple defects seen as spinal deformity, left upper limb abnormality with lower limb deformity (congenital talipes equino varus). Antenatal ultrasound will identify the majority of abdominal wall defects accurately. Hence Antenatal Ultrasound and screening for maternal AFP are important for follow up and management.

Key Word: Gastrochisis, deformities.


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INTRODUCTION

Gastrochisis is the most common major congenital abdominal wall defect along with omphalocele.¹ Gastrochisis 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 occasionally parts of other abdominal organs are herniated outside the abdominal wall with no covering membrane or sac. Etiology of gastrochisis are usually

considered to be the result of a vascular insult.² We report a fetus with a large left-sided gastrochisis which incorporated the liver, stomach, small intestine, colon and spleen, along with other malformations.

CASE REPORT

Mrs X 23 yr old primipara, unbooked case, with 35 weeks of gestation had no antenatal check up or ultrasound scan throughout pregnancy. She presented with labour pain and leaking PV, delivered a pre term dead anomalous male baby vaginally. Baby had large paraumbilical defect with herniation of stomach, small and large bowel loops, liver, gallbladder, and left kidney [Figure 1 & 2]. Left upper limb was rudimentary with two buds of fingers [Figure 3], Both lower limbs were pointing downwards and turned inside [Figure 4]. Left side hip was small with scoliotic curvature of vertebral column. Rudimentary male genital organs were seen. Umbilical cord was short and had two vessels. Head, face, neck, thorax, and other upper limb was normal.



Figure 1: Left Sided Gastrochisis



Figure 2: Contents of Gastrochisis



Figure 3: Rudimentary Upper Limb With Two Finger Buds



Figure 4: Talipes Equino Varus With Scoliosis

DISCUSSION

Gastroschisis is a congenital anterior abdominal wall defect, adjacent and usually to the right of the umbilical cord insertion which has no sac covering and no associated syndromes. Its incidence rate is 0.3-1 in 10,000 births.¹ It is rare when occurring to the left of the umbilical cord. Only a handful of similar cases have been reported.³⁻⁶ It has been hypothesized that involution of the right umbilical vein, which normally occurs during embryogenesis, results in decreased viability of the surrounding mesenchyma. Regression of the left umbilical vein or left-sided omphalomesenteric artery might lead to a left sided gastrochisis.⁷ It has to be differentiated from omphalocele, which usually is covered by a membranous sac and is frequently associated with other structural and chromosomal anomalies. However gastrochisis may be associated with gastrointestinal anomalies such as intestinal atresia, stenosis, and malrotation.⁸ Singal R *et al.* reported a rare case of a newborn baby with an abdominal wall defect, together with multiple congenital abnormalities and diagnosed as gastrochisis. There were multiple defects seen as spinal deformity, imperforate anus, esophageal fistula, and lower limb deformity (congenital talipes equinovarus) along with the webbing of neck. There were also ischemic changes present over the left upper limb in the form of cyanosis. The diagnosis made was gastrochisis and Omphalocele along with spinal deformity.⁹ Among the reported cases evisceration of the stomach, small and large intestine is common, and evisceration of the liver, kidney, urinary bladder and

ovary have all been reported^{10,11,12} Spinal bony abnormalities range from 30% to 44%. Scoliosis is associated with arthrogryposis (2.5% to 34% incidence)¹³ The true etiology of congenital clubfoot is unknown. Cytogenic abnormalities (e.g. – congenital talipes equinovarus (CTEV) deformity is usually seen in syndromes involving chromosomal deletion.

CONCLUSION

Gastroschisis usually carries a good prognosis. Hence, these fetuses benefit from early diagnosis which can be done by Antenatal ultrasound. Delivery in tertiary care hospital with intensive care management of neonate and improved surgical care has increased the survival upto 80%. Gastroschisis may be associated with gastrointestinal anomalies. The present case was a left sided gastrochisis with spinal and limb deformities The presented case is a rare association of left sided gastrochisis with both spinal and limb anomalies in primipara.

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