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Case Report

Prenatal Diagnosis and Management of Isolated Recto-Urethral Prostatic Fistula: A Case Report

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ABSTRACT

Background: Recto-urethral prostatic fistula (RUPF) is a rare form of anorectal malformation (ARM). Its prenatal diagnosis and management with a minimum consequence are challenging. This study aimed to present diagnostic and therapeutic modalities in a patient with RUPF.

Case report: A 32-year-old pregnant woman with no relevant medical or surgical history at 27 weeks of gestation was referred to our department of pediatric surgery. Prenatal ultrasound showed loop dilatations and enterolithiasis. Fetal magnetic resonance imaging (MRI) imaging confirmed the diagnosis of ARM and suggested the presence of a recto-urinary fistula. There was no other associated malformation. Parents decided on the continuation of pregnancy after counseling. A 2300 g male was born at 37 weeks of gestation in February 2019. Colostomy followed by laparoscopic pull-through were performed. Expectations of the physician and parents were met after a one-year follow-up period.

Conclusion: Fetal MRI had the potential to diagnose ARM more accurately than ultrasound. Moreover, laparoscopic pull-through was safe and feasible.

Keywords: Anorectal malformations, MRI, Prenatal diagnosis, Therapeutics, Ultrasound

Introduction

Anorectal malformation (ARM) is a rare condition among newborns, and more serious malformations result in higher numbers of associated defects. Many of these associated anomalies may be seen in the utero, such as renal anomalies, esophageal atresia, skeletal lesions, and cardiac malformations. Prenatal diagnosis of the ARM is difficult and its rate is within the range of 16-42% among the cases. Recto-urinary fistula (bladder and urethra) presents less than 25% of ARM. Isolated RUPF is uncommon and prenatal diagnosis is challenging (1-4). There are controversies concerning laparoscopic-assisted pull-through (LAPT) or posterior sagittal pull-through in the case of RUPF (1,2, 5, 6). This study presents diagnostic and therapeutic modalities in a patient with RUPF. It should be mentioned that written informed consent of parents was obtained.

Case report

A 32-year-old woman, gravida 1, para1, with no relevant medical or surgical history, was referred to our department of pediatric surgery at Hedi Chaker Hospital in Sfax, Tunisia. Ultrasound performed at 27 weeks of gestation (WG) showed dilated bowel with intraluminal hyperechoic structures (Figure 1A). It should be mentioned that no other abnormalities were found. The T1 and T2 weighted images obtained through magnetic resonance imaging (MRI) were needed to confirm the rectal obstruction at 28 WG. The fetal rectum and sigmoid were dilated to 13 and 20 mm, respectively, and there was a heterogeneous fluid signal on T2 weighted images (WI) (Figure 1B).

Prenatal diagnosis made it possible to consult the couple, inform them about the prognosis of the detected anomalies, and arrange for delivery and

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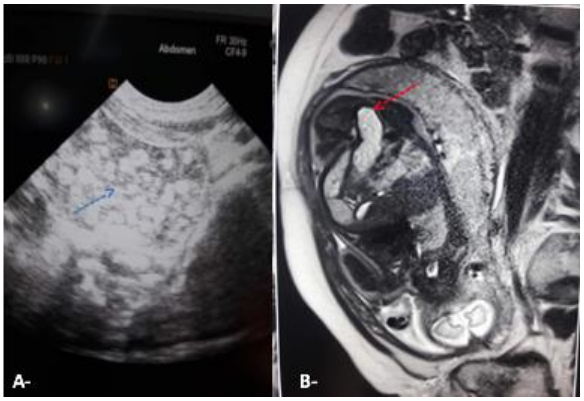

Figure 1. A- Fetal ultrasound showed enterolithiasis (→) **B-** MRI showed a dilated rectum and sigmoid and abnormal hyperintense fluid signal on a sagittal T2-weighted image

then surgery in Hedi Chaker Hospital. This hospital is a tertiary care unit located in Tunisia which is a North-African developing country.

A 2300 g male neonate was born at 37 WG in February 2019. Physical examination revealed an imperforated anus, plate perineum, and a distended abdomen. Nasogastric tube ruled out associated esophageal atresia. The cross-table lateral film taken after 24 h of life showed distal gas high in the pelvis. A sigmoid colostomy was performed and the urine came from the distal end of the colostomy. The distal colostogram showed a large recto-urethral fistula.

