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CASE REPORT

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Wharton-Sheares-George vaginoplasty for Müllerian agenesis: A case report

Tonny O. Opar[1*], Johnstone S. Miheso[2], Alfred O. Osoti[1], Eunice M. Njeru[3], Robert I. Mwangi[3]

[1] Department of Obstetrics and Gynecology, University of Nairobi, Nairobi, Kenya
[2] Department of Obstetrics and Gynecology, The Nairobi Hospital, Nairobi, Kenya
[3] Department of Obstetrics and Gynecology, Mater Misericordiae Hospital, Nairobi, Kenya

***Correspondence**: tonnyopar91@gmail.com

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Abstract

Background: Müllerian agenesis is a rare embryological disorder that affects 1 in 4500-5000 female livebirths. It may result in anomalies of the uterus, vagina, or both necessitating surgical management.

Case presentation: A 23-year-old nulliparous presented to the gynecological ward with amenorrhea, severe lower abdominal pain, and bloating. Physical examination revealed well-developed breasts in Tanner stage v. The vagina was noted as a blind-ending dimple with a patent urethral opening. Pelvic magnetic resonance imaging revealed hematometra in the rudimentary right uterine horn. She was scheduled for diagnostic laparoscopy and Wharton-Sheares-George vaginoplasty. A review four weeks later revealed vaginal stenosis, which was managed laparoscopically, with the excision of the rudimentary uterine horn. Her postoperative recovery was uneventful. Her external genitalia were normal, and the vaginal mucosa was typical, with a depth of 6cm and she is currently using Syracuse vaginal dilators and on long-term follow-up.

Conclusion: Wharton-Sheares-George vaginoplasty is a minimally invasive, quick, reliable, and safe procedure for patients with Müllerian agenesis requiring vaginoplasty

Keywords: amenorrhea, Müllerian agenesis, neovagina, Wharton-Sheares-George vaginoplasty

Introduction

Müllerian agenesis or Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is a condition that results from embryological underdevelopment of the Müllerian duct, with resultant agenesis or atresia of the vagina, uterus, or both (1). It affects 1 in 4500- 5000 female livebirths (2). Müllerian agenesis may occur with other congenital anomalies, mainly those of the urinary tract and the skeletal (3). Primary amenorrhea is a common clinical presentation; however, pubertal development and growth are normal (4). Patients may also present with chronic cyclic pelvic and abdominal pain, and pelvic mass may develop due to obstructed outflow tract (5). Management entails psychosocial support, nonsurgical, or surgical treatment of the anatomical anomalies (6). Various neovagina creation methods have been used; however, there is no consensus (7). This is a case of a 23-year-old nulliparous with primary amenorrhea secondary to Müllerian agenesis that was managed using Wharton-Sheares-George vaginoplasty.

Case presentation

A 23-year-old nulliparous presented to the gynecological ward with amenorrhea, severe lower abdominal pain, and associated pressure pain. The pain radiated to the back and was associated with nausea and lack of appetite. The initial occurrence of the lower abdominal pain was ten years prior when she also presented with vomiting. A pelvic ultrasound revealed hematometra, and she was put on analgesics. A year later, she presented with debilitating lower abdominal pain associated with nausea and back pain, during which diagnostic laparoscopy and drainage of hematometra were performed to relieve the symptoms. She was put on the combined pill, which was later changed to the injectable contraceptive, Depo-Provera a few months before the current presentation. However, she stopped the injections due to severe side effects (shortness of breath and general body malaise). She had a childhood history of asthma, for which she was on budecort and montelukast. She was allergic to nonsteroidal antiinflammatory drugs and was on amitriptyline for depression.

On admission, she was generally fair, with typical vital signs. She had normal and well-developed secondary sexual characteristics in the Tanner stage v. An abdominal examination revealed a tender lower abdomen but no obvious masses. A perineal examination revealed well-developed external genitalia and a blind-ending vagina dimple with a patent urethral opening (**Figure 1**). A digital rectal examination revealed a 4-5cm tender mass. An impression of Müllerian agenesis with hematometra was made. Her karyotype was 46, XX. Pelvic magnetic resonance imaging revealed hematometra in the right rudimentary horn, a 1.7 x 1.8cm right adnexal endometrioma with moderate free fluid in the pouch of Douglas. Her preoperative full hemogram, urea, electrolytes, and creatinine tests were within normal reference ranges. She was counseled and scheduled for examination under anesthesia, diagnostic laparoscopy with or without reconstruction of the vagina, and drainage of the hematometra by a multidisciplinary team. The patient was made aware that the primary aim at this initial stage was to relieve her pain by draining the hematometra, but attempts would be made to create neovagina if feasible. She was also made aware that natural birth would not be possible, but intercourse would be achieved after the creation of neovagina. In vitro fertilization, surrogacy, and adoption options were discussed. She was keen to keep the rudimentary uterine horn.

