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Case report

Perforation of sigmoid neovagina in a patient with Mayer-Rokitansky-Küster-Hauser syndrome

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ABSTRACT

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is a congenital aplasia of the uterus and upper part of the vagina in females. Treatment includes surgical creation of a functional neovagina. Perforation of the neovagina is extremely rare with only handful of cases reported in transgender patients post gender reassignment surgery. We report a first case of sigmoid neovaginal perforation in MRKS patient. The patient presented with progressively worsen abdominal pain and multiple intra-abdominal abscesses due to perforation of sigmoid neovagina. She was treated with surgical drainage and antibiotics and recovered clinically. Although exceedingly rare, we should keep history of sigmoid neovaginoplasty and possible perforation in patients with MRKS presenting with abdominal abscesses.

Introduction

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome, present in 1 in 5000 live births, is a congenital aplasia of the uterus and upper part of the vagina in females with normal secondary sex characteristics and female karyotype. It is generally diagnosed during adolescence when the patient present with primary amenorrhea. Treatment options includes creation of a functional neovagina. There are various different surgical and non-surgical methods available for vaginal construction [1]. Sigmoid vaginoplasty is considered safe procedure for vaginal agenesis with good cosmetic results. The properties of sigmoid colon (large lumen, thick walls resistant to trauma, adequate secretions for lubrication, not requiring prolonged dilatation and short recovery time) makes it a good candidate for vaginal construction [2]. Perforation of the neovagina is extremely rare with only handful of cases reported. On detailed review, it appears all the cases occurred in transgender patients. There has been no documented case of sigmoid neovaginal perforation in a female with MRKS.

Case

A 27-year-old woman with Mayer Rokitansky Syndrome (Mullerian agenesis), who underwent sigmoid neovaginoplasty nine years ago, presented to the clinic with lower abdominal pain and bilateral pelvic pain of two weeks duration. She denied routinely irrigating or dilating her neovagina, and normally had penetrative sexual intercourse every couple of weeks with her husband. However, due to life stressors, she had not had intercourse in a few months. CT imaging of the abdomen demonstrated tubular, heterogenous, fluid-filled structure, measuring 7.1 cm × 20 cm, extending from the pelvis to the lower abdomen, with blind ending (Fig. 1). An outpatient referral to the gynecologist was arranged but the patient's abdominal pain acutely worsened over the next 24 hours. She presented to the local Emergency Department (ED) with diaphoresis and significant distress due to pain. The vitals were unremarkable on presentation. Complete blood count (CBC) revealed leukocytosis of 15.6 k/mm^3 with absolute neutrophils of 9.8 k/mm^3. Repeat abdominal CT demonstrated increasing inflammatory process in the pelvis surrounding the reconstructed vagina (Fig. 2). The patient received empiric intravenous (IV) piperacillin-tazobactam at the outside hospital and then transferred emergently to our children's hospital associated with her urologist. On arrival to our facility, she was notably hypotensive (blood pressure BP 87/54 mmHg), tachycardic (heart rate HR 150 s/minute), afebrile (temperature 98.4 °F), and tachypneic (respiratory rate 31 s/minute) with oxygen saturation of 95 % on room air. She received four IV fluid boluses. The antimicrobials were empirically changed to IV ceftriaxone, IV vancomycin, and IV metronidazole. The patient was taken emergently to the operative room (OR) with the urology and general surgery teams for exploratory laparotomy, cystoscopy, and vaginoscopy. Intra-operatively, she had normal bladder and

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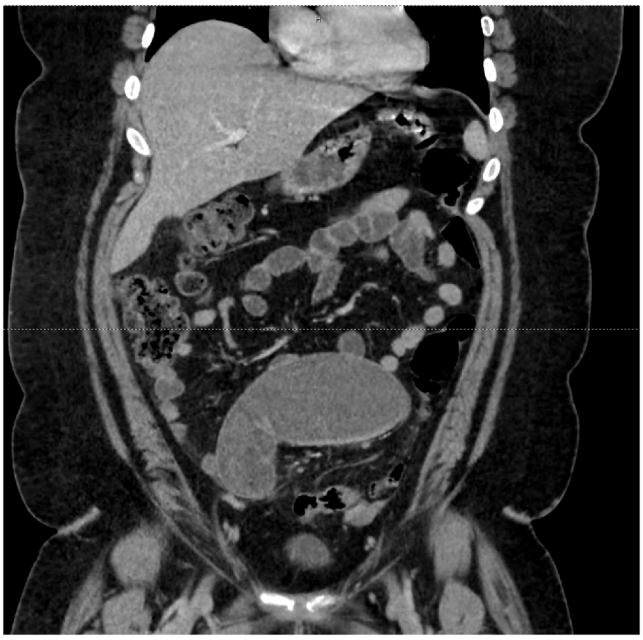