At the age of six months, he had an LAPT, and a urinary catheter was inserted into the bladder. The operation was performed using a homemade glove port and straight instruments inserted through a single umbilical incision (7). Rectal mobilization was performed after minimal dissection of the posterior wall of the rectum using hook cautery. The rectal wall was separated from the urethra, and the fistula was closed by clips. The center of the sphincter was determined with the use of electric stimulation; a trocar was passed under direct vision into the presumed place of the anus to dilate the route for rectal pull-through. Under the guidance of laparoscopic visualization, the distal rectum was grasped and pulled down (Figure 2).

The postoperative course was simple. The urinary catheter was maintained in place for five days. It should be noted that the follow-up period after closure of colostomy is 13 months. The case experience no recurrence of fistula and no mucosal prolapse or perianal erosion. It is noteworthy that there was no need for a laxative. Renal ultrasound was normal, and the Parents and surgeon were satisfied with the short-term results.

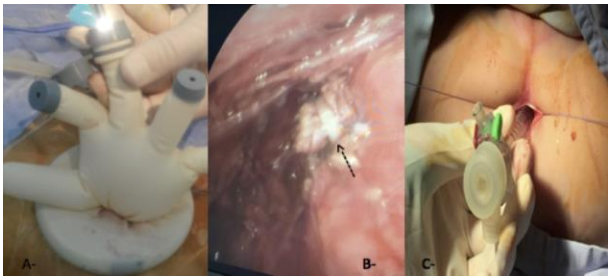

Figure 2. A- Homemade glove port inserted through the umbilical incision **B-** intra-operative view showing the fistula closed using clips (→) C- Center of the sphincter was determined with the use of electric stimulation, then a trocar was passed under direct vision into the presumed place of the anus to dilate the route for rectal pull-through

Discussion

The present report demonstrated a case of isolated RUPF diagnosed by fetal MRI and treated using LAPT in a North-African developing country. It is worth mentioning that this anorectal malformation is rare. Opinions vary about the most useful imaging study and the appropriate sequence of performance in the case of ARM. The sensibility and specificity of ultrasound and MRI to diagnose this rare ARM during pregnancy are controversial (1-4). The ability to identify and characterize ARM on prenatal imaging is important since it may give future parents information about the type of anomaly and associated malformations, increase their psychological compliance, and allow them to plan delivery and surgery in a specialized center or request the medical interruption of pregnancy.

Prenatal diagnosis of ARM is based on indirect signs, such as the U or V segment of dilated bowel, presence of hypoechoic intra-abdominal cystic mass, presence of intraluminal enterolithiasis, and the absence of the anal ring after its maturation (i.e., 30 weeks of gestation). Presence of the anal ring may rule out the presence of anal atresia. These signs are not constant and may not be specific to ARM (3, 8, 9).

In this case, elements that suggested the diagnosis of ARM were the dilated loops on ultrasound and the presence of enterolithiasis. The anal ring, in this case, was not described. Enterolithiasis is an extremely rare sonographic sign. In the most comprehensive case series summarizing 20 cases of prenatal diagnosis of enterolithiasis, 14 cases were attributed to the anorectal malformation spectrum (10). Enterolithiasis presents in ultrasound with the pathognomonic sign of singular or multiple intraluminal hyperechogenic structures within a dilated bowel loop. It must be distinguished from

meconium peritonitis where hyperechogenic structures are extraluminal.

The physiopathology of enterolithiasis is poorly understood. It may be explained by digestive enzymes or a mixture of the calcium-phosphate present in the urine and meconium. Based on previous studies, the majority of cases with enterolithiasis had recto-urinary fistula (9-11). The case presented in this study supports the hypothesis that the prenatal diagnosis of enterolithiasis raises the probability of the presence of a recto-urinary fistula. Enterolithiasis has been the first sonographic feature that led to fetal MRI in the largest case series of enterolithiasis (10).

