CASE REPORT

Closing left gastroschisis with vanishing left testis

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SUMMARY

We report a baby boy with gastroschisis with left non-palpable undescended testis who had a defect on the left side of an intact and normal umbilical cord and had associated testicular atrophy and abnormalities of the ductus deferens. They were successfully managed by primary repair and had uneventful recovery. Subsequent inguinal exploration confirmed blind ending vas deferens and vanishing left undescended testis. Our case confirms vascular accidents at the narrow abdominal wall defect can lead to vanishing testis following attempts at closing gastroschisis making the defect narrow and compromising the blood supply to the testis.

BACKGROUND

Gastroschisis is a para umbilical defect of the anterior abdominal wall, which is nearly always located to the right of the umbilicus. It is generally small, without a membrane, isolated defect through which bowel eviscerates. There are only 21 cases of left-sided gastroschisis reported in the world literature.^{1–16} We wish to add one more case of left-sided gastroschisis which had the associated abnormalities of the vas deferens with ipsilateral testicular atrophy/infarction.

CASE PRESENTATION

A baby boy was born to a healthy 33-year-old multipara. He had a prenatal diagnosis of gastroschisis and was delivered at 37+2 weeks by a lower segment emergency caesarean section. The birth weight was 2740 g. At birth the patient was noted

to have a left-sided gastroschisis with hypoplastic left hemiscrotum and non-palpable left undescended testis (figure 1). A 3 cm defect was noted towards the left of the umbilicus. The heart sounds were on the left chest and there was no evidence of situs inversus.

INVESTIGATIONS

All laboratory investigations including the complete blood count, renal profile and coagulation screen were normal. Baby gram and renal ultrasound were normal. There was no evidence of left testis in the left hemiscrotum or in the groin on ultrasound scan.

DIFFERENTIAL DIAGNOSIS

A differential diagnosis of left gastroschisis with left-undescended testis or a closing gastroschisis with vanishing left testis and ruptured exomphalos was considered. The patient was re-examined for situs inversus which was absent.

TREATMENT

After a period of resuscitation and stabilisation the patient was taken to the theatre. Examination under anaesthesia after cleaning the prolapsed viscera revealed following findings.

The stomach was prolapsed and the small bowel shortened with a 2 cm mesenteric defect noted. The small and the large bowels were patent and splenules were noted in the small bowel mesentery. The liver and spleen were contained within

Figure 1 Preoperative photograph showing the left-sided gastroschisis with absent of the left testis in the scrotum or groin and atrophic testis seen at the base of the defect.

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the abdominal cavity. The bowel was gently reduced into the abdominal cavity and the abdominal wall closed in layers. A lesion in the small bowel mesentery was sent for histological analysis.

He underwent reduction of all viscera into the abdominal cavity followed by primary closure of his left-sided anterior abdominal wall defect with a purse string suture at the base of the umbilicus and leaving a sizable stump of the umbilical cord with a clamp on it leaving a natural umbilical scar.

OUTCOME AND FOLLOW-UP

Postoperatively the patient was ventilated for 1 day and received total parenteral nutrition for 16 days. The patient made an uneventful recovery and was discharged home after 19 days in the hospital. At 6-month follow-up, he had a small incisional hernia and the left testis was not palpable in the scrotum or groin.

One year later the patient was re-admitted on an elective basis for repair of an incisional hernia as a result of the original surgery and an exploration of his left groin as his left hemiscrotum was seen to be empty and there was no palpable left undescended testis.

At surgery the left inguinal canal was empty with no processus vaginalis. A blind ending structure was seen that was resected and sent for histological analysis.

Histology from the first operation showed that the lesion taken from the small bowel mesentery contained normal epididymis plus vascular connective tissue, with an adjacent portion of tissue which had undergone complete infarction leaving its exact nature difficult to ascertain. The results of a histology from the second operation confirmed that the blind ending structure was vas deferens with no testis or associated structures. The conclusion was that closing left gastroschisis has led to strangulation of blood supply to the left undescended testis leading to vanishing left testis and blind ending vas.