Fig. 1. Day 0: CT Abdomen/Pelvis ordered by primary care physician showed tubular heterogenous fluid filled structure extending from pelvis to lower abdomen, measuring 7.1cm × 20cm, with blind ending.

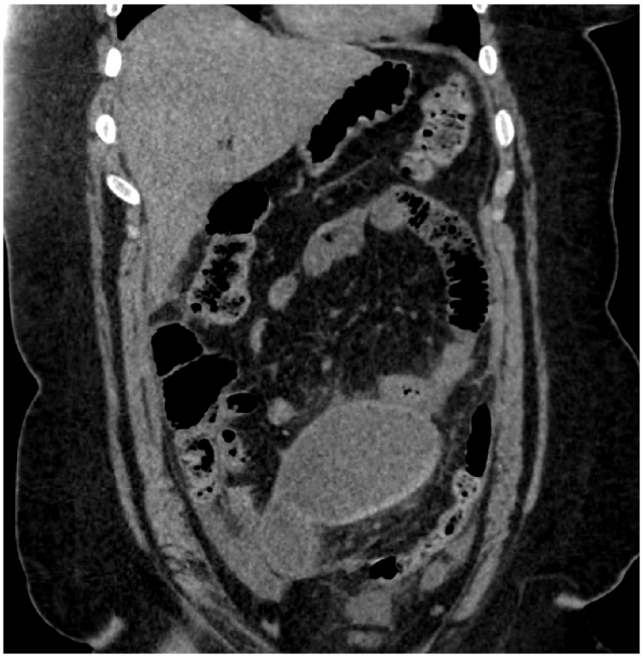

Fig. 2. Day 2: CT Abdomen/Pelvis ordered by Emergency Department, showed increasing inflammatory process in the pelvis surrounding the reconstructed vagina.

urethra but an entirely obliterated introitus, and diffuse intra-abdominal spillage of the mucus and perforated sigmoid neo-vagina. About one liter of purulent fluid was drained in the OR, and three intrabdominal drains were placed at that time.

Post-operatively, the patient remained intubated requiring mechanical ventilation and in septic shock requiring three vasopressor agents. The antimicrobials were transitioned to IV cefepime, IV vancomycin, and IV metronidazole due to preliminary peritoneal culture growing gram-negative rods. The blood cultures

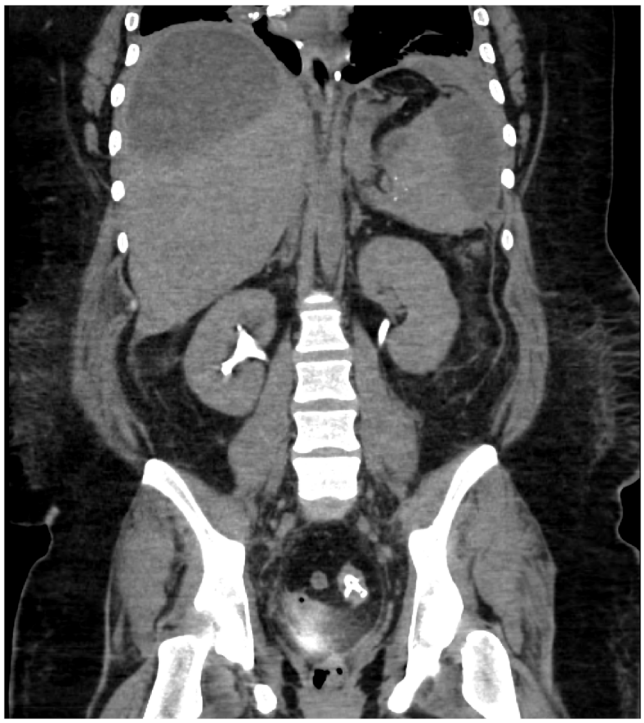

Fig. 3. Day 25: CT Abdomen/Pelvis showed abscess formation, largest next to the liver (15.6 cm), transcutaneous drainage catheter in pelvis, open anterior midline wound with wound vacuum.

