Laporan Kasus

Vaginoplasty and Fimbrioplasty on Vaginal Agenesis with Hematocolpos, Hematometra, and Hematosalpinx

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Abstrak

Agenesis vagina kongenital adalah kelainan langka pada saluran genital wanita yang disebabkan oleh perkembangan embriologis duktus Müllerian. Manifestasi klinis bervariasi tergantung pada luasnya anomali, tingkat keparahan, dan usia pasien saat datang. Dilaporkan seorang gadis berusia 16 tahun yang datang ke Klinik Uroginekologi RSUP Dr. Hasan Sadikin Bandung dengan keluhan utama nyeri perut siklik yang hilang timbul selama 2 tahun terakhir dan belum menstruasi. Pasien dirujuk dari beberapa rumah sakit swasta dengan riwayat laparotomi eksplorasi sebelumnya. Pasien memiliki perkembangan seksual sekunder yang normal, tetapi tidak ditemukan kantong vagina pada pemeriksaan panggul. Pada pasien tersebut ditemukan hematokolpos 10,08x6,80x6,02 cm dan hematometra 14,41x12,92x10,62 cm dari pemeriksaan ultrasonografi. Pada laparotomi eksplorasi, ditemukan hematosalping bilateral dengan uterus normal. Dilakukan tindakan vaginoplasti, salpingektomi parsial bilateral, dan fimbrioplasti bilateral. Dalam laporan kasus ini, penulis berhasil melakukan vaginoplasti dan fimbrioplasti untuk mengevakuasi hematokolpos, hematometra, dan hematosalping. Vaginal molding dimasukkan ke dalam rongga neovagina untuk mencegah vagina kolaps spontan dan dipertahankan in situ selama 2 minggu setelah operasi.

Kata kunci: agenesis vagina, hematokolpos, hematometra, hematosalping

Abstract

Congenital vaginal agenesis is a rare abnormality of the female genital tract caused by abnormal embryological development of the Müllerian ducts. Clinical manifestations vary depending on the extent of the anomaly, severity, and the patient's age at presentation. It has been reported that a 16-year-old presented to the Urogynecology Clinic at Dr. Hasan Sadikin General Hospital, Bandung, with a chief complaint of cyclic abdominal pain that had been intermittent for the past 2 years and no history of menstruation. The patient was referred from several private hospitals with a history of previous exploratory laparotomies. The girl had normal secondary sexual development. No vaginal sac was found on pelvic examination. A hematocolpos measuring 10.08 x 6.80 x 6.02 cm and a hematometra measuring 14.41 x 12.92 x 10.62 cm were found on ultrasound. An exploratory laparotomy revealed bilateral hematosalpinx with a normal uterus. Vaginoplasty, bilateral partial salpingectomy, and bilateral fimbrioplasty were performed. In this case report, the authors successfully performed vaginoplasty and fimbrioplasty to evacuate the hematocolpos, hematometra, and hematosalpinx. Vaginal molding was then inserted into the neovaginal to prevent spontaneous vaginal collapse and maintained in situ for 2 weeks after surgery.

Keywords: *hematocolpos, hematometra, hematosalpinx, vaginal agenesis*

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INTRODUCTION

Congenital vaginal agenesis is a rare anomaly of the female genital tract caused by the failure of formation of the sinovaginal bulb from the fusion of the caudal end of the Mullerian duct system and the urogenital sinus. It occurs in 0.001–0.025% of the population.[1,2] It may be

an isolated anomaly or may occur in association with several other anomalies, such as complete androgen insensitivity syndrome (CAIS) or *Rokitansky-Mayer-Kuster-Hauser syndrome* (MRKH).[1,3] Many classifications for congenital uterovaginal anomalies exist and are useful for guidance in the diagnosis and management of this condition. The American Society of Reproductive Medicine (ASRM) separates the anomalies into seven classes.[4,5]

Clinical manifestations of vaginal agenesis vary, but commonly, the patient comes for treatment due to abdominal pain and no menstruation. Diagnosis and management of vaginal agenesis is challenging. There is still no agreement on the gold standard for vaginal agenesis management. Besides relieving the abdominal pain, the goals of management are preservation of sexual and fertility.[2,5] Based on the American Congress of Obstetricians and Gynecologists, nonsurgical creation of a neovagina is the first-line management of vaginal agenesis. However, the failure rate of this procedure is high.[6-8] Thus, many patients have been assessed and considered for surgery as first-line therapy. The risks of surgery are high, with 30–70% of studies reporting complications such as stricture formation or injury to surrounding structures. Therefore, neovagina creation requires an individualized approach, depending on the severity of vaginal agenesis and patient preference.[6]

We have a case report of distal vaginal agenesis that causes hematocolpos, hematometra, and hematosalpinx. It was associated with primary amenorrhea and cyclic abdominal pain. The diagnosis of vaginal anomaly was based on a careful gynaecological examination and ultrasound examination. In this case, due to the abdominal pain worsening, surgical treatment was carried out as early as possible. We performed vaginoplasty and partial bilateral salpingectomy to evacuate hematocolpos, hematometra, and hematosalpinx. In addition, we performed bilateral fimbrioplasty to preserve fertility.

