CASE REPORT

Inferior gastroschisis

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SUMMARY

Gastroschisis is a common defect of the central abdominal wall, nearly always located to the right of the umbilicus. We report a baby boy with gastroschisis who had a defect on the inferior side of an intact and normal umbilical cord which was managed by primary repair and an uneventful recovery. Such a case has never been reported in the literature. This case contributes towards enlarging the spectrum of gastroschisis.

BACKGROUND

Gastroschisis is a congenital abdominal-wall defect, which typically occurs to the right of the umbilicus, is small in size and is an isolated defect. There are 13 cases of left-sided gastroschisis,^{1–10} three cases of left-sided abdominal wall defect in left hypochondrium,^{11–13} one case of epigastric lesion¹⁴ and one case of bilateral symmetrical abdominal wall defects, however none of inferior or infraumbilical location has been reported in world literature.^{1–15}

CASE PRESENTATION

A baby boy weighing 3160 g was delivered at 37 weeks of gestation by emergency caesarean section because of fetal distress after artificial rupture of membranes. There was no meconium in the liquor. The mother was 25 years old and along with the father was non-consanguineous. Antenatal scans at 13 and 17 weeks detected gastroschisis following which she was counselled. The baby boy was born in good condition at Leicester Royal Infirmary and required no resuscitation. His APGAR score was satisfactory. He was resuscitated and prepared for immediate surgical repair.

The clinical examination showed a moderate-sized bowel evisceration through the defect which was covered with bowel bag.

INVESTIGATIONS

Urine dipstick was normal. Complete blood count, coagulation screen, renal, liver and bone profiles were normal. Ultrasound scan for abdomen and kidneys was normal.

DIFFERENTIAL DIAGNOSIS

Inferior gastroschisis and ruptured exomphalos minor were considered.

TREATMENT

After his condition was stabilised, he was taken to the operating theatre for reduction of bowel and repairing the defect. The eviscerated organs included small and large bowel loops which were not thickened, oedematous, shortened, matted or having a peel. The defect was inferior to the

umbilicus, the eviscerated bowel could be reduced into the abdominal cavity successfully after gentle stretching of the abdominal wall. During operation no situs inversus viscerum was found and the liver and falciform ligament were normal. There was non-rotation of the midgut. The umbilical vein was patent and entered the liver at the falciform ligament. The normal three-vessel cord included a vein and two arteries.

The defect could be closed primarily. Both the testes were descended. Rectal washout with tween 80 and bowel decompression was carried out.

OUTCOME AND FOLLOW-UP

Postoperation recovery was steady and the feeds were started on day seven and were gradually increased. The baby boy's inferior gastroschisis scar below the umbilicus healed primarily (figure 1). He tolerated feeds and responded positively. He was on full feeds on day 18 when total parenteral nutrition (TPN) was stopped. He was discharged home. On follow-up, he remained asymptomatic, and is thriving well with normal gastrointestinal function.

DISCUSSION

In most patients with gastroschisis, the abdominal wall defect occurs on the right side of the umbilicus. Only 12 cases of left-sided gastroschisis have been reported in the literature^{1–10} with three being abdominal defects away from the umbilicus situated in the left hypochondrium.^{11–13}

There is one case of supraumbilical midline defect¹⁴ and one of bilateral symmetrical defect in the supraumbilical region of the anterior abdominal wall.¹⁵ However, we could not find a single case of isolated inferior or infraumbilical gastroschisis.

Figure 1 Postoperative photograph showing primarily healed inferior gastroschisis scar just below the umbilicus. Note the superior, right and left side of the umbilicus is completely free of any defect/scar.

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Although multiple hypotheses have been proposed, the embryo genesis of gastroschisis is controversial and largely unproven. Here we report an unusual, previously unreported case of gastroschisis adding to the spectrum but challenging existing theories.

DeVries proposed that abnormal atrophy of the right umbilical vein resulted in a mesenchymal defect at the junction of the body stalk and the body wall.¹⁶ This theory best explains right-sided lesions and left-sided lesions with situs inversus.

The pathogenesis of left-sided gastroschisis is a mirror image variation of the right-sided lesion and its overwhelming female preponderance, the left hypochondrial defects, supra umbilical midline and bilateral defects and that of our case with infraumbilical defect largely remains speculative and may be better understood when more becomes known about the role of genes, influence of hormones and the environment in the causation of the defects.

Learning points

- ▶ Gastroschisis should be considered as a diagnosis when there is congenital abdominal wall defect around the umbilicus but the umbilical ring is intact and umbilical cord normal.
- ▶ Inferior variety is atypical, typical being to the right of the umbilicus.
- ▶ This variety has not been reported earlier in live born babies.
- ▶ This case adds to the spectrum of the varieties of gastroschisis around the umbilicus.
- ▶ Prenatal diagnosis of gastroschisis is possible but the inferior variety can only be established on postnatal examination as the defect is small and difficult to locate.

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Competing interests None.

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