CASE STUDY

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Congenital rare anterior abdominal wall defect affecting the perineum- a case study

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General Note
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ABSTRACT

Abdominal wall defects with gastroschisis not uncommon but gasroschisis associate with abdominal viscera herniating within the boundaries of perineum is uncommon. A male still born foetus was procured from local government hospital. The foetus showed

severe form of anterior abdominal defect and abdominal viscera herniating as caudally through the perineum, lower limb and spine deformities. This type of case is important for discussion of rare developmental malformations for post graduates, gynaecologists, paediatricians, etc.

Key words: equinovarus, gastroschisis, malformation, scoliosis, perineum

1. INTRODUCTION

Congenital anterior abdominal wall defects include omphalocele, gastroschisis, body stalk anomaly and prune-belly syndrome. Majority cases of omphalocele are associated with other serious structural defects and chromosomal abnormalities. Whereas, gastroschisis is usually an isolated lesion and is not associated with other structural defects and abnormal karyotype. Body stalk anomaly is also an isolated anomaly with rare association with chromosomal anomalies, but is lethal. Prune-belly syndrome is mostly associated with obstructive uropathy and severe maldevelopment of urinary tract. The final outcome of these defects is significantly affected by the presence of additional structural and / or chromosomal abnormalities (Axt R 1999 , Boyd PA 1998). Prenatal diagnosis is important for the parents as well as for the gynaecologists, paediatricians, and radiologists for discussion for such a type of rare development malformations.

2. CASE REPORT

A primi gravida of 20 weeks gestation delivered a stillborn male foetus. A male still born foetus was procured as a part of dissertation. In this case pelvic & lower limb abnormalities with equinovarus (Figure 1a, Figure 1b) are observed in addition to the abnormally large abdominal wall defect. This is a very rare case with severe gastroschisis leading to the exposure of not only intestines but also other abdominal viscera like liver, spleen and bladder herniating through the perineum right lateral to the external genitalia (Figure 1c) and scoliosis of spine (Figure 1d), a combination that has not been previously reported in the literature.. The umbilicus was attached to the right of abdominal wall defect. There was no history of consanguinity. Radiograph of the foetus showing abnormal short spine and pelvic bones (Figure 2a), and curved spine (Figure 2b).

3. DISCUSSION

Normal development of the anterior abdominal wall depends on the fusion of four ectomesodermic folds; cephalic, caudal and two lateral folds. Failure of lateral body folds to migrate centrally results in omphalocele. If the anomaly of the ventral wall is more extensive and, in addition to exomphalos involves cephalic embryonic fold then it results in pentalogy of Cantrell. Similarly, if the lateral fold defect is associated with caudal fold failure, it results in exstrophy of bladder or cloaca. Gastroschisis is found either incidentally during second trimester ultrasound scan (or during targeted ultrasound scan) or because of elevated MSAFP (Holmgren G 1984). The diagnosis can be made with endovaginal sonography as early as 12 weeks, however caution should be taken not to diagnose gastroschisis prior to 11 weeks' gestation due to the normal evisceration of the fetal bowel as part of normal embyogenesis at that time)(Guzman ER 1990, Redford DH 1985)

The severity ranges from a small vesicocutaneous fistula in the abdominal wall or simple epispadias to complete exstrophy of the cloaca involving exposure of the entire hindgut and the bladder (Mirk P 1993, Kutzner DK 1988). Serial ultrasound follow-up is important because later in pregnancy bowel obstruction, peritonitis, bowel perforation, and fetal growth restriction may occur (Paidas MJ 1994).

4. CONCLUSION

This case may be a rare foetal anomaly of anterior abdominal wall and evisceration herniating through perineal muscles. Very rarely is gastroschisis associated with herniation of other organs, and makes the prognosis worse. Survival rate of patients with abdominal wall defects has gradually improved; the outcome is largely dependent on coexisting anomalies.