CASE

A 16-year-old presented in our Urogynecology Clinic with the chief complaints of on-and-off cyclical abdominal pain for the last 2 years and absence of menses. The patient was referred to us from a private hospital with a history of an exploratory laparotomy performed before. The patient reported that she has never had sex in her life.

The clinical examination found normal secondary sexual development (Tanner stage 3). A convex abdomen with tenderness in the hypogastric area was found. The vulva was grossly normal with pubic hair, clitoris, urethral opening and vaginal dimple (Figure 1). A digital rectal examination revealed good sphincter tone; a tender mass was palpable. We found a hematocolpos measuring 10.08 x 6.80 x 6.02 cm (Figure 2) and a hematometra measuring 14.41 x 12.92 x 10.62 cm on ultrasound (Figure 3). The transperineal ultrasound showed the distance between the probe and the end of the hematocolpos to be 1.68 –3.38 cm. Hematosalpinx was difficult to assess by abdominal ultrasound. The patient was diagnosed with 1/3 distal vaginal agenesis.

The patient was planned to undergo abdominoperineal repair after taking proper consent and explaining to the guardian of the patient regarding the possibility of hysterectomy. The patient was cleaned and draped in the lithotomy position under general anesthesia. We performed a reassessment in the operating room; no vaginal pouch was found.

Firstly, we performed vaginoplasty by clamping the anteroposterior part of the vagina, then incising horizontally. Using a Sims speculum, an incision was made at the proximal end of the vagina for 1 cm, then bluntly expanded to the side and inward, about $\pm$1000 cc of reddish-brown thick blood came out. Using Kocher clamps, the proximal anteroposterior vagina is directed and pulled distally. We performed interrupted suturing with PGA 2.0 suture between the anteroposterior vaginal mucosa and the labia minora. The mold was used with a silicon catheter into the neovagina and was kept *in situ* for 2 weeks after surgery.

On exploratory laparotomy, the patient was found to have bilateral hematosalpinx with grossly normal uterus and ovaries. The right salpinx increased with size 5 x 4 x 1 cm and the left salpinx increased with size 4 x 3 x 1 cm (Figure 4). We performed a partial bilateral salpingectomy to evacuate hematosalpinx. To preserve fertility, we also performed fimbrioplasty.

DISCUSSION

Vaginal agenesis is a rare case and the prevalence rate in Indonesia has not been reported. It commonly results as a part of a complex anomaly or an isolated developmental defect. Clinical manifestation is commonly cyclic pelvic pain due to menstrual outflow obstruction.[3] Since this patient has normal breast development, the patient typically presents in adolescence with primary amenorrhea. A detailed investigation is required to diagnose and differentiate this condition from other causes of primary amenorrhea.

In a clinical setting, distal vaginal agenesis is identical to an absent vaginal pouch.[3,4] This might be thought of as a lower transverse vaginal septum. In a transverse vaginal septum, the septum is usually no more than 1 cm thick. It can completely obstruct the vagina and can cause hematocolpos (accumulation of menstrual blood in the vaginal cavity) and hematometra (accumulation of menstrual blood in the uterus).[5,9] Similar to a transverse vaginal septum, it can present a diagnostic challenge, as the symptoms are often not typical, including lower abdominal pain, low back pain, chronic constipation, or urinary retention.

An obstructive genital anomaly should be routinely sought in all young women with primary amenorrhea. Another differential diagnosis is an imperforate hymen, which can be easily distinguished from a transverse vaginal septum on physical examination. The bluish appearance with a bulge is observed between the lips and slight pressure applied suprapubically causes visible distension of the imperforated hymen.[9]

Since most young women do not engage in sexual activities in Indonesia. Therefore, to assess the size of hematocolpos in more detail and to evaluate its etiology, abdominal, transperineal, or transrectal ultrasound, or MRI is often used before attempting management.[5] Magnetic resonance imaging (MRI) is an ideal modality for evaluation of Mullerian duct anomaly due to its high accuracy and detailed delineation of uterovaginal anatomy, an accuracy of up to 100% in the evaluation of Mullerian duct anomaly.[10]

The best management for vaginal agenesis is still controversial.[3] The goals of vaginal agenesis management are to relieve the abdominal pain due to The obstructed outflow tract, sexual function, and fertility but it vary according to each individual. In this case, due to the diagnosis of 1/3 distal vaginal agenesis, we performed vaginoplasty to evacuate hematocolpos and hematometra. It continued by vaginal dilation post-operatively to prevent vaginal stenosis. We made an incision at the proximal end of the vagina for 1 cm, then bluntly expanded to the side and inward.

