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Case Report

A case of left sided gastroschisis with pulmonary hypoplasia

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ABSTRACT

Gastroschisis is a congenital paraumbilical anterior abdominal wall defect resulting in herniation of abdominal wall contents. Incidence of gastroschisis is ~5 per 10,000 live births. Usually, it is right sided and left sided gastroschisis is rare. Gastroschisis in itself has a good prognosis, but the prognosis may vary with the severity of associated conditions. This article reports a case of left sided gastroschisis associated with pulmonary hypoplasia.

Keywords: Gastroschisis, Left-sided gastroschisis, Congenital anomaly, Case report

INTRODUCTION

Gastroschisis is a congenital paraumbilical anterior abdominal wall defect resulting in herniation of abdominal wall contents. Incidence of gastroschisis is approximately 5 per 10,000 live births. Usually, it is right sided and left sided gastroschisis is rare.[1]

CASE REPORT

A 28 years old female, gravida 2, para 1, living 1 at 21+2 weeks of gestation with no other known comorbidities presented with complaints of pain abdomen on and off since day before. Her anomaly scan showed mild prominence of right renal pelvis with no other major congenital anomalies, gave no h/o addictions/substance abuse. There was no h/o congenital anomaly in her family.

As per history and examination findings she was diagnosed to be a case of inevitable abortion, and was delivered vaginally as per institutional protocols. She expelled a dead male fetus of 345 g and placenta of 190 g.

On gross examination, small intestine was found to be herniating from a defect of approximately 3 cm at the left side of the abdominal wall. Contents were not covered by membrane. No other gross obvious anomaly was observed.

Figure 1: Anterior view.

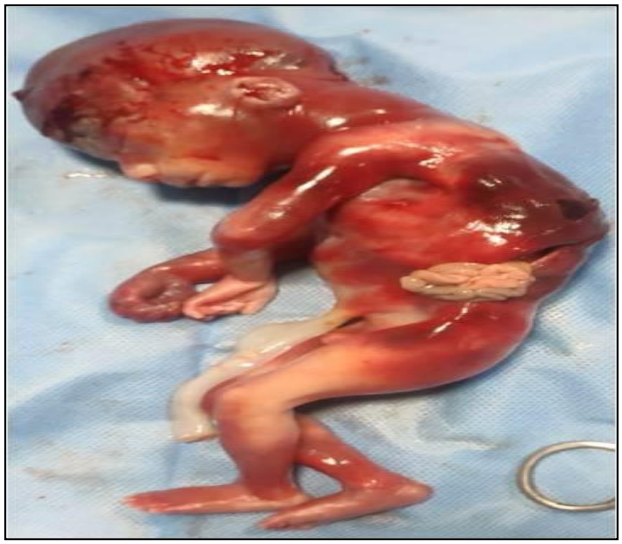

Figure 2: Lateral view.

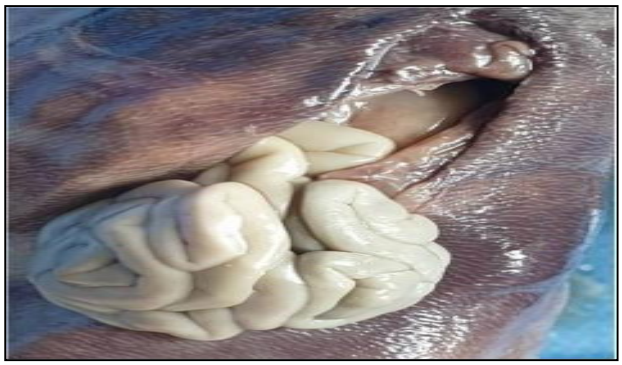

Figure 3: Gastroschisis.

Fetal autopsy, placental biopsy and infantogram were done. Placental biopsy was reported to be normal. Fetal autopsy was suggestive of pulmonary hypoplasia with left sided gastroschisis. Infantogram demonstrated no other anomaly except for the herniation of abdominal content.

DISCUSSION

Gastroschisis is a congenital abdominal wall defect, which is almost always seen as right sided. Risk factors for gastroschisis are younger maternal age, cigarette smoking, illicit drug use and genetic polymorphisms.[2]

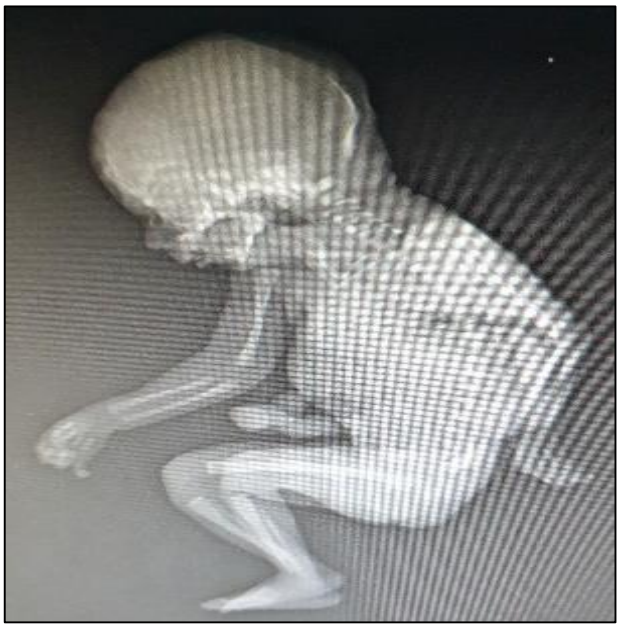

Figure 4: Infantogram.

