

CLINICAL CASE

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Neovagina formation, Vecchietti technique, in cases of vaginal agenesis due to Rokitanski syndrome

Formación de neovagina, técnica Vecchietti, en agenesis vaginal por síndrome de Rokitanski

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ABSTRACT

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is a rare congenital anomaly of the genital tract characterized by agenesis of the uterus and the upper portion of the vagina, which may present with various residual structures, but with normal sexual characteristics and karyotype. It has a multifactorial etiology with a strong genetic component. It is often suspected in cases of primary amenorrhea, and magnetic resonance imaging (MRI) is essential for diagnosis. Treatment involves both non-surgical and surgical techniques. We present the case of a 29-year-old female patient diagnosed with MRKH syndrome who underwent a hysterectomy followed by creation of a neovagina using the open Vecchietti technique.

Keywords: Uterine diseases, Hematometra.

RESUMEN

El síndrome de Mayer-Rokitansky-Küster-Hauser (MRKH) es una rara anomalía congénita del tracto genital, caracterizada por la agenesia de útero y porción superior de vagina que puede presentar distintas variaciones de remanentes, pero con caracteres sexuales y cariotipo normal. Tiene una etiología multifactorial, con un fuerte componente genético. Se suele sospechar en un cuadro de amenorrea primaria y para el diagnóstico es indispensable la resonancia magnética. El tratamiento se basa en técnicas no quirúrgicas y quirúrgicas. Se presenta el caso de una paciente de 29 años de edad con diagnóstico de síndrome de MRKH a quien se le realizó histerectomía más una neovagina con técnica de Vecchietti abierta.

Palabras Clave: Enfermedades uterinas, Hematómetra.

INTRODUCCIÓN

Mayer-Rokitansky-Küster-Hauser syndrome, also known as Müllerian agenesis or vaginal agenesis, is a rare congenital anomaly of the genital tract. Its incidence is approximately 1 in 4,500–5,000 women⁽¹⁾. It is characterized by agenesis of the uterus and the upper two-thirds of the vagina, but the presentation can range from the complete absence of these two structures to the presence of uterine remnants with or without endometrial tissue, and the lower third of the vagina, which may consist of simply a small depression between the labia or may be up to 5 to 6 cm in length⁽²⁾. It is also characterized by the normal physiological development of secondary sexual characteristics and a normal female karyotype of 46 XX⁽³⁾. Two clinical forms are distinguished: The typical form, or Type I, also known as Müllerian agenesis⁽⁴⁾, is characterized by the absence of a vagina, a uterus represented by symmetrical uterine remnants, and normal fallopian tubes and ovaries. The atypical form, or Type II, in which the uterine rudiments are asymmetrical or absent; there is hypoplasia or aplasia of one or both fallopian tubes; and it is frequently associated with renal and skeletal anomalies⁽⁵⁾. The latter form is the least common, with an incidence of approximately 30%⁽⁶⁾.

The etiology of MRKH syndrome remains elusive; it appears to be

a multifactorial disorder; however, increasing reports of familial clustering point toward genetic causes, and the use of various genomic techniques has allowed for the identification of promising recurrent genetic abnormalities in some patients^(7, 8, 9).

MRKH syndrome typically presents as primary amenorrhea in an adolescent female with normal external genitalia and normal pubertal development. Both the ovaries and their function are normal. In 2–7% of cases, a uterus (rudimentary or obstructed) with a functioning endometrium is present. Consequently, patients may experience symptoms such as cyclic pelvic pain⁽¹⁰⁾.

Magnetic resonance imaging is considered the gold standard for diagnosis; it is noninvasive and superior to computed tomography and ultrasound⁽¹¹⁾.

The American Society for Reproductive Medicine offers three treatment options: medical management, mechanical dilation, and surgical treatment⁽⁴⁾. The American College of Obstetricians and Gynecologists (ACOG) recommends mechanical vaginal dilation as the first-line treatment, although patient compliance may be low. Surgical procedures include vaginoplasties that use various autografts, such as the McIndoe vaginoplasty (skin graft), the Baldwin vaginoplasty (intestinal graft), the Davydov vaginoplasty (peritoneal graft), and the Williams vulvovaginoplasty (labia majora flaps). More recently, vaginoplasties using cultured autologous vulvar tissue and tissue-engineered biomaterials have been suggested⁽¹²⁾. An alternative surgical procedure is Vecchietti's laparoscopic vaginoplasty, initially described as an open procedure; its primary objective is to create a neovagina by gradually stretching the patient's vaginal cutis. This involves inserting an olive-shaped instrument into the vaginal fornix, which is connected to nylon traction threads that, after passing through the pelvic peritoneum, emerge through the abdomen and are subsequently secured to a progressive traction device. The operation is performed laparoscopically or via laparotomy. Once in place, the nylon threads are gently pulled before being connected to a traction device placed on the abdomen; the threads are set to progressive traction, ap-

proximately 1 cm per day, for 7 days. On the seventh day, the Vecchietti device is removed, and the woman must use vaginal dilators for 30 minutes a day to maintain vaginal length⁽¹³⁾.