Figure 1a Showing liver herniation and equinovarus of left foot.
Figure 1b Defective lower limbs
Figure 1c Showing gastroschisis and abdominal viscera herniating through perineum wall
Figure 1d Showing scoliosis

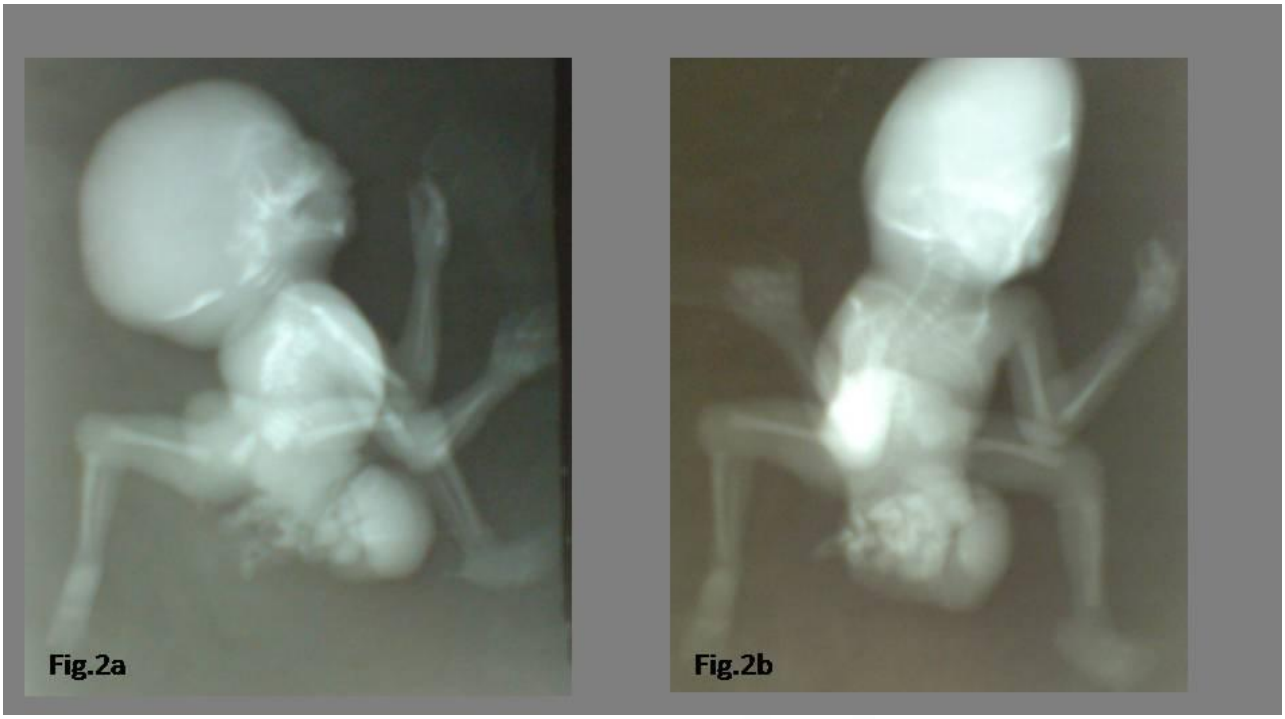

Figure 2a showing abnormal spine and deformed pelvic bones
Figure 2b showing scoliosis of spine

REFERENCE

1. Axt R, Quijano F, Boos R et al . Omphalocele and gastroschisis: Prenatal diagnosis and peripartal management. A case analysis of the years 1989-1997 at the Department of obstetrics and Gynecology, University of Homburg / Saar. Eur J Obstet Gynecol Reprod Biol. 1999 Nov, 87(1): 47-54
2. Boyd PA, Bhattacharjee A, Gould S et al. Outcome of prenatally diagnosed anterior abdominal wall defects. Arch Dis Child Fetal Neonatal Ed. 1998 May; 78 (3): F 209-13.
3. Guzman ER. Early prenatal diagnosis of gastroschisis with transvaginal ultrasonography. Am J Obstet Gynecol. 1990; 162:1253–4. [PubMed]
4. Holmgren G, Sigurd J. Prenatal diagnosis of two cases of gastroschisis following α-fetoprotein (AFP) screening. Acta Obstet Gynecol Scand. 1984; 63:325–8. http://doi.org/b3fsp5. [PubMed]
5. Kutzner DK, Wilson WG, Hogge WA. OEIS complex (cloacal exstrophy): prenatal diagnosis in the second trimester. Prenat Diagn 1988; 8:247-253.
6. Mirk P, Calisti A, Fileni A. Prenatal sonographic diagnosis of bladder exstrophy. J.Ultrasound Med 1993;5:291-293
7. Paidas MJ, Crombleholme TM, Robertson FM. Prenatal diagnosis and management of the fetuses with an abdominal wall defect. Semin Perinatol 1994, 18:196
8. Redford DH, McNay MB, Whittle MJ. Gastroschisis and exomphalos: precise diagnosis by midpregnancy ultrasound. Br J Obstet Gynaecol. 1985; 92:54–9. http://doi.org/cvk69h. [PubMed]