A Challenging Case of Anorectal Malformation With a "Twist"

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Introduction

Anorectal malformations (ARM) occur in about 1:2000 – 5000 live birth and it remains one of the more commonly encountered congenital anomalies. It has a wide range of associated conditions as well as variable presentations. We present a case of anorectal malformation with abnormal position of sigmoid colon resulting in a challenging stoma creation. .

Case Summary

A term newborn male infant who underwent investigation antenatally for a cystic lesion suspicious of sigmoid colon was noted to have imperforate anus with no perineal fistula at birth. There was no meconuria up to 24hours of life. Detailed antenatal scan at 22 weeks showed a right sided cystic mass. A fetal MRI at 26 weeks gestation showed a grossly dilated rectum and sigmoid with beaking towards the perineum and unable to trace upwards beyond sigmoid colon. The small bowel is in normal position and less likely to be malrotation. Patient was planned for left proximal sigmoid colostomy but the sigmoid colon was grossly dilated and fixed to the right lateral wall. A decision was made to convert to a full laparotomy.

Intraoperatively there was a grossly dilated sigmoid colon which was fixed to the right lateral abdominal wall pushing the caecum to the left, resulting in a twisted configuration of large bowel causing a 180' rotation about the rectum. The duodenojejunal junction was in normal position and the small bowel was not dilated. Incidentally he was noted to have a patent urachus and a rectourethral fistula was identified during urinary catheter insertion. The large colon was mobilized and repositioned to normal configuration. Caecopexy was performed at right lower quadrant after appendicectomy and a loop sigmoid colostomy was created in left lower quadrant. Patent urachus was excised. Patient was nursed in neonatal ICU (NICU) and discharged home on post operative day 5.

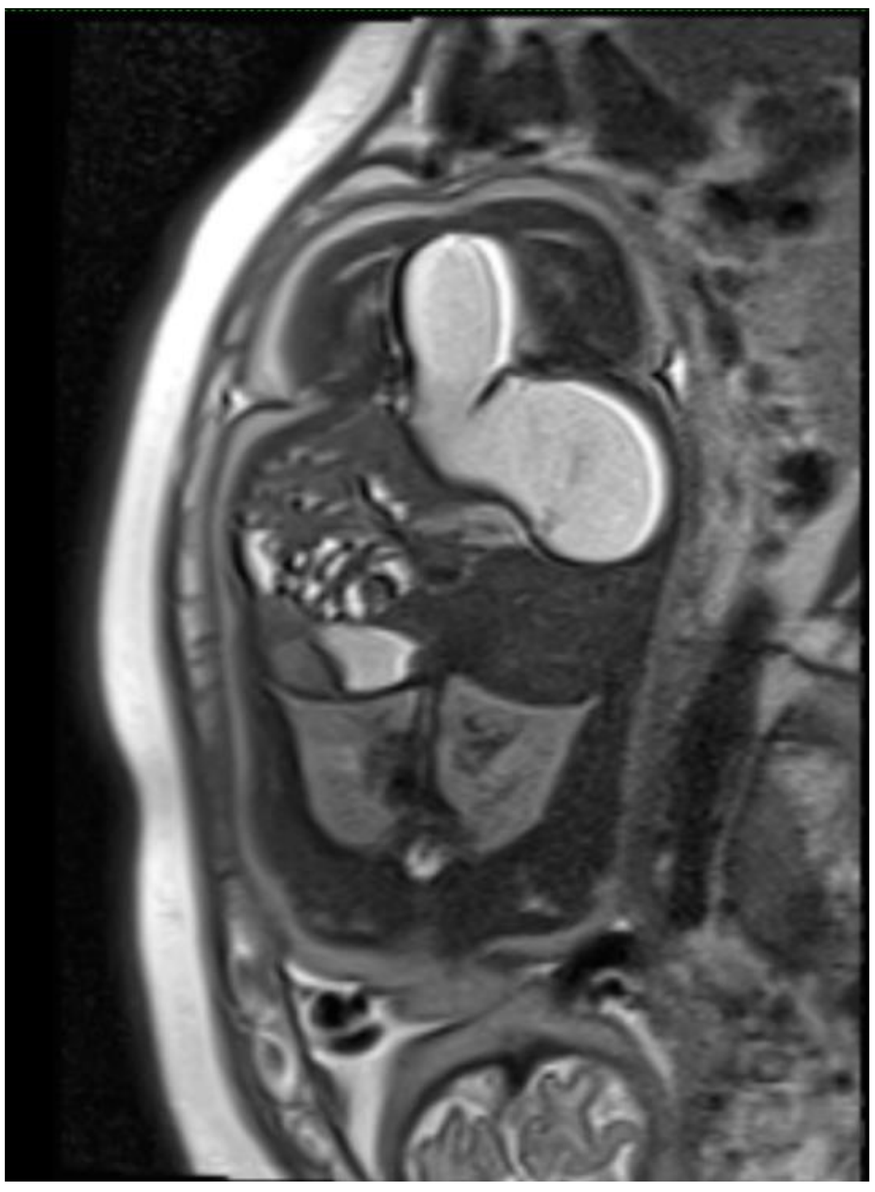

Figure 1. Antenatal MRI showing a grossly distended sigmoid colon on right side of abdomen.