remained negative. Once the peritoneal cultures finalized to *Bacterioides thetaioaomicron*, *Bacteroides caccae*, and *Actinomyces* species, the antimicrobials were changed to IV piperacillin-tazobactam on day 7 of her hospitalization. She was weaned off vasopressors and extubated on day 8. She was transferred to the general floor on day 12 and Infectious Diseases team was consulted for antimicrobial management. Additional susceptibilities for the *Bacteroides* species were requested to potentially narrow her antibiotics. While the susceptibilities were pending, the patient was discharged home, on day 15, with abdominal wound vacuum, and IV piperacillin-tazobactam for four weeks and plan for close follow-up with her primary care doctor, urologist and infectious diseases physician.

Unfortunately, two weeks post-discharge, she experienced generalized malaise and diffuse abdominal pain and readmitted with sepsis. Laboratory data revealed white blood count 13.2 k/cmm^3, absolute neutrophil count 9.5 k/cmm^3, D-dimer >5000 ng/mL DDU, and lactate 0.9 mmol/L. CT of the chest, abdomen and pelvis showed bilateral pleural effusions with loculated left pleural effusion and multiple new abdominal abscesses, largest next to the liver (15.6 cm), transcutaneous drainage catheter in pelvis, open anterior midline wound with wound vacuum (Fig. 3). Due to hypoxemia, she was transferred to the intensive care unit (ICU) and IV piperacillin-tazobactam was continued. She underwent placement of a right perihepatic drain with aspiration of 20 mL of purulence, but unsuccessful drainage of peri-splenic collection due to narrow window per interventional radiologist. Blood cultures remained negative. Interventional radiology was reconsulted and were able to drain 350 mL of pus from the right perinephric abscess and 90 mL of pus from her perisplenic abscess. Broad-spectrum PCR was sent on the drained fluid from the abscesses. She improved clinically, and on day 32 from the initial presentation, the antimicrobials were narrowed to IV ampicillin-sulbactam at discharge based on the susceptibilities from the original surgical cultures. Broad

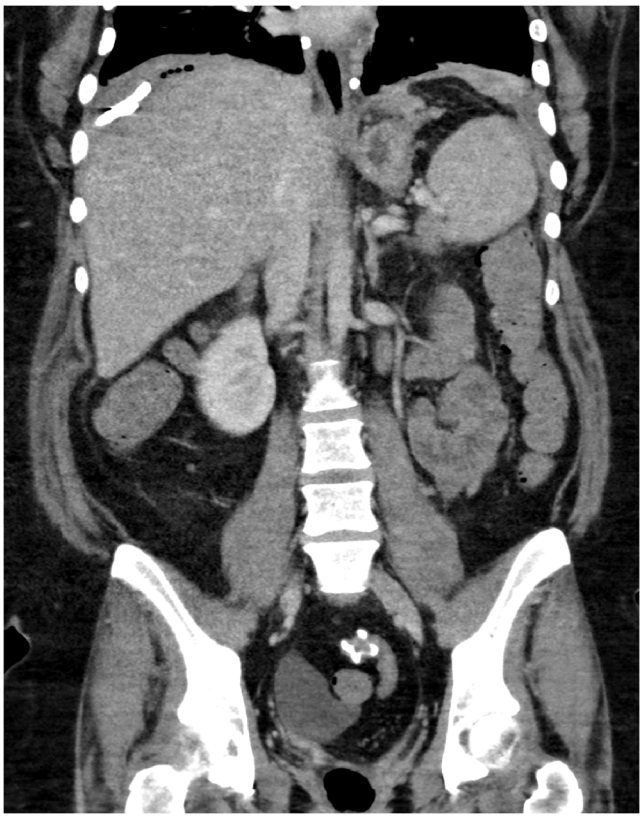

Fig. 4. Day 60: CT Abdomen/Pelvis showed decreased size of the right and left subphrenic fluid collections.

spectrum PCR from the fluid was positive for *Gleimia europaea* (formerly *Actinomyces europaeus*), *Alistipes onderdonkil, Varibaculum timonense*, and *Jonquetella anthropi*. She followed up at the adult infectious diseases clinic on day 44 and continued on ampicillin-sulbactam with plans to reimage on day 60. On clinic follow up, the patient had improved clinically, and the repeat CT abdomen demonstrated decreased in the size of right and left subphrenic abscesses. She was transitioned from IV ampicillin-sulbactam to oral amoxicillin-clavulanate for several weeks until complete resolution of the abscesses (Fig. 4).

