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Case report

A case report: Common channel anomaly with vaginal agenesis and rectal stone after posterior sagittal anorectoplasty (PSARP)

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ABSTRACT

Introduction and importance: We reported a case of common channel anomaly complicated with vaginal agenesis and rectal stone, which is a long-term complication of PSARP, a combination of very rare conditions with high morbidity and especially difficult treatment.

Presentation of case: A 15-year-old female presented with a chief complaint of cyclic abdominal pain. The patient had no history of menstruation before. Physical examination showed a common channel. The diameter of the common channel was approximately 1 cm and 0.3 cm in length. A sound (±2 mm thickness) was inserted to the small opening between the urethra and anal mucosa with the length of the canal 6 cm. At the end of the opening, a stone-like structure was felt. The management of this case was abdominal hysterectomy with right salpingectomy and stone evacuation.

Discussion: Cloacal malformation is thought to be associated with vaginal agenesis since both malformations are considered to have a similar pathophysiologic background. Treatment can be performed using posterior sagittal combined with laparotomy approach. After surgery, patients will need a long-term follow-up since the conditions may be associated with many possible urologic and gynecologic comorbidities, including recurrent urinary tract infections, hematosalpinx, and vesicourethral reflux, including stone formation.

Conclusion: Our case was considered one of the most complex common channel anomalies because it was complicated with a rare condition called vaginal agenesis and rectal stone after PSARP. A multidiscipline approach was necessary.

1. Introduction and importance

Cloacal malformation is a very rare congenital defect characterized by the urethra, vagina, and rectum fusion into one common channel and orifice. It is considered the most complex anorectal malformation, and its treatment is one of the biggest challenges even to the most experienced multidisciplinary team of physicians. It is estimated to occur in every 50.000 live births [1,2]. Until now, the etiology of cloacal malformation is still unknown [3].

The uncertainties get even more complex when another anomaly may present concurrently with cloacal malformations, one of which is vaginal agenesis. Vaginal agenesis or Mullerian agenesis is a type of congenital anomaly in which there is an absence of the vagina coupled with maldevelopment of the uterus. It was found that vaginal agenesis occurs in every 1 of 5000 live births [4,5]. It may present as a cyclic abdominal pain caused by hematosalpinx, hematometra, hematocolpos, or endometriosis [6].

In this study, we reported a patient with cloacal malformation of common channel anomaly with incidental finding of vaginal agenesis and rectal stone with cyclic abdominal pain as the chief complaint. The rarity of this case with rectal stone makes this case interesting.

This case report has been reported in line with the SCARE 2020 criteria [7].

2. Case presentation

2.1. Patient information

A 15-year-old Indonesian female presented with a chief complaint of cyclic abdominal pain. The patient was a junior high school student and was unmarried. She was referred to our hospital with vaginal agenesis and suspected genital malformation. The cyclic abdominal pain had occurred since 1 year ago and had worsened in recent months.

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2.2. Clinical findings and investigations

General state and vital signs were normal. The patient had a BMI score of 21.4 kg/cm^2 and Tanner stage of M3P2. Her abdominal pain occurred in monthly intervals with a worsened symptom. She had histories of anal surgery and colostomy (PSARP). No abdominal bulge had ever presented during the abdominal pain, and she never had menstruation before. Breast and pubic growth were within normal limits.

She urinated around three times per day and defecated once daily. The patient could not hold watery stools and flatus. However, she could still hold solid feces. The patient was the third child in her family, and there was no familial history of a similar condition.

She had twice surgery procedures in infancy due to anal atresia (PSARP). She had a Mitrofanoff silicone catheter due to bilateral hydronephrosis and megaurethra. The patient consumed analgetic routinely, and she had no history of drug allergy. Physical examination showed minimal pubic hair and asymmetric of the labia minora; a common channel was present with no hymenal ring. The anal mucosal could be seen 3 cm below the urethral meatus with no anal sphincter tone. The diameter of the common channel was approximately 1 cm, and the length was 0.3 cm. A sound (±2 mm thickness) was inserted into the small opening between the urethra and anal mucosal. The canal length was found at approximately 6 cm. At the end of the opening, a stone-like structure was felt (Fig. 1).