On laparoscopy, the mesonephric duct was revealed; the ovaries were normal, and bilateral vestigial tubes were observed. The right side of the uterus was missing but with a larger left side (**Figure 2**). A right simple cyst and an endometrioma of 5cm were found (**Figure 3**). Bilateral dilatation of the mesonephric duct was done, starting from Hegar dilator sizes 2.5 to 14. The septum between Hegar dilators was then incised using a blunt and sharp dissection to the uterine horn. A colpotomy was performed, the lower part of the uterine horn was transfixed, and dilators were used to drain the hematocolpos. Laparoscopy was repeated to assess for any complications. The vagina was packed with a vaginal mold (**Figure 4**). Postoperatively, she was put on antibiotics, analgesics, antacids, stool softeners, and hematinics. She was discharged on the seventh postoperative day and advised on vaginal mold insertion nightly for three months after applying xylocaine and estrogen cream to the mold. A clinical review one month postoperatively revealed that she was developing vaginal stenosis. She was scheduled for surgery, where the vaginal adhesions were released. Transvaginal laparoscopic removal of the rudimentary horn was also performed. The external genitalia were normal, typical vaginal mucosa with a depth of 6cm (**Figure 5**). Her postoperative recovery period was unremarkable, and she was discharged on the third postoperative day. The patient is currently using Syracuse vaginal dilators and is on long-term follow-up.

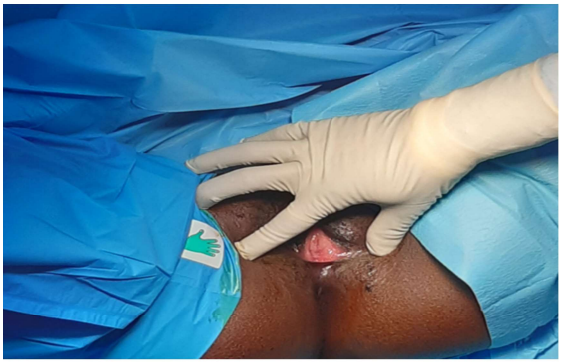

Figure 1: Perineal examination revealed a blind-ending vagina with a patent urethral opening.

Discussion

Müllerian duct abnormalities can result in minor anatomical variations to total aplasia. Mayer-Rokitansky-Küster-Hauser syndrome is a common cause of such abnormalities. This was a classic case of MRKH syndrome. It afflicts about 90% of patients who have vaginal agenesis. The patients have a normal female phenotype and can develop primary amenorrhea in adolescence with karyotype 46, XX (6). The two forms of MRKH syndrome are type 1, which presents with isolated Müllerian

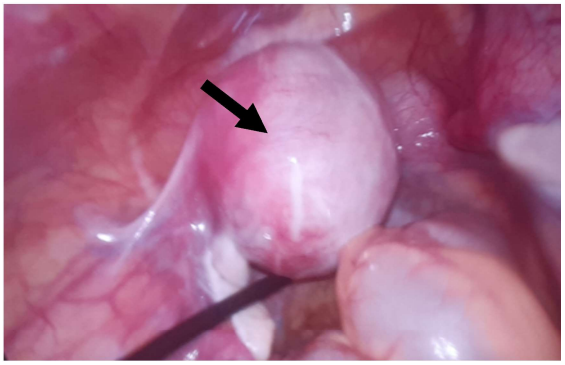

Figure 2: Ruderimentary uterine horn (arrow).

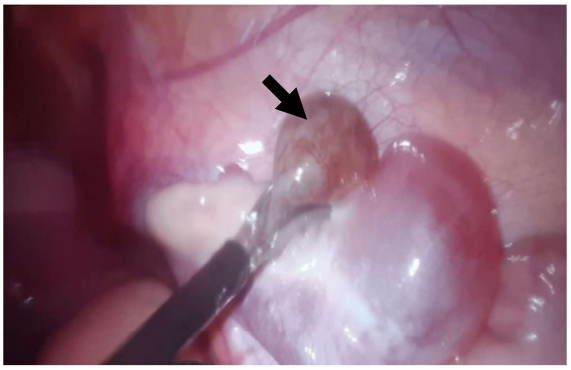

Figure 3: Endometrioma seen on laparoscopy (arrow).