Various theories have been proposed to explain the etiology of gastroschisis, most prevalent of which is vascular insult to the right umbilical vein. During the normal development, right umbilical vein undergoes involution around sixth week of the gestation. Vascular insult as well as subsequent thrombosis results in right-sided gastroschisis.[3] Other theories proposed include early body wall abnormal malformation, yolk sac failure, and prenatal rupture of the physiological hernia.[4] Barisic et al showed in their study showed that in 106 cases of gastroschisis, 59% had live birth, 12% were intrauterine fetal demise as well as the 29% pregnancies were terminated.[5] Left-sided gastroschisis is a rare entity, and its etiology is mostly unknown. Proposed theories for left sided gastroschisis are early regression of umbilical vein, a disorder of right-left axis orientation and abnormal folding of body wall.[6] A summary of cases of left-sided gastroschisis reported till date has been given below (Table 1).

Table 1: Summary of cases of left-sided gastroschisis reported.

Years	Authors	Sex	Gestational age (weeks)	Associated anomalies
1988	Blair et al[7]	M	-	None
1989	Hirthler et al[2]	F	27	Hyaline membrane disease
1989	Hirthler et al[2]	M	Term	None
1993	Toth et al[8]	F	35	None
2000	Thepcharoennirund et al[9]	F	36	None
2000	Thepcharoennirund et al[9]	F	40	None
2001	Pringle et al[10]	M	34	Left testis herniating through defect
2002	Fraser et al[11]	M	28	None
2002	Ashburn et al[12]	F	37	None
2004	Ameh et al[13]	M	Term	None

Continued.

Years	Authors	Sex	Gestational age (weeks)	Associated anomalies
2004	Orpen et al[14]	-	Term	Pseudoexstrophy, ASD, PDA, ureteral reflux
2004	Wang et al[15]	F	-	Situs inversus
2004	Yoshioka et al[16]	F	38	None
2004	Yoshioka et al[16]	F	34	Necrosis of herniated bowel
2006	Gow et al[17]	M	39	None
2007	Parsun et al[18]	F	24 (terminated)	Multicystic renal dysplasia
2008	Suver et al[19]	F	34	Absent corpus callosum, optic dysplasia, panhypopituitarism, intestinal atresia
2008	Suver et al[19]	F	35	Cerebral arteriovenous malformations
2008	Suver et al[19]	F	34	ASD, pulmonary valve stenosis
2009	Punia et al[20]	M	26 (Intra uterine fetal demise)	Meromelia of all four limbs
2010	Patel et al[21]	F	34	Small left colon syndrome
2012	Shi et al[22]	M	35	Liver/Stomach/Spleen herniation, VSD, scoliosis, small chest
2013	Mandelia et al[23]	M	Term	PDA
2013	Patel et al[23]	M	37	Hypoplastic left hemiscrotum, atrophic left testis
2015	Shin et al[24]	M	35	PDA, ASD, peripheral pulmonary stenosis
2015	Lubala et al[25]	F	-	Mirror image gastroschisis in female monochorionic twins
2015	Hombalker et al[26]	M	-	Cecal agenesis, short gut, malrotation
2017	Rahul et al[28]	F	Term	Intestinal atresia, perforated proximal ileum
2017	Litman et al[2]	F	34	Persistent superior vena cava, Left talipes equinovarus deformity, hypoplastic right third digit, right supernumerary $4^{th}/5^{th}$ digit
2017	Kalenga et al[29]	F	-	None
2017	Soomro et al[30]	F	36	Unspecified heart murmur
2017	Singh et al[31]	F	-	Meckel's Diverticulum
2017	Nam et al[32]	F	35	Situs inversus totalis
2018	Schierz et al[33]	M	33	Persistent right umbilical vein, right aortic arch
2018	Sullivan et al[34]	F	37	Septo-optic dysplasia
2020	Muta et al[35]	F	35	None
2020	Muta et al[35]	M	36	Umbilical hernia
2020	Masden et al[36]	F	37	Omphalocele
2021	Cannon et al[37]	F	41	None
2022	Nayak et al[38]	M	17-20 weeks old preserved specimen	None
2022	Abdullah et al[39]	F	24	None
2022	Srivastava et al (Current report)	M	21 (Intra uterine fetal demise)	Pulmonary hypoplasia

CONCLUSION

Left sided gastroschisis is usually associated with extra-intestinal congenital anomalies including choledochal cyst, cleft lip, cleft palate, pulmonary hypoplasia, ASD, PDA, etc. It can be diagnosed with ultrasound at about 20 weeks of gestation with specificity of ~95%, and surgical management can be planned. Gastroschisis in itself has a good prognosis, but the prognosis may vary with the severity of associated conditions.

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