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BRIEF COMMUNICATION

Congenital Pouch Colon: A Case Report

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Abstract Congenital pouch colon (CPC) is a rare condition in which a variable length of colon is converted into a pouch which communicates with the urogenital tract. We report the antenatal sonographic findings of a boy found to have CPC type I postnatally, which was suspected to be an anorectal malformation in utero. This entity has been described in the literature mainly from the Indian subcontinent. Therefore, it is an important differential diagnosis to be kept in mind in fetuses with abnormal large bowel pattern.

Keywords Congenital pouch colon · Ultrasound · Fetus · Bowel · Anorectal malformation

Case

31-Year-old lady was referred for suspected fetal bladder outlet obstruction due to the observation of bilateral hydronephrosis and an overdistended urinary bladder during the course of a sonogram to assess fetal growth. There was no history of consanguinity. The couple's first child was a healthy five-year old boy. Their medical and family history was unremarkable. Sonogram revealed a fetus corresponding to 34 weeks of gestation. There was mild right pyelectasis (renal pelvis—8 mm) and moderate left hydronephrosis (renal pelvis—10 mm, calyceal dilatation—2 mm). The sagittal length of the fetal urinary bladder was 3.9 cm (Fig. 1a). The posterior urethra was not dilated. Liquor was adequate. The small bowel was seen primarily in the center and right side of the abdomen below the liver with the superior mesenteric artery coursing towards it (Fig. 1b). A hypoechoic, meconium filled, sac like structure was seen in the fetal abdomen on left side caudal to the stomach (Fig. 1c). Large bowel could not be seen separately. The perianal muscular complex was not delineated (Fig. 1d).

Diagnosis of an anorectal malformation was made. The patient delivered a 2.8 kg boy at 38 weeks by spontaneous vaginal delivery. The baby passed urine within 6 hours of birth. Examination of his perineum revealed absent anus with a dimple in its place. The external genitalia and urethral opening were normal. On operation, the small bowel was seen on the right side and the colon was entirely absent. The ileum was opening into a pouch on the left side. The pouch was communicating with the urinary bladder through a fistula. A diagnosis of congenital pouch colon (CPC) type I was made. The pouch and fistula were excised. The baby was discharged with an ileostomy. He is 5 months old and awaiting definitive surgery.

Discussion

CPC is an uncommon condition in which a variable length of the colon is replaced by a pouch-like structure that communicates with the urogenital tract through a fistula [1]. It is associated with anorectal agenesis. The condition has a male preponderance with a male to female ratio of 2.25:1–7:1 [2, 3]. Majority of the cases of CPC have been reported from India. It has been classified into various types. The most widely used classification of CPC given by Narasimharao et al. [4] divides the condition into four groups based on the length of normal colon present

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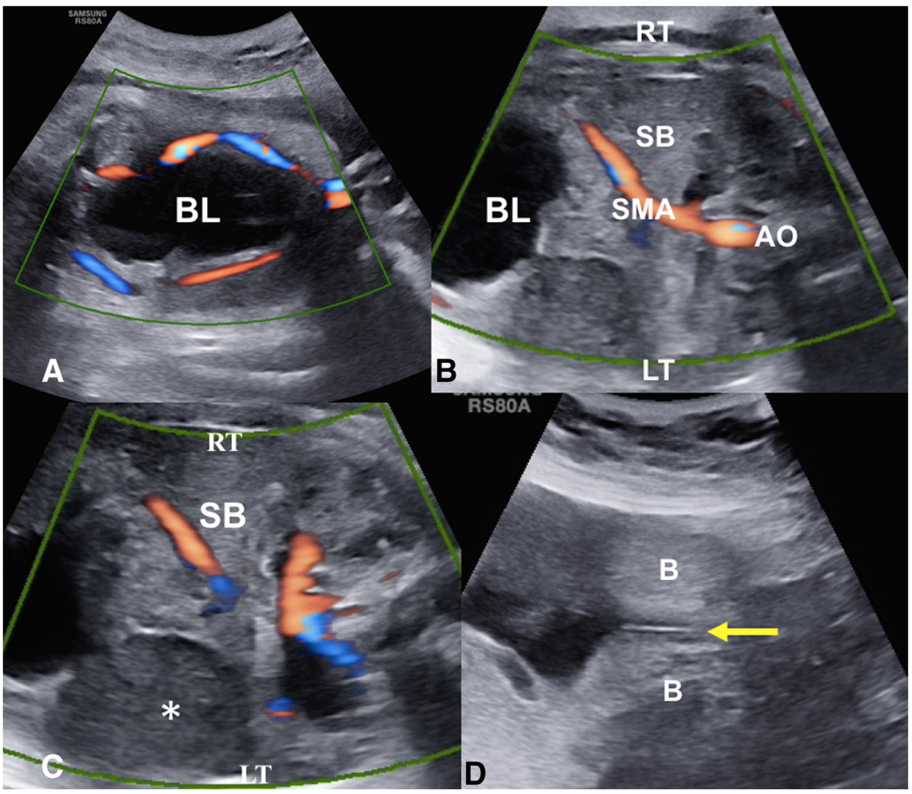