2.3. Diagnostic assessment

Ultrasound examination revealed that the uterine cavity was tilted to the right with a rudimentary left side and unicornuate uterus, filled with echoic mass in accordance to hematometra. No vaginal lumen and cervical canal were detected. A normal ovary was seen in the right adnexa. Medially to the right ovary, a cystic mass was present with the size of 6.7 × 3.5 cm, with a normal view in the left adnexa. It was concluded to be unicornuate uterus with rudimentary left side with hematometra and hematosalpinx on the right side of the uterus with cervical and vaginal agenesis (Fig. 2).

MRI examination showed that the uterus was anteflexed, with hematometra and hematosalpinx with cervical and vaginal atresia. Both ovaries were normal in shape and size. A common channel connected with the urethra and small canal mimicking the rectum and the artificial anal canal was not connected to the common channel. The bladder was within normal limits. Rectal stone was seen in the space, which resembled the rectum (Fig. 3).

2.4. Therapeutic intervention and outcomes

Preoperative investigation showed normal laboratory and thorax radiology results. This procedure was performed by an experienced Urogynecologist and Urologist. This case was managed by abdominal hysterectomy and right salpingectomy concurrent with cystoscopy. Cystoscopy was performed to examine the vaginal patency and the connection between the vaginal canal and the uterus. There was no patent vaginal canal and no connection between the small vaginal opening with the uterus on cystoscopy findings. The bladder was within normal limits, and megaurethra was also found. The bladder mucosal was smooth with no sign of hyperemia. In the other space below the external meatus urethra, the cystoscope was inserted, and we found a stone 2 cm at the proximal space with the size of 3 × 3 cm. This space was suspected as the remnant of the rectum before PSARP (Fig. 4).

Histopathology examination with tissue taken from the retrovesical tissue showed rectal tissue lined with single-layered columnar epithelia with goblet cell and chronic inflammation cells of colon space resembling the rectum tissue; no remnant of Mullerian duct was seen. Another tissue sample that was taken from the uterus showed a uterus without a cervix (Fig. 5).

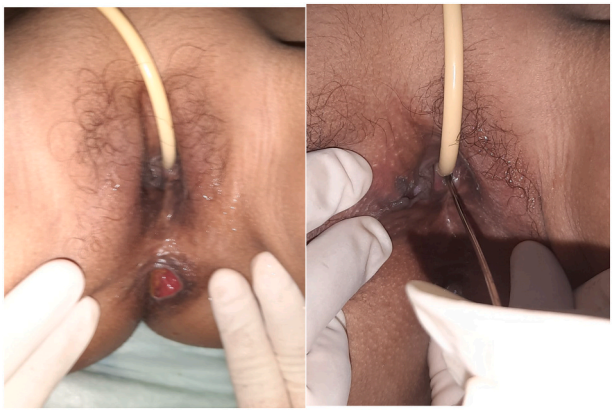

Fig. 1. Physical examination.

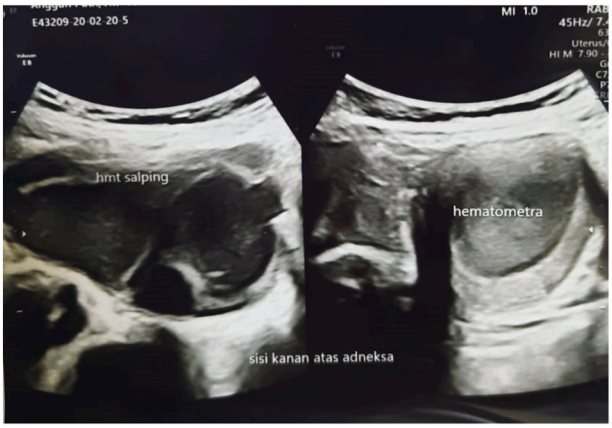

Fig. 2. Ultrasound examination.