The MRI is not frequently used during the prenatal period and indications are yet to be established for fetuses (3, 8, 9, 12). In a normal fetus, urine has a homogeneous hyperintensity and is hypointense on T2 and T1 WI, respectively. The normal meconium appears hyperintense on T1 WI and hypointense on T2 WI. Fetuses with ARM may mainly have a recto-urinary fistula (in males), increased signal intensity in the rectum, and decreased signal intensity in the bladder (4,12). The normal maximum distal colon diameter increases with gestational age from approximately 8 mm at 24 weeks to approximately 16 mm at 35-38 weeks. The rectum should be closely apposed to the bladder regardless of gender, and it should extend at least 10 mm below the bladder neck (12).

In our case, there was a heterogeneous signal on T2 WI in the rectum and bladder as a result of the mixing of urine and meconium. It should be mentioned that the distal colon was dilated, and the rectum was located higher than the bladder base. These findings allowed the evaluation of the anomaly with greater accuracy than ultrasound. The limitation of MRI is that fetal MRI for ARM should be performed at 20 weeks of gestational age. Before this time, the distribution of meconium cannot be accurately defined within the colon and rectum. Another limitation of MRI is the lack of availability in some centers.

The treatment and prognosis depend on the type of malformation and associated anomalies. The surgeon may perform a primary repair or colostomy. An ARM with a recto-urinary fistula is a common indication for colostomy [1]. For the second step of surgical repair, some surgeons perform posterior sagittal anorectoplasty (PSARP) while others prefer laparoscopic-assisted LAPT. The LAPT for high imperforate anus was first reported by Georgeson et al. in 2000 (13).

Recent publications have focused on the comparison between laparoscopy and the PSAR; however, the number of patients in the studied groups was limited (1, 5, 12-15). Bischoff et al. stated that laparoscopy is an option to replace a necessary laparotomy in 10% of male patients. According to them, the ideal indications for laparoscopy were the recto-bladderneck fistulas and some selected recto-urethral prostatic fistulas. In male patients, the LAPT would better be contra-indicated in the recto-perineal fistula, recto-vestibular fistula, imperforate anus without fistula, and recto urethral bulbar fistula, as it risks injury to the urethra or formation of a posterior urethral diverticulum (1, 5).

A recent meta-analysis showed that LAPT is a safer and more effective surgical procedure, compared to PSARP for the treatment of high/intermediate anorectal malformations (6). Son et al. found that minimal laparoscopic dissection of the posterior wall of the rectum, similar to what was performed on our case, may reduce the prevalent rectal prolapse (16%) after the PSAR (16, 17). Tran QA et al. evaluated the outcomes of LAPT for high-type ARM. They concluded that LAPT was effective on a low rate of complications without a urethral fistula or diverticulum during a mean follow-up period of 71.5 months (17).

To the best of our knowledge, there is no series comparing the results of PSARP and LAPT for this particular anatomic form. In our case of RUPF, LAPT allowed good visualization and safe closure of the fistula. Moreover, it allowed a precise pull-through of the rectal pouch within the center of the complex. The LAPT also avoided denervation injury to the muscle complex and division of the anal sphincter. Postoperative outcomes and cosmetic results were satisfactory.

Limitations of the study are the short follow-up period since a long-term follow-up is needed. In fact, the risk of postoperative anal incontinence or chronic constipation is important. All patients with ARM must be followed to be sure that the kidney function does not deteriorate over time. In addition, their anorectal function should be assessed as well. The LAPT was safe for this single case and large series are still recommended to confirm the feasibility and benefits of LAPT in the management of patients with RUPF.

In conclusion, the use of MRI in the second and third trimester of pregnancy combined with ultrasound may help to identify the anomaly. It also allows realistic estimation of the prognosis, better prenatal counseling, and planning postnatal management. Till now, there has been no evidence

regarding the optimal method of ligation and division of the fistula and accurate creation of the pull-through canal. The LAPT with minimal dissection of the posterior rectal wall was a safe and feasible procedure in this case. The LAPT is still evolving and further technical refinement and further large studies are needed to achieve better outcomes for RUPF.

Conclusion

Fetal MRI had the potential to diagnose ARM more accurately than ultrasound. Moreover, laparoscopic pull-through was safe and feasible.

Acknowledgments

None.

Conflicts of interest

None.

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