CASE PRESENTATION

A 29-year-old woman referred from the city of Caraz due to chronic pelvic pain and a "vaginal septum," with no prior additional tests. During the medical history interview, it was noted that the pelvic pain occurs cyclically, approximately every 30 days. She also reported primary amenorrhea and painful sexual intercourse, for which she was admitted to the hospital for evaluation. The patient reported that she underwent surgery 2 years ago for a right ovarian cyst; no further details were provided.

Physical examination revealed external genitalia with apparently normal sexual development, except for a vaginal canal 2 cm in length (Figure 1), and the cervix could not be palpated. Hormone levels were measured (TSH, free T4, prolactin, LH, FSH, estradiol, free testosterone), with results within normal ranges. Karyotype result: 46XX. The abdominal ultrasound showed no abnormalities.

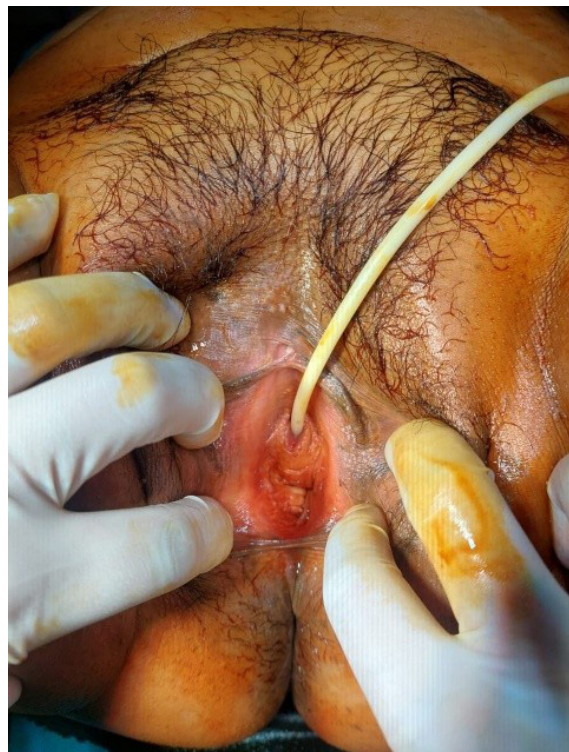


FIGURE 1: NORMAL-LOOKING EXTERNAL GENITALIA. VAGINAL LENGTH OF 2 CM.



The 3D pelvic ultrasound revealed the absence of the upper third of the vagina, as well as the absence of the cervix and isthmus; it also showed an enlarged uterus due to a hematometra. The MRI showed an enlarged uterus, along with a rudimentary cervix and the absence of a vagina (Figure 2).

A diagnosis of MRKH syndrome is considered: cervical agenesis and partial vaginal agenesis; surgical treatment is recommended for the patient due to her chronic pain. In addition, the differences between non-surgical and surgical treatments for vaginal dilation are explained to her, and she agrees to proceed with surgery as the first-line treatment. A laparotomy is performed for hysterectomy and neovaginal reconstruction. Intraoperative findings included an 8x8 cm unicornuate, ovoid uterine remnant enlarged by the presence of blood within it; in addition, a left endometrioma was present, along with the absence of the cervix and the upper two-thirds of the vagina. This was confirmed by vaginally inserting a No. 14 Nelaton catheter and instilling saline solution, which demonstrated the absence of communication. A transverse vaginal septum was ruled out due to the absence of the entire upper vagina and the lack of communication between the uterine remnant and the lower third of the vagina. This uterine remnant was in contact with the upper end of the vesicorectal space. Following the hysterectomy, a neovagina was constructed using the Vec-