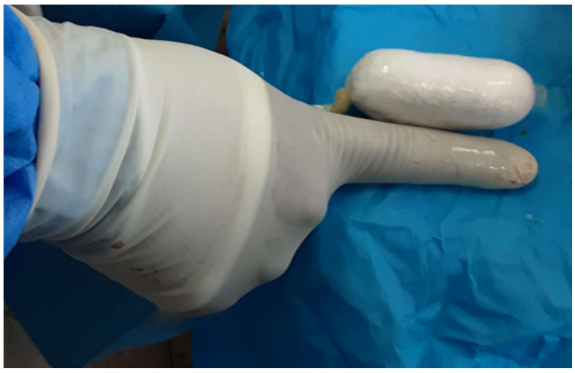

Figure 4: Improvised vaginal mold.

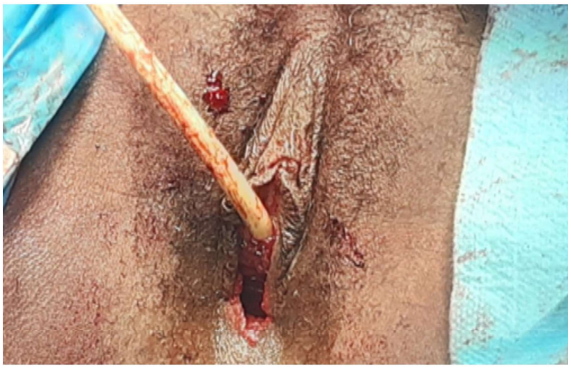

Figure 5: External genitalia post-vaginoplasty showing the introitus (arrow).

agenesis, and type 2, in which Müllerian agenesis primary amenorrhea in adolescence with karyotype 46, XX (6). The two forms of MRKH syndrome are type 1, which presents with isolated Müllerian agenesis, and type 2, in which Müllerian agenesis occurs together with other associated congenital anomalies. The most common are urinary and skeletal, especially scoliosis (7). Imaging studies (pelvic ultrasound, pelvic magnetic resonance) and hormone assays (luteinizing hormone, follicle-stimulating hormone, estradiol, and testosterone) are essential for the MRKH diagnosis (1,2). Radiography, audiography, electromyography, and pelvic laparoscopy can be requested depending on the symptoms (1). Differential diagnoses include androgen insensitivity, isolated vaginal atresia, and congenital absence of the vagina and uterus. Treatment can be delayed until patients mature emotionally and can start sexual activity (1). The condition can cause severe somatic and psychosocial deficits such as depression, necessitating psychological monitoring. Treatment options may be surgical or nonsurgical, depending on patient needs, functional needs, and motivation. Fertility counseling should be done. Alternative pregnancy options, such as gestational surrogacy and adoption, should also be discussed (6). Assisted reproductive techniques can be used with a gestational carrier or adoption (3). Advanced techniques, such as uterine transplantation, have also been implemented elsewhere (7).

Vaginoplasty is usually undertaken per the patient's wishes following counseling. Various vaginoplasty techniques have been used over time (7). The Wharton-Sheares-George technique is based on Sheares' principle in which a perineal skin is inserted as an isograft using the Müllerian ducts for orientation. It is also based on the Wharton principle, which states that if a vaginal insert is placed in the vagina for extended periods, vaginal epithelium regeneration can occur spontaneously (1). George modified the technique and omitted the creation of the perineal skin graft flap, therefore, making the operation simpler and safer (6). Laparoscopy is usually performed to exclude visceral injury. Initially, the examination is performed to identify a vaginal dimple, which serves as an anatomical landmark. Lateral and dorsal to the dimple are rudimentary Müllerian ducts in the plane between the bladder and the rectum. A channel is created using Hegar dilators starting with 2.5mm to 14mm on each side. The median raphe between dilators is transected using monopolar energy. The neovagina is inspected to ensure hemostasis. Two fingers should be inserted comfortably inside the vagina. An inflatable dilator should be inserted and stitched to keep it in for seven days. After this, it can be kept nightly for three months. Laparoscopy is repeated at the end to

check for any injury (1). Complications are minimal in Wharton-Sheares-George vaginoplasty; however, the risk of cystocele and rectocele and lifelong dilatation to avoid vaginal stenosis are the disadvantages of this method. The patient, in this case, developed vaginal stenosis, which was arrested early during the clinical review. The creation of the neovaginal axis in this technique makes it suitable for uterus transplantation; thus, the potential for women with MRKH to experience pregnancy (7).

Conclusion

Wharton-Sheares-George vaginoplasty is a minimally invasive, quick, reliable, and safe procedure for patients with Müllerian agenesis requiring vaginoplasty.

Consent for publication

Informed consent for publication was obtained from the patient.

Declarations

Conflict of interests

The authors declare no conflicts of interest.

Funding

None

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