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Case Report

Cervical Os Marsupialization in the Initial Management of a Rare Case of Obstructive Uterine Didelphys with Ipsilateral Renal Agenesis

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ABSTRACT

INTRODUCTION: Uterine didelphys is an uncommon Mullerian anomaly that often is associated with obstructive pathology. Without early intervention, obstruction can progress to potentially fatal complications including pyometra and sepsis. Due to the rarity and variability of this condition, treatment options have not been systematically studied.

CASE DESCRIPTION: In this case report, we discuss a 16-year-old female who presented with acute sepsis secondary to pyometra in the setting of chronic dysmenorrhea and purulent vaginal discharge. On imaging, she was found to have uterus didelphys with right renal agenesis and a hypoplastic right cervix along with a pelvic fluid collection. During exam under anesthesia, no obstructing hemivagina was found, however the right cervical os was identified with purulent material draining from it and identified as the site of obstruction. The right cervical os was extended and its edges marsupialized. The patient recovered well postoperatively with a course of antibiotics and continued to be followed in an outpatient setting, where she was started on Depo Provera for menstrual suppression. She remained asymptomatic for months, but after discontinuing the Depo for some time, began to experience pelvic pain and purulent vaginal discharge again. While she agreed to resume Depo treatment, definitive treatment with right hemihysterectomy was discussed and eventually undertaken.

DISCUSSION: Although hemihysterectomy may be required in many cases, the technique of marsupialization of a cervical os is unique and may be a new approach to patients with non-classic anatomic presentations of obstructive symptoms with uterine didelphys.

KEYWORDS: uterine didelphys, Mullerian anomaly, pyometra, marsupialization, hemihysterectomy

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INTRODUCTION

Mullerian duct anomalies encompass a wide range of anatomical variations. One of the rarest of these is uterus didelphys, which has a reported incidence between 0.1% and 3.8%. This condition results from failure of Mullerian duct fusion resulting in two separate uterine cavities, two cervices, and either a full or partial vaginal septum. Many uterus didelphys cases are also accompanied by ipsilateral renal anomalies due to the intimately associated development of the two organ systems. Depending on the level at which Mullerian duct fusion has failed, a partial vaginal septum or hypoplastic cervix may obstruct more proximal parts of the reproductive tract. Patients with obstructive uterus didelphys typically present several years after menarche with dysmenorrhea due to the progressive accumulation of menstrual material in the obstructed hemivagina, termed hematocolpos.[1] Management involves pain control and menstrual suppression until vaginal septum excision and hematocolpos drainage can be performed, with hemihysterectomy reserved for recurrent or complicated cases.[2] However, without early intervention, prolonged pressure in the hematocolpos may perforate the vaginal septum and allow the ascension of pathogenic bacteria that cause reproductive tract infection.[3,4] These patients may develop pyometra, a rare and potentially fatal accumulation of purulent material in the uterus that can progress to uterine perforation, acute peritonitis and sepsis.[5] In this paper, we describe the case of an adolescent female presenting with pyometra

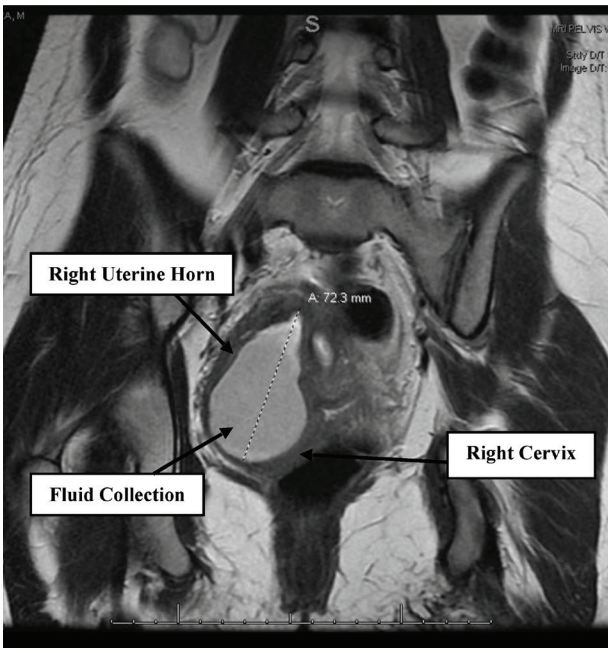
in the setting of undiagnosed uterine didelphys with unilateral cervical hypoplasia and ipsilateral renal agenesis. We outline her initial treatment with cervical marsupialization, follow-up care, and definitive treatment with right hemihysterectomy.

CASE SUMMARY

Our patient is a 16-year-old G0P0 who presented to the emergency department with a 3-day history of bilateral abdominal pain accompanied by fever, nausea, and decreased appetite. The patient also reported a thin, malodorous, yellow vaginal discharge of about a year duration that was occasionally copious enough to warrant use of a tampon. The patient endorsed a history of heavy and painful periods accompanied by bloating and fatigue. The patient had never been sexually active and had no history of sexually transmitted infection. On presentation, the patient was tachycardic, tachypneic, and febrile to 38.2 °C with physical exam significant for bilateral lower abdominal tenderness more pronounced on the right and mild distension of the lower abdomen. Speculum exam revealed a bulge on the right vaginal wall with tenderness to palpation. Abdominal and pelvic ultrasounds were significant for absence of the right kidney and a suspected bicornuate uterus measuring 9.8 x 3.9 x 3.9cm with complicated fluid in the lower uterine canal. MRI of the pelvis revealed uterine didelphys with fluid collections or abscesses in the pelvis and an obstructive right cervix. At this time, the patient was diagnosed with acute sepsis from possible pyometra and was started on a regimen of clindamycin, ampicillin, and gentamicin along with IV fluids and medications for pain control.