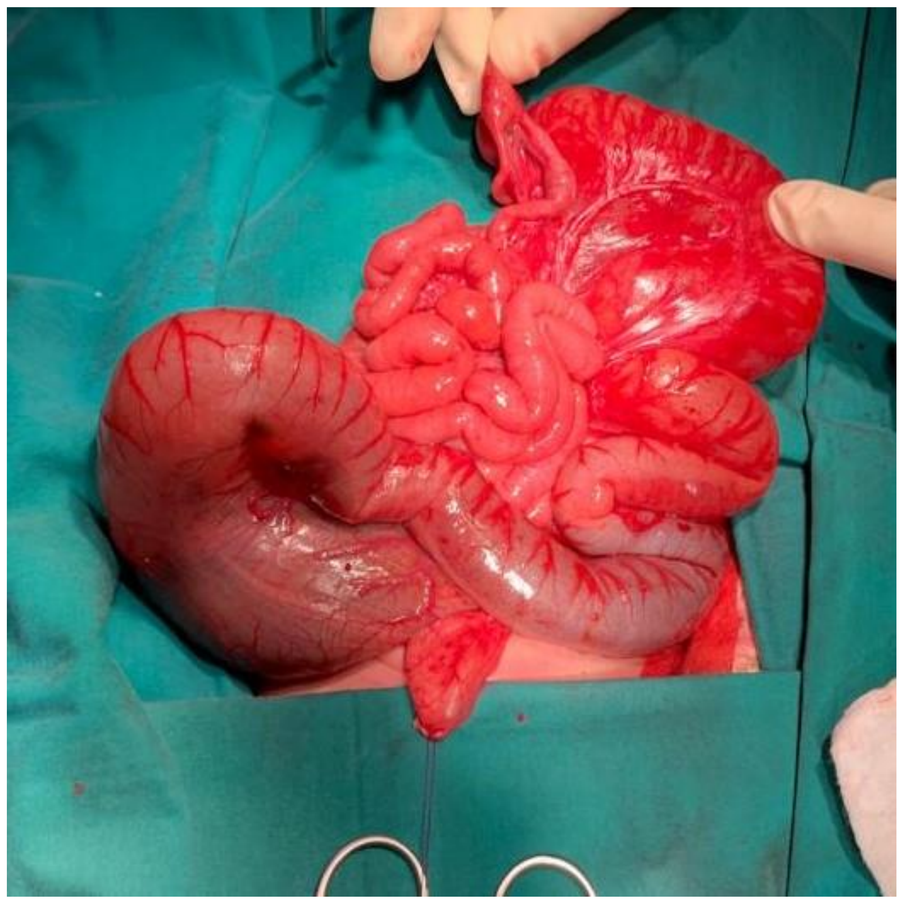

Figure 2. Sigmoid colon is grossly dilated and fixed to right lateral wall. Figure above shows after mobilization from right lateral wall i.e Toldt's line. The distended sigmoid colon displaced the caecum to the left hypochondrium. Urinary bladder is seen retracted inferiorly.

Discussion

Anorectal malformation is a form of congenital malformation with wide range of associated anomalies (VACTERL, Trisomy 21, Currarino syndrome etc.)[1]. Urgency for intervention is determined on the basis of ability to pass meconium [2]. Malrotation associated with anorectal anomalies only occur in 4.8% of the cases [3], and usually this would require additional procedure to establish gastrointestinal continuity. It is interesting in this case to note that the caecum was first encountered upon a left lower quadrant incision, raising a suspicion of possible malrotation before a full laparotomy revealed the entire anatomical configuration.

However , this abnormal positioning of the bowel is not due to malrotation. So far, there has only been one reported case of this type of bowel arrangement and it was found in asymptomatic adult cadaver during routine anatomy dissection. No similar findings have been reported in neonatal group. The embryological basis for this variant is thought to arise from defective fixation between 12-17th week of intrauterine life[4-5].

Conclusion

This case highlights the rarity of anatomical variant other than malrotation in congenital anomalies such as anorectal malformation where an abnormal fixation of the sigmoid colon resulted in a near closed loop obstruction of the distal bowel. A laparotomy was needed to complete the mobilization and decompression.

References

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