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Case Report

Antenatal Diagnosis of Vesicocolic Fistula

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Abstract

Urorectal septum malformation sequence (URSMS) is a rare congenital anomaly characterized by lack of perineal or anal opening, external genital defects, and various urogenital and colonic abnormalities. This condition results from deficient separation of the cloaca and the failure of the membrane to rupture. Here, we present a unique case of URSMS with prenatal diagnosis, which is extremely challenging to identify before birth. Prenatal ultrasonography revealed the presence of intraluminal enterolithiasis in the sigmoid colon, indicating high anal atresia and a vesicocolic fistula. The diagnosis was confirmed postnatally by clinical examination and ascending cystourethrogram. This case highlights the importance of prenatal screening in detecting rare congenital anomalies, and the role of ultrasonography in distinguishing enterolithiasis from other conditions. Moreover, it underscores the significance of precise fistula location determination for optimal neonatal management of anorectal malformations.

Keywords

- urorectal septum malformation sequence
- prenatal diagnosis
- vesicocolic fistula
- intraluminal enterolithiasis
- ascending cystourethrogram
- anorectal malformations

Introduction

The term "urorectal septum malformation sequence" (URSMS) describes a rare congenital condition characterized by the absence of a perineal or anal opening, external genital defects, as well as abnormalities in the urogenital system and colon. URSMS represents a collection of anomalies that arise due to insufficient separation of the cloaca and the failure of the membrane to rupture. This condition was initially documented by Escobar et al[1] and has an estimated incidence rate ranging from 1 in 50,000 to 1 in 250,000 births.[1]

The range of severity of URSMS varies from partial to full forms. Partial URSMS is a milder variation in which there is only one perineal opening serving as a common exit for both faeces and urine. The exact cause of URSMS remains unclear, but it has been suggested that it may result from an incomplete division of the cloaca and abnormal development of the urorectal septum.[1]

Prenatal diagnosis of URSMS is extremely challenging. Features such as enterolithiasis (presence of calculi or stones in the intestines) and vesicocolic fistulae (abnormal connections between the urinary bladder and colon) are infrequent but highly specific indicators of URSMS.

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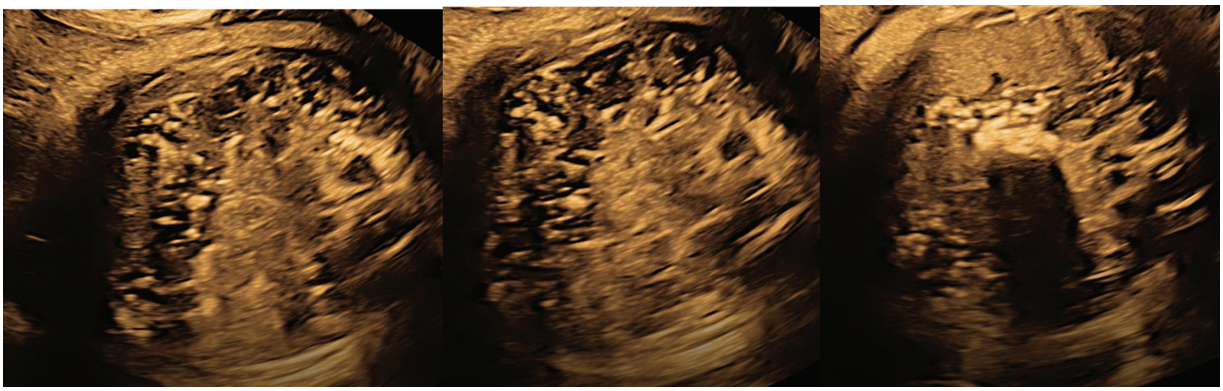

Fig. 1 Prenatal ultrasound in the third trimester showing large colon with echogenic and floating fecolith.

Neonatal anorectal anomalies are frequently linked to fistulas, particularly in cases of imperforate anus. These congenital rectourinary fistulas in neonates can manifest as rectovesical, rectourethral at the prostatic or bulbar level and rectourethral at the distal urethra. An ascending cystourethrogram is used to visualize the abnormal passage connecting the rectum and the prostatic urethra.[2]

The presence of meconium calcification within the fetal and neonatal bowel is a rare discovery, usually occurring in the context of intestinal blockages. This condition stands out because calcifications are located inside the intestinal lumen, setting them apart from calcifications resulting from bowel perforation or meconium peritonitis.

The earliest documented case of neonatal intraluminal enterolithiasis dates back to 1813, as reported by Fournier and later cited by Walker in 1948.[3] In this case, the author described a situation involving rectal atresia, where the enterolith had passed through a rectovaginal fistula.