Fig. 1 Fetus with congenital pouch colon type I at 34 weeks. **a** Color Doppler image of the pelvis showing the urinary bladder (BL) flanked by the umbilical arteries on either side, **b** oblique coronal image of the abdomen showing small bowel loops (SB) in the center and to the right (RT) with the superior mesenteric artery (SMA) coursing towards them. Aorta (AO), Left (LT), **c** hypoechoic meconium filled pouch (asterisk) in the abdomen on left (LT) side and small bowel (SB) on right (RT) side, **d** non visualization of the target shaped perianal muscular complex in between the buttocks (B). Instead, an echogenic line (arrow) seen

proximal to the pouch. Type I CPC, seen in the present case, is the most severe form with total absence of the colon whereas type IV CPC has a near normal colon with only its terminal portion converted into a pouch. Saxena and Mathur [5] added a new type, type V wherein there are two pouches interposed by a normal segment of the colon and the distal pouch ends in a fistula with the urogenital tract. The fistula opens into the urinary bladder in males. In females, it may open in the cloaca, vagina or the vestibule.

The exact embryogenesis of CPC is unknown. Several theories have been put forth. It was initially believed that the pouch was formed as a consequence of obstruction. Since the pouch fails to regress even after a colostomy, this theory was discarded. Chadha et al. [1] have proposed that it occurs due to a vascular insult at the time of the partitioning of the cloaca by the urorectal septum. Wakhulu et al. [6] postulated that it is a stage in the development of cloacal exstrophy. Many anomalies have been reported in association, namely genitourinary, gastrointestinal, vertebral and cardiac [7]. Hydronephrosis and vesicoureteric reflux are the most common. In females, it is often associated with a cloacal anomaly.

This case illustrates the antenatal sonographic findings of a baby with type I CPC which is the first in literature to the best of our knowledge. It highlights the importance of a careful examination of the morphology as well as the location of the fetal bowel. Normal fetal small bowel lies in the mid and lower abdomen caudal to the liver and stomach (Fig. 2). In the present case, small bowel was seen in the center and right side of the abdomen caudal to liver. The normal hypoechoic, meconium filled colon can be easily identified in the periphery of the small bowel in the third trimester. Non visualization of the normal colon and perianal muscular complex along with the presence of a meconium filled sac—like structure are clues to the diagnosis of CPC. It is pertinent to remember that there are associated malformations, most common being

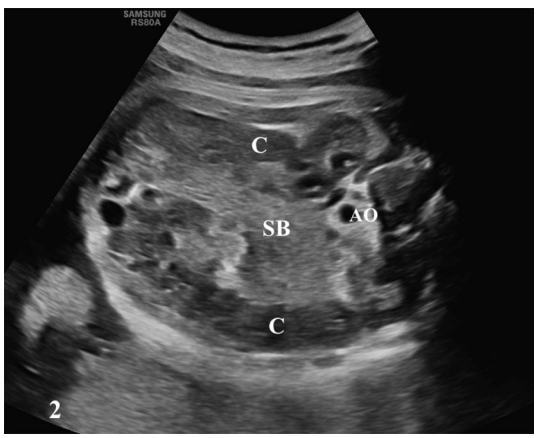

Fig. 2 Bowel pattern in a normal fetus at 34 weeks. Small bowel loops (SB) in the abdomen with the hypoechoic, meconium filled colon (C) in the periphery. Aorta—AO

genitourinary. Sometimes, these might be the initial sonologic findings drawing our attention. This case underscores the importance of a meticulous examination of the fetus when a seemingly innocuous observation (for instance, pyelectasis) is made on ultrasound. Since the gastrointestinal and the genitourinary tract develop in close proximity in the embryo, their anomalies are often concomitant. Therefore, an abnormality in one system should prompt a closer scrutiny of the other.

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