DISCUSSION

In most patients with gastroschisis, the abdominal wall defect occurs on the right side of the umbilicus. Occasionally a mirror image variation on the left side is reported and recently a case each of midline epigastric defect, bilateral pararectal defects and cases of left hypochondrial abdominal wall defects have been described.^{17–20}

Of 12 reported cases till 2010; 8 were girls, 1 boy and in 3 sexes were not mentioned.

Evisceration of the stomach, small and large intestine is common and evisceration of the gallbladder, urinary bladder, uterus, ovary and testis have all been reported. All cases could be closed primarily but one case required staged closure and almost all except one had no associated anomalies.²¹

Pringle²² described two patients: one with left and the other with right-sided gastroschisis, each associated with the respective testis 'hanging out of the defect on the testicular vessels and vas'. In each of these cases the testes were successfully salvaged. It is possible that association of the gastroschisis and cryptorchidism is incidental and strangulation at the level of the defect may have caused testicular infarction and/or atrophy. This is being explained by the concept of closing gastroschisis described recently with the help of MRI scans.²³

Although multiple hypotheses have been proposed, the embryogenesis of gastroschisis is controversial and largely

Figure 2 Schematic diagram showing full thickness defect in the anterior abdominal wall in the para umbilical area. Note vulnerability of gonadal vessels for vascular compromise as first step towards closing gastroschisis. ARS, anterior rectal sheath; RM, rectus muscle; PRS, posterior rectal sheath; CG, closing gastroschisis; LT, left testis with vas and vessels very close to closing defect.

unproven. Our cases with large, left-sided defects associated with multiple splenoculi in our case in addition to vas deferens and testicular pathology certainly adds to the spectrum of gastroschisis but at the same time it challenges the existing theories. Left-sided massive mesodermal insult during embryogenesis may have resulted in all these left-sided anomalies in our case.

DeVries²⁴ 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. This theory best explains right sided lesions and left-sided lesions with situs inversus.

There are three stages in the evolution and course of this anomaly. *Gastroschisis* is defined as a full-thickness abdominal wall defect (usually right-sided) with intestinal prolapse and occasionally other viscera. A *closed gastroschisis* or *vanishing midgut* is one in which the defect itself may close around the viscera causing exit or entry level intestinal atresia and ischaemia or midgut infarction.²⁵ Recent serial ultrasound and MRI has led to unfold the secret of the spectrum of *closing gastroschisis* or *vanishing gonads* in which infants are born with various stages of closing of gastroschisis defect and affects the vascularity of organs of evisceration close to the defect and the ability of the organ to resist ischaemia. Close antenatal monitoring may prevent severe bowel loss in some cases²⁶ but the testis has very low threshold of tolerating ischaemia and vanishing testis may be difficult to diagnose prenatally and hence may not be able to salvage it (figure 2).

In conclusion we believe that this hitherto unreported observation alerts us to the need to examine the vas deferens in male children undergoing repair of the left-sided gastroschisis.

Learning points

- ▶ Gastroschisis have the right paraumbilical defect and left mirror image gastroschisis is very rare.
- ▶ This is the first case of closing left gastroschisis leading to left vanishing testis due to vascular compromise at closing defect.
- ▶ Most cases reported with the left gastroschisis were females but our case is a male infant.
- ▶ Spontaneous descent in undescended testis associated with gastroschisis to a varying degree is common provided the testis remains viable.
- ▶ If testis is undescended in a case of gastroschisis. An active search for any tissue near the defect and accompanying vas should be made during surgery and sent for histopathology to ascertain its nature and extent of underlying process.

Contributors All the authors have made substantial contributions to the conception and design of this paper, search of the literature, the acquisition, analysis and interpretation of the data, to drafting the article or revising it critically for important intellectual content and to the final approval of the version to be published.

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