CASE REPORT CRSLS

Evaluation of Post-Menopausal Bleeding in Two Patients with Sigmoid Neovaginoplasty

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ABSTRACT

Introduction: Mayer-Rokitansky-Kuster-Hauser syndrome or vaginal agenesis, is the rare congenital absence of the vagina with varying degree of hypoplasia of the Mullerian duct system and uterine development. One of the reconstructive surgical options for a vaginoplasty involves the usage of the large bowel.

Case Description: We report two cases of patients who have had a sigmoid neovaginoplasty presenting many years later with postmenopausal bleeding.

Discussion: The authors describe the evaluation that can be performed by the gynecologist and gastroenterologist to initiate the work-up for this rare presentation.

Key Words: Mullerian Agenesis, MKRH, Neovagina, Postmenopausal bleeding.

INTRODUCTION

Mayer-Rokitansky-Kuster-Hauser syndrome (MRKH), also known as vaginal agenesis, is a rare congenital disorder that affects woman with an incidence of 1/5000 female births.[1–3] It involves the failure of the uterus and vagina to develop properly in patients whose ovarian functions remain intact with normal external genitalia.[4] As a result, women with this condition develop normal secondary sexual characteristics and experience normal pubertal changes (normal breast development and growth of pubic hair) but do not menstruate, which is the initial clinical sign of MRKH syndrome. This disorder is usually detected in late adolescence with failure of menstruation and is the second most common cause of primary amenorrhea.[5]

The severity and range of this syndrome can vary significantly and is generally broken down into type I (absence of the uterus, cervix, and upper one-third of the vagina) or type II (involvement of additional organ systems most commonly the renal structures and spine) classification.[6] Because of the nature of this condition with the associated age group at presentation, MRKH syndrome can cause significant psychological conditions with depression and anxiety.[2] To address these concerns, multiple vaginal reconstructive surgical techniques have been described over the years including: skin grafts, peritoneal grafts, musculofasciocutaneous flaps, and bowel flaps.[7] Given the unique characteristics such as the location and proximity to the operative site, morphology, and similar tissue consistency, the sigmoid colon has been used for neovaginal reconstruction. The thicker mucosa layer provides resistance and durability that can make it less susceptible to trauma from coital activity.[8,9]

Primary carcinoma of the vagina is quite rare contributing to only 2% of all malignant neoplasms of the female genital tract.[10] In addition, given the rarity of MRKH syndrome, malignancy arising in the neovagina is extremely rare. To date, there has been only eight cases of adenocarcinoma arising in the neovagina involving bowel flaps described in the literature.[3,11–17] Although this malignancy is very uncommon, as more patients who have undergone this procedure are entering their postmenopausal years, and with a growing number of gender-affirming surgeries

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taking place yearly, physicians may see a growing number of patients with a sigmoid neovagina who may present with postmenopausal bleeding. The authors present two case reports for consideration involving the work-up of these patients.

CASE REPORTS

In Case 1, a 71-year-old female who was referred by her primary care physician (PCP) for postmenopausal bleeding which occurred intermittently for a few months. This patient never had a Papanicolaou test and received her care predominantly by her PCP for many years. After her doctor retired and she was evaluated by her new PCP, she was referred. A thorough pelvic examination was normal, and a "vaginal" Papanicolaou test revealed negative cytology. Human papilloma virus (HPV) serotyping was not readily available at the time (2002) and not performed. An magnetic resonance (MR) image of the abdomen and pelvis did not reveal renal or other organ anomalies, and a blind loop of bowel consistent with her history of a sigmoid neovaginoplasty was seen. This work-up confirmed MRKH type 1. The patient was scheduled for a vaginoscopy using normal saline, and few targeted biopsies were performed which revealed negative pathology. This case was performed with a gastroenterologist in the room. This patient underwent a colonoscopy prior to the vaginoscopy which was also negative.

Case 2 involved a 69-year-old patient referred by her obstetrician/gynecologist for postmenopausal bleeding occurring intermittently for several weeks. The patient stated that she has "never had a Papanicolaou test in my life." A careful examination revealed suspected vaginal stenosis, but the reconstruction appeared intact with no breakage, tear, or laceration (**Figure 1**). A thorough pelvic examination was normal, and a pediatric speculum was used and a "vaginal" Papanicolaou test was obtained for cytology which was negative. HPV serotyping was negative as well. An MR image of the abdomen and pelvis did not reveal renal or other organ anomalies, and a blind loop of bowel consistent with her history of a sigmoid neovaginoplasty was seen. No other pelvic or adnexal pathology was seen. This work-up confirmed MRKH type 1. The patient had a negative colonoscopy six months prior and underwent a vaginoscopy under intravenous sedation using normal saline as the distension medium. A few biopsies were performed which were negative (**Figure 2**). This procedure was performed with an experienced gastroenterologist in the room.