Discussion

We report the first documented case of perforation of sigmoid neovagina in a female patient with Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome. Imprarato et al. conducted the largest case series of post sigmoid vaginoplasty follow-up in MRKS patient. The study included total of 62 patients, with 58 MRKS patients and 4 females with scarred vagina due to previous radiation therapy for vaginal or cervical cancer. There were no infections reported in MRKS group. There was a case of necrotizing fasciitis in non-MRKS group. The patient was treated with surgery, penicillin, clindamycin and diverting colostomy and complete recovery was achieved [3]. In another study of 42 transgender patients, the long-term complications of intestinal vaginoplasty included introital stenosis in 6 patients (14.6 %) and mucosal prolapse in one patient (2.4 %) but no perforation was reported. The literature review of nine studies between 1997–2007 with 278 transgender patients had eight cases of superficial wound infection, one case of necrotizing fasciitis and two cases of neovaginal perforation [4].

There are three additional case reports of perforation of sigmoid negovagina in male to female transgender patients. Ligouri reported a case of acute peritonitis due to introital stenosis that caused perforation [5]. Another case of neovaginal perforation was described by

Amirian et al. The trigger was accidental introduction of air by douche pump. The patient was treated with IV cefuroxime, metronidazole, and gentamicin for 7 days and later switched to oral amoxicillin and metronidazole for 10 days [6]. Shimamura reported neovaginal perforation in a male to female transexual patient. The patient underwent drainage of intra-abdominal abscess and treated with antibiotics for 8 weeks. The pathogens or antimicrobials were not described [7].

Birse et al. studied neovaginal microbiome of transgender females post gender reassignment surgery and found *Porphyromonas*, *Peptostreptococcus*, *Prevotella*, *Mobiluncus*, and *Jonquetella* to be the prominent organisms, while cis vaginas primarily had *Lactobacillus* and *Gardnerella* [8]. Abdominopelvic actinomycosis is a rare disease. Actinomyces is normal flora of oral, gastrointestinal and pelvic mucosa but becomes pathogenic when mucosal barrier is disrupted and can lead to tissue granulation, dense fibrosis and abscess. Even in these cases, the colon perforation is a rare event. Actinomycosis is generally treated with IV penicillin for 2–6 weeks followed by 6–12 months of oral antimicrobials [9].

Recent advances in microbiologic taxonomy, using genotypic methods have identified many new Actinomyces species. *Actinomyces europaeus* is one of the recognized pathogens. It has been reported to cause breast abscess, brain abscess and necrotizing fasciitis [10]. Approximately 75%–95% of the actinomyces infections are polymicrobial. The most common co-isolates are Actinobacillus actinomycetemcomitans, Eikenella corrodens, Fusobacterium, Bacteroides, Capnocytophaga, Staphylococcus, Streptococcus, and Enterococcus spp. The choice of antimicrobials covering the co-pathogens is particularly important in invasive infections and lower-abdominal infections. A combination beta-lactam plus beta-lactamase inhibitor is indicated in these infections [11].

Our patient had mixed gram-positive and gram-negative anaerobic infection with actinomyces species (*Gleimia europaea*), bacteroides species (*Alistipes onderdonkil, Varibaculum timonense*) and synergistetes species (*Jonquetella anthropic).* The patient was treated with surgical drainage and antibiotics and recovered clinically. Although exceedingly rare, we should keep history of neovagina and possible perforation in patients with MRKS presenting with abdominal abscesses.

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Ethical approval

Not applicable.

Consent

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

Author contribution

Aneesha Kamath: (1) the acquisition of data and interpretation of data, (2) drafting the case (3) final approval of the version to be submitted.

Saira Butt: (1) the conception and design of the study (2) drafting the article including introduction and discussion (3) revising the article for important intellectual content, (4) final approval of the version to be submitted.

Declaration of Competing Interest

All authors have no conflicts of interest.

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