A literature search revealed 48 cases of intraluminal calcifications; all of them had intestinal obstruction. Anal atresia or stenosis was present in 36 cases and 29 had a rectourinary fistula.[4]

Pathophysiology of intraluminal meconium enterolithiasis is obscure but prolonged stasis and admixing of urine and meconium have been suggested to be a plausible cause of colonic calcifications in cases with imperforate anus. The presence of fetal urine is believed to bring a change in the calcium phosphate in the meconium and this results in enterolithiasis.[5]

Case Report

A primigravida presented to the Department of Foetal Medicine at Artemis Hospitals in the third trimester for routine fetal well-being ultrasound.

Fetal ultrasound showed dilatation of the entire colon. This was filled with echogenic enteroliths (►**Fig. 1**). On further delineation, echogenic fecoliths were seen floating in the sigmoid colon. When urine comes from the fetal urinary bladder into the sigmoid colon, it makes the fecoliths echogenic and float suggestive of high anal atresia with vesicocolic fistula.

A fistulous connection between the fetal urinary system and sigmoid colon was observed with passage of urine, which made the enterolith echogenic and float in the colon (►**Fig. 2**).

Antenatal ultrasound of the fetus failed to show the "target sign" that is suggestive of high anal atresia which was confirmed by postnatal examination (►**Fig. 3**).

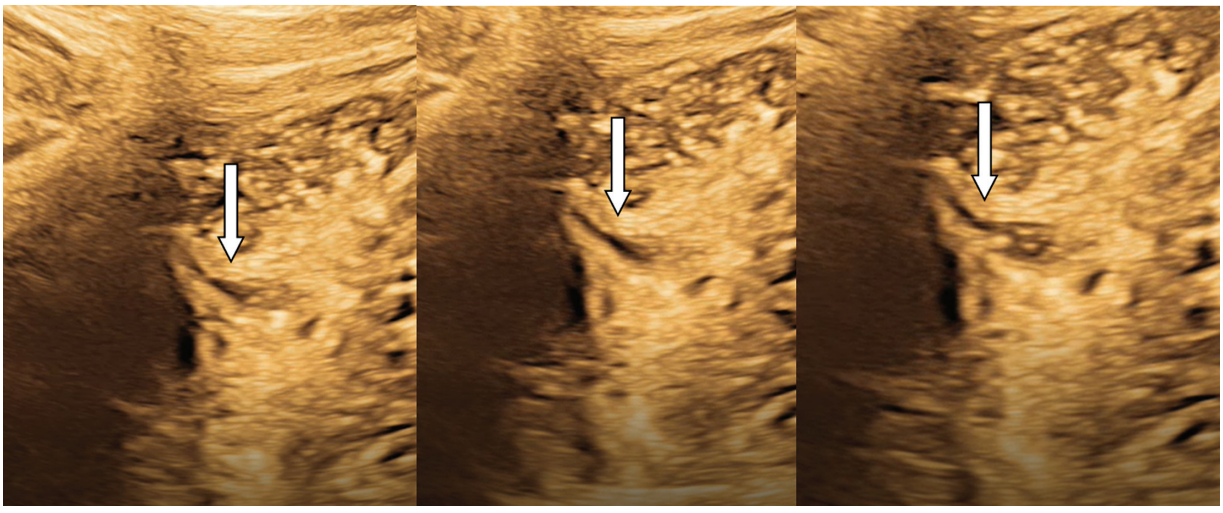

Fig. 2 Ultrasound showing patent fistula with passage of urine through the channel and echogenic enterolith.

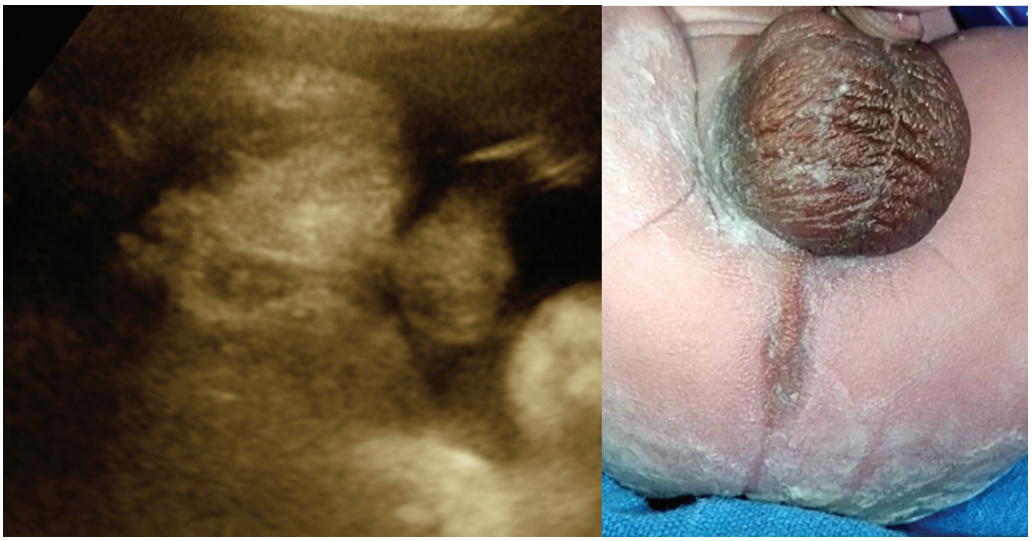

Fig. 3 Antenatal ultrasound with nonapparent "target sign" and postnatal picture showing anal atresia.