Surgical vaginoplasties can be categorized into four main types: creation of a perineal pouch, lining a neovaginal space, laparoscopic procedures, and intestinal vaginoplasty.[4] Linder BJ et al. reported the success of vaginoplasty with a split-thickness skin graft in a 24-year-old patient with vaginal agenesis.[11] In Indonesia, Moegni *et al.* (2021) reported secondary pyosalpinx eleven days after performing amniotic graft neovagina surgery due to vaginal agenesis with bilateral hematosalpinx in a 12-year-old teenager. Then, adhesiolysis, bilateral salpingectomy, and omentectomy were performed to evacuate bilateral pyosalpinx.[12] Therefore, in this case, we performed partial bilateral salpingectomy to evacuate hematosalpinx and to prevent secondary pyosalpinx.

To preserve fertility, also performed fimbrioplasty. No literature explained the success rate of fimbrioplasty in vaginal agenesis. However, fimbrioplasty had been performed, in addition to vaginoplasty with a modified Singapore flap to evacuate hematocolpometra.[2] Fimbrioplasty and neosalpingostomy had a good outcome, with the pregnancy rate of these treatments being 28.4% in distal tubal obstruction. In this case, it was a young woman who was not sexually active. Follow-up regarding postoperative neovaginal, counseling about the use of vaginal dilators, and further discussion about the fertility plan were needed.

A success of customized vaginal stent use after vaginal agenesis surgery. The 2-year follow-up revealed that the use of the single, customized stent was cost-effective, increased adaptability, duration of wear, and the patient's acceptance of the treatment.[13,14] Of the various types of vaginal agenesis surgery, it has not yet been decided which type of surgery has a better outcome and fewer complications.[15]

CONCLUSION

Vaginal agenesis is one of Mullerian anomalies for which the prevalence rate in Indonesia has not been reported. It commonly results as a part of complex anomalies or as an isolated developmental defect. Clinical manifestations vary and must be carefully assessed to differentiate from other vaginal anomalies. There is still no agreement on the gold standard for vaginal agenesis management. We successfully performed vaginoplasty and partial bilateral salpingectomy in evacuating the hematocolpos, hematometra, and hematosalpinx. Additionally, fimbrioplasty is used for fertility preservation.

Figure 1. No vaginal pouch was found

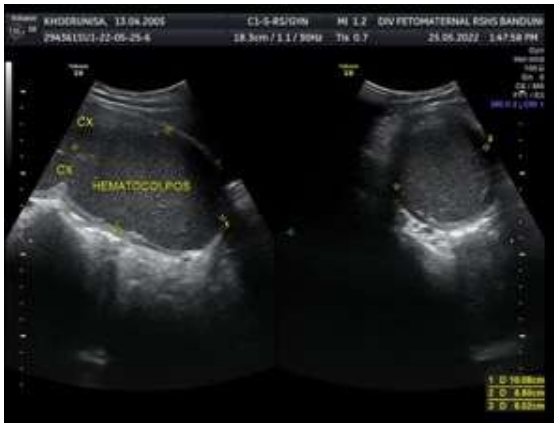

Figure 2. Hematocolpos seen on ultrasound

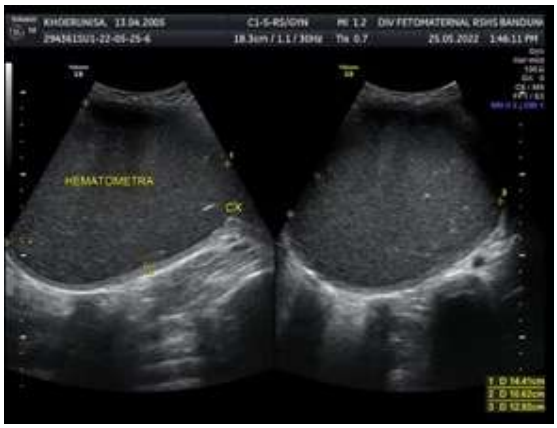

Figure 3. Hematometra seen on ultrasound

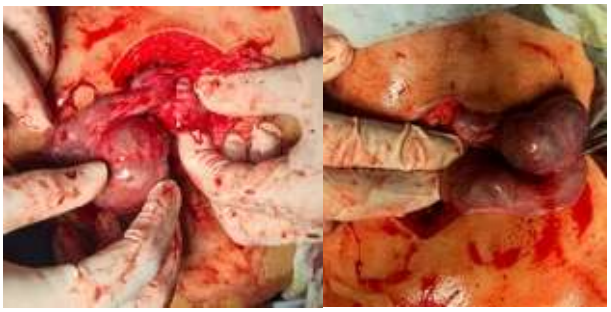

Figure 4. Bilateral hematosalpinx with normal ovaries

INFORMED CONSENT

Written informed consent for the publication of this case report and its accompanying images was obtained from the patient.

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