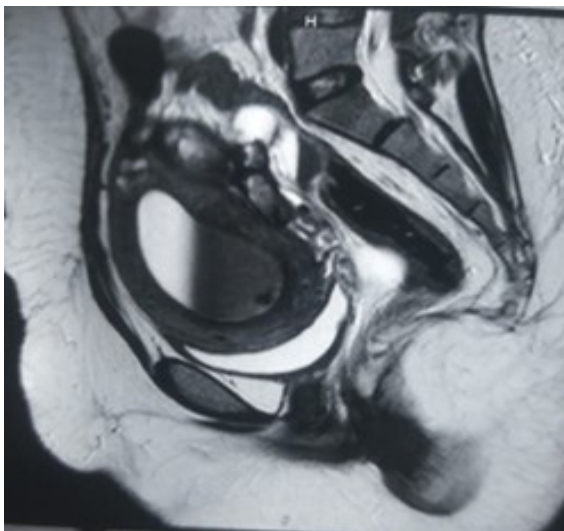


FIGURE 2: MRI. AN ENLARGED UTERUS WITH PROBABLE HEMATOMETRA IS EVIDENT. THE CERVIX AND UPPER VAGINA ARE ABSENT.

chietti technique by placing a sterile olive in the vagina, anchored by two polyglactin sutures, which were inserted vaginally toward the Douglas pouch, traveling subperitoneally along each flank until they emerged through the skin (Figures 3 and 4). These sutures were secured to the abdominal wall, with daily traction of 1 cm applied for 7 days. Upon removal of the sutures, a vaginal length of 6 cm was achieved, and the patient was instructed to continue using a latex vaginal dilator at night for 2 months. During hospitalization, a complication of surgical site infection arose, which was resolved with wound care and antibiotics. The patient was discharged without further complications. The pathology report indicated a remnant of the uterine body with superficial and deep adenomyosis, with no cervix present. Additionally, the left ovary showed hemorrhagic parenchyma and ovarian endometriosis.



FIGURE 3: PLACEMENT OF AN INTRAVAGINAL DEVICE, SECURED WITH A POLYLACTIDE SUTURE.



FIGURE 4: SUTURE EMERGING FROM EACH SIDE OF THE PATIENT (GREEN ARROW)



At the follow-up visit 2 months after surgery, the patient had a vaginal length of 7 cm and resumed her sex life.

DISCUSSION

MRKH syndrome is increasingly recognized as a condition with a complex, multifactorial etiology; furthermore, genetic abnormalities that affect the embryological profile contribute significantly to its development.

In 2024, Matemanosak P, et al., published a retrospective study of 96 patients with MRKH syndrome diagnosed and treated at a tertiary referral hospital in Thailand between 2000 and 2022; they found that 80.3% had Type I and 19.2% had Type II. Furthermore, 21.8% had total vaginal atresia and 78.2% had vaginal hypoplasia.⁽¹⁴⁾

Despite significant advances in reconstructive surgery, there is no consensus regarding the ideal treatment for these patients. Treatment should be aimed at creating a functional vagina.

Although dilation may be chosen as a first-line treatment, as described by Chen N et al.; among 97 patients who underwent vaginal dilation between 2020 and 2023 in China, the success rate was 77% (75 patients), with an increase from 1 to 3 cm before dilation to 6 to 8 cm afterward, associated with the active participation of the sexual partner⁽¹⁵⁾. Surgery may be indicated in cases where nonsurgical therapy has failed, or when the patient prefers it as the initial treatment, as in this case.

When discussing the different treatment options with the patient, it is important to emphasize that there is no quick fix for achieving a functional vagina and that surgical options still require ongoing postoperative dilation—through regular sexual intercourse or vaginal dilators—to ensure a satisfactory long-term outcome.

Yang X, et al., compared the Vecchietti and Davydov techniques in a total of 53 women who underwent surgical neovaginal reconstruction between 2009 and 2019, with a 2-year follow-up. They achieved vaginal lengths of 8.2 cm and 8.5 cm, respectively,

concluding that the Vecchietti technique is a procedure that requires less operating time and may prove to be a relatively simple procedure with much lower operative morbidity⁽¹⁶⁾. The optimal age for this surgery has not been established, but it should be performed once sexual maturity has been reached.

Martens L, et al., published a meta-analysis of 33 studies involving nearly 1,500 patients comparing the Vecchietti and Davydov techniques, finding that 12 months after surgery, vaginal length was 8.7 cm and 8.3 cm, respectively, and that the surgical time was 126 minutes with the Davydov technique compared to 40 minutes with the Vecchietti technique.⁽¹⁷⁾

However, the best surgical option continues to depend on the experience of the surgical team and the patient's preferences. After the creation of the neovagina, patients can lead a normal sexual life⁽¹⁸⁾.

In Peru, there are very few studies on surgical techniques for neovaginal reconstruction, so this case may pave the way for more similar publications. It is worth noting that there are gynecologists with strong surgical skills in hospitals not only in Lima, and this helps decentralize highly complex procedures for the benefit of patients.

ARTICLE INFORMATION

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