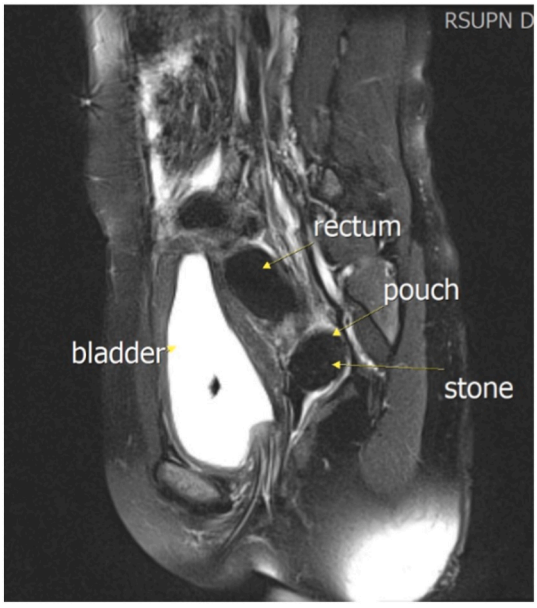

Fig. 3. MRI examination.

2.5. Follow up and outcome

The patient regularly came to the urogynecology and urology

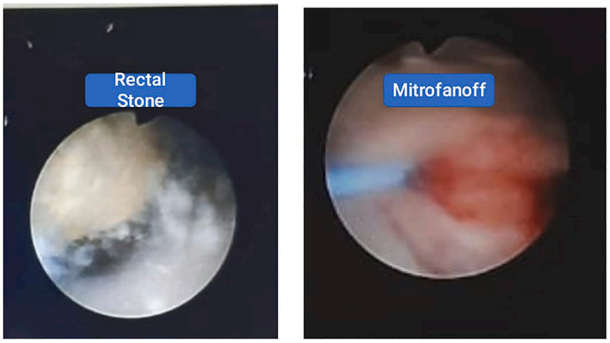

Fig. 4. Cystoscopy view.

outpatient clinic until 1 year after the procedure for follow-up. The patient had been able to perform daily activities properly without cyclic pain. Her Mitrofanoff catheter was still maintained by the urologist. A neovaginal procedure would be performed when the patient plans to get married.

3. Discussion

Cloacal malformation is a severe malformation of the anorectal region thought to be caused by the failure of urogenital sinus division in the early step of embryogenesis. The failure of cloacal region formation resulted in the fusion of several tracts derived from the cloaca and urogenital sinus into a single channel. The cloaca is a perineal structure that presents in 4 weeks old embryo. It will give rise to the urogenital sinus and anorectal sinus divided by the urorectal membrane. The separated structures will then open up in the perineum with each own opening. Arrest in the differentiation and development of the structures will result in cloacal malformations, and the timing of the arrest onset will determine the morphology of cloacal malformation [8,9].

Although the subtypes are diverse, the imaging feature of the cloacal malformation subtypes is quite similar to each other. Cystic mass with an air-fluid level in the pelvic region is a classic finding, and the mass may be septated. On physical examination, meconium accumulation in the vagina can be observed since the urethra, vagina, and anus will all have connected to a single cloacal chamber. The obstruction will hinder urine excretion. However, urine production will increase along with age. The impaired outlet of the urine combined with an increasing rate of urine production will result in the accumulation of urine in the genitourinary tracts, which ultimately causes hydronephrosis, as was seen in our case. At an early age, these changes are often subtle and may manifest only in older age [10,11,12,13].