DISCUSSION

MRKH is a rare and largely sporadic congenital syndrome. Patients with this syndrome have a chromosomal pattern of 46XX with normal functioning ovaries; phenotypical female appearance consisting of normal external genitalia, breast, and pubic hair development; however, vaginal, cervix, and uterine development will vary, and can be associated with other malformations.[18,19] The differential diagnoses to consider include 5-α reductase deficiency, androgen insensitivity syndrome, hermaphroditism, congenital adrenal hyperplasia, Turner syndrome, and Mullerian-inhibiting substance deficiency.[5,20]

When post-menopausal patients with a history of MRKH who underwent a sigmoid neovaginoplasty present with vaginal bleeding, the blood is not originating from the uterus but from the sigmoid tissue itself. Due to its rarity, some physicians feel uncomfortable to initiate a work-up in these patients. The patient in case 1 did not have any documented medical records involving her surgery. The

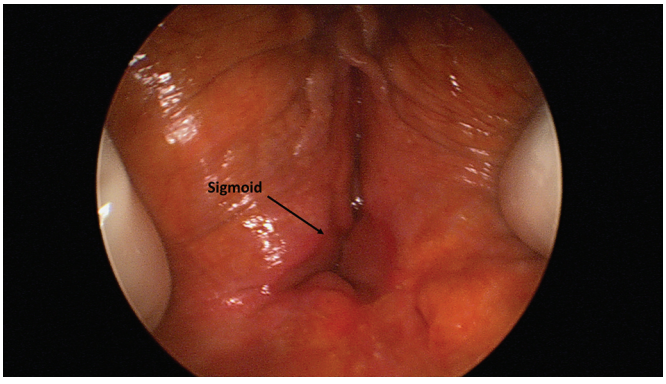

Figure 1. Intact anastomosis of neovagina.

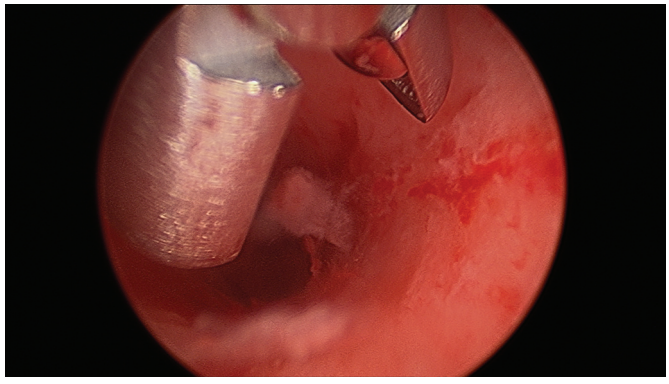

Figure 2. Biopsy via Vaginoscopy.

patient in case 2 did; however, the operative report was in German. Basic work-up should consist of a detailed and comprehensive medical history including any potential hormone treatments, and a meticulous physical and pelvic examination. Cytology can easily be obtained in the office, and an imaging study, preferably abdominal and pelvic MR imaging, should be obtained to assess other potential structural anomalies. Lastly, since the source of the bleeding may be originating near the apex of the blind pouch, rather than performing a random biopsy, a visually guided biopsy by performing a vaginoscopy is highly recommended. Since essentially bowel tissue is being assessed (and not necessarily vaginal tissue), performing the scope with an experienced gastroenterologist is recommended.

CONCLUSION

It is fortunate that adenocarcinoma arising in sigmoid colon neovagina is extremely rare. Given the timing and the refining techniques of colon vaginoplasty over the years, a growing number of patients who underwent this procedure are now entering their postmenopausal years. The incidence of postmenopausal bleeding in these patients will rise. Coupling this with the significant growth of gender-affirming surgeries taking place in just the past 20 years,[21–23] physicians will need to be aware of nontraditional approaches to carefully evaluate patients in order to rule out pathology. Continued cytology monitoring (apical pap smears) can be considered for these patients. Gynecologists should be aware and mindful of the work-up when such cases are encountered. The authors recommend the consideration of the work-up as shared and presented in these case reports.

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