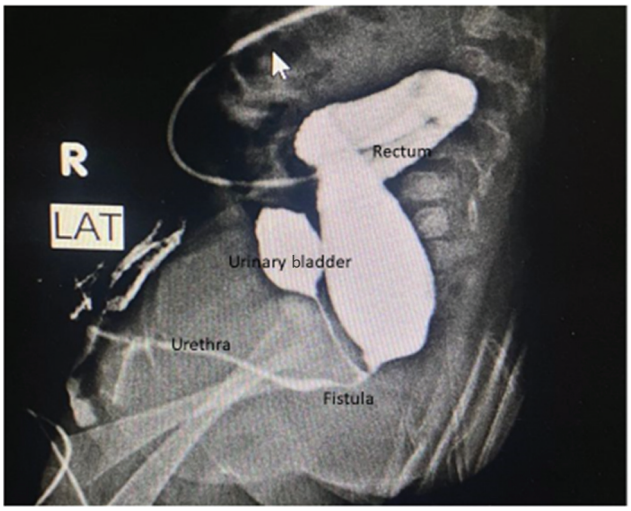

Fig. 4 Postnatal cystourethrogram depicting fistulous track between the colon and urethra.

Postnatal ascending cystourethrogram showed the fistulous tract between the colon and the urethra corroborating the antenatal findings (►**Fig. 4**).

Thus, a fetal vesicocolic fistula was identified prenatally, conformed by meconuria in the immediate postnatal period and further confirmed by cystourethrogram.

Discussion

Prenatal ultrasonography has helped identify an intraluminal calcification and differentiate between enterolithiasis and meconium peritonitis; this was first described by Anderson et al in 1988.[6] The sonographic sign of enterolithiasis is the dilated colonic loops containing calcified foci.[7] Different authors have reported antenatal enterolithiasis-associated malformations and associated fistulas (►**Table 1**).

Anorectal malformation with rectourethral fistula can be managed by primary neonatal reconstruction without

Table 1 Prenatal diagnosis of enterolithiasis and other malformations by different authors

Sl. no.	Reference	Sex	VACTERL	Other anomalies	Fistula
1	Anderson et al[6]	F	AL	URSMS	Rectovesical
2		M	VARL	URSMS	Rectourethral
3	Grant et al[7]	F	AR	Choledochal cyst, imperforate hymen, urachal cyst	No
		F	A	Hydrometrocolpos	Rectourethral
4	Mandell et al[8]	?	VAC	–	Rectourethral
		M	A	–	Rectourethral
5	Simma et al[9]	M	ACR	Splenic duplication	Rectovesical
6	Sepulveda et al[10]	M	ACR	Short small bowel, malrotation of mesentery, absence of penis	–
7	Achiron et al[11]	F	A	–	Vesicovaginorectal
8	Present case	M	A	–	Rectourethral

Abbreviations: A, imperforate anus; C, cardiac anomalies; F, female; L, limb anomalies; M, male; R, renal anomalies; TE, tracheoesophageal fistula; URSMS, urorectal septum malformation sequence; V, vertebral defects.

colostomy. So, to prevent any urethral injury during anorectoplasty, the fistula's location is very important. To date, voiding cystourethrograms are used to determine the presence and location of fistulas in neonates with anorectal malformations. Ultrasounds can accurately detect but cannot precisely locate the fistulas in neonates with anorectal malformation. When primary neonatal reconstruction of anorectal malformation without colostomy is planned, voiding cystourethrogram provides the precise information required about fistula location.[11]

Fetal extraluminal calcifications are commonly reported and are usually the result of intrauterine intestinal perforation with intraperitoneal extravasation of meconium due to intestinal obstruction and/or atresias. Intraluminal calcification of meconium is rare and results from the admixing of stagnant urine and meconium in utero. Intraluminal calcifications in dilated loops of intestine with associated urinary tract abnormality are highly suggestive of a rectourinary fistula.

Mandell et al[8] reported two cases of imperforate anus with rectourinary fistulae diagnosed antenatally.[8] Anorectal malformations are rare to occur and even rarer to be diagnosed antenatally. It is suspected if the distal bowel appears dilated and more so if the intraluminal contents are echogenic. Correia et al reported a case with dilated sigmoid in the first trimester and echogenic intraluminal calcifications in the second trimester with vesicorectal fistula.[13]

Ours is the fourth case of rectovesical fistula with high anal atresia with echogenic and floating enterolith identified prenatally.

Conflict of Interest

None declared.

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