The severity of the malformation heavily affects the clinical outcome. Patients with cloacal dysgenesis usually do not survive due to renal failure [14,15]. The main goal of cloacal malformation treatment is to relieve the ongoing genitourinary obstruction, reduce the risk of vaginal perforation, and prevent sepsis. The position of the anus is a significant factor affecting the outcome. The presence of a normally positioned anus is a sign of a favorable prognosis since it indicates intact innervation and pectinate line. The common channel length also affects the clinical outcome. A common channel longer than 3 cm is harder to repair and thus is a sign of poor prognosis. Pena et al. found that common channel anomaly patients with lengths longer than 3 cm have a higher risk of having urologic complications. Patients with a common channel can be treated using posterior sagittal combined with a laparotomy approach. Patients with a common channel length greater than 3 cm require a combination of techniques for the treatment to be successful [16]. Diagnostic cystoscopy and vaginoscopy can be done to determine the length of the common channel. After surgery, the patients will need a long-term follow-up since the conditions can be associated with many possible urologic and gynecologic comorbidities, including chronic and recurrent constipation, recurrent urinary tract infections, hematosalpinx, kidney stone formation, infertility, and vesicourethral reflux and stone formation [17].

The cloacal malformation is thought to be associated with vaginal agenesis since both malformations are considered to have a similar pathophysiologic background. In vaginal agenesis, hindered Mullerian duct development causes the female reproductive organ development to be incomplete. In many cases, only a short vagina or small duct-like structure mimicking the vagina can be seen, and the uterus may be rudimentary. The disconnection of the malformed vagina with the uterus is a common finding [18,19,20]. Patients with a congenital cloacal malformation may need to undergo complex reconstruction of the rectourogenital tracts. The current surgical approach for cloacal repair was derived from the posterior sagittal anorectoplasty (PSARP), described by Peña and De Vries. In our case, PSARP was done without vaginal reconstruction in the infancy period. The patient had felt genital complaints after the menarche period. On examination, we found congenital urogenital anomalies with a history of PSARP, hematometra on the unicornuate uterus, and hematosalpinx without cervix and vagina. As for the associated anomalies, we found megaurethra with inserted Mitrofanoff catheter by the urologist in this patient. A rectal stone was found in space resembling a remnant of the rectum before PSARP. We suspected the stone was formed by reflux of urine from vesicourethral into space remnant of the rectum. Unfortunately, we did not perform any laboratory analysis of the stone.

4. Conclusion

To our knowledge, our case was one of the most complex common channel anomaly cases because it was complicated with other rare disorders. In these very rare conditions, a collaboration of highly experienced and specialized urogynecologist, pediatric surgery specialists, and urologist is necessary since the treatment is complex and requires surgical intervention from many different and highly specialized fields.

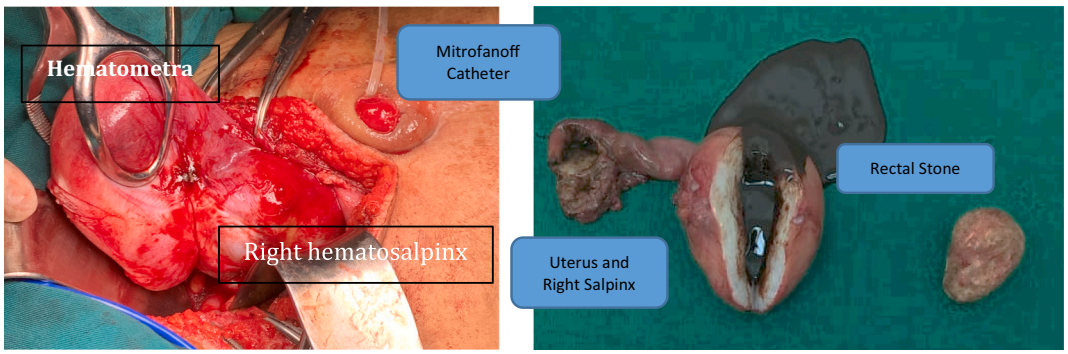

Fig. 5. Hematometra, hematosalpinx, and rectal stone of the patient seen during surgery.

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Ethical approval

Not required.

Informed consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Tyas Priyatini: concept, operator, data analysis, drafting and revising, final approval.

Roziana: data collection, data analysis, writing the paper.

Guarantors: Tyas Priyatini, Roziana.

Declaration of competing interest

None declared.

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