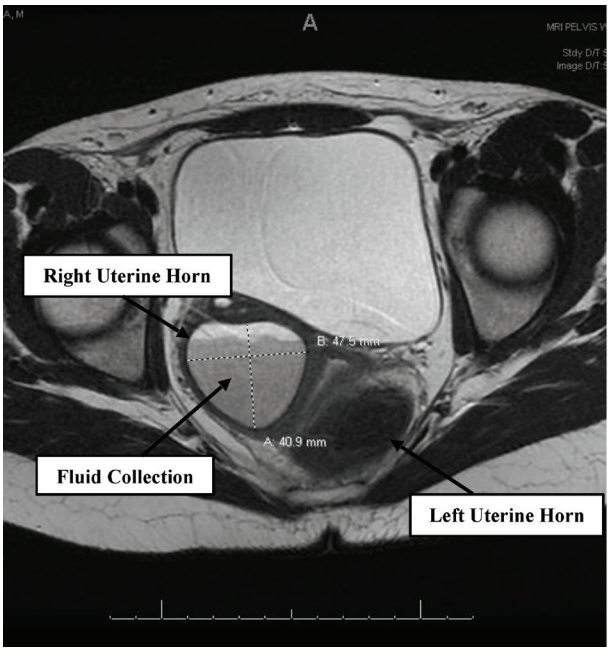
The following day, the patient underwent examination under anesthesia. Bimanual exam revealed right-sided pelvic fullness and fluctuance at the right vaginal side wall and a large amount of purulent material drained from the vagina at the conclusion of the exam. Further examination revealed a normal-appearing left-sided cervix with no clear opening in the right vaginal wall from which the discharge originated. Vaginoscopy with vaginal hydrodistension was then performed and revealed a small opening in the vaginal mucosa on the right side wall from which purulent fluid was draining. This opening was deduced to be the external os of the right cervix and was progressively dilated to allow for hysteroscopy of the right uterus, which showed a normal appearing endocervical canal and endometrium. A uterine sound of the right uterus revealed a total length of 10 cm. The right external os was then extended laterally and marsupialized

FIGURE 1. Coronal MRI of the Pelvis



This coronal view of the pelvis shows a fluid collection measuring 72.3mm in height within the right uterine horn obstructed by the stenosed right cervix.

FIGURE 2. Axial MRI of the Pelvis



This axial view of the pelvis shows both the left and right uterine horns with a fluid collection measuring 40.9mm x 47.5mm present within the right uterine horn.

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by stitching the edges of the incision to the vaginal mucosa. Her post-operative course involved continued fever spikes that resolved with adjustment of the antibiotic regimen. By the fourth day post operatively, the patient remained afebrile, was tolerating oral intake, voiding spontaneously, ambulating and her pain was controlled, and she was discharged home in stable condition.

About 2 weeks later, the patient returned to the outpatient clinic with only mild pelvic cramping and continued abnormal vaginal discharge without odor. Her incisions were clean, dry, and intact. Speculum exam revealed copious purulent discharge draining from a sutured area on the right vaginal wall. On bimanual exam, some tenderness was elicited at this site, but no obvious defect was palpable. At this time, the patient was started on Depo-Provera (DMPA) to decrease menstruation and the buildup of fluid in the right uterine horn. At a second follow-up visit months later, surgical inflammation had subsided sufficiently to perform a more extensive pelvic exam. This revealed a normal vagina and midline left cervix. The right cervical os was flush with the right vaginal side wall and deviated to the right. Both cervical ostia were patent, and the left cervical length was greater than the right. No vaginal septum could be appreciated. Due to the patient's significant clinical improvement and lack of symptoms at this time, further surgical intervention was not warranted, and the patient continued on Depo for menstrual suppression. However, over the next year, the patient missed several Depo shots and began gradually experiencing the return of symptoms. Additionally, because the patient desired future fertility, reproductive endocrinology and infertility (REI) was consulted. It was recommended that the right uterine horn be removed to minimize the risk of cervical stenosis and recurrent pyometra and thus injury to the remaining reproductive organs that could compromise fertility. Due to the recurrence of symptoms on Depo and the recommendation from REI, the patient elected to undergo right hemihysterectomy.

The right hemihysterectomy was performed laparoscopically. Upon abdominal cavity entry, omental and peritoneal adhesions were observed over the right fallopian tube, appendix, cecum, right pelvic brim, and right anterior and posterior cul-de-sacs. Following lysis of adhesions, the reproductive organs were visualized and a broad-based connection between the uterine horns was noted. It was decided that a supracervical hysterectomy rather than complete hysterectomy would be performed due to the proximity of the two cervices and risk of compromising the left cervix and therefore future fertility. An incision was made at the midline between the uterine horns and was extended across the right internal os to remove the right uterus. Special care was taken to use minimal energy while dissecting the midline to avoid damage to the left uterine horn. The cervical canal was cauterized and the cervical bed was imbricated over the os. The specimen was removed through a colpotomy in the posterior vaginal fornix which was then closed. The patient recovered well postoperatively and continued to be asymptomatic at follow-up several months later.

DISCUSSION

In this case, we describe an adolescent presenting with pyometra secondary to undiagnosed uterus didelphys with a stenosed right cervix and ipsilateral renal agenesis. Our patient presented similarly to many patients with obstructive uterus didelphys, with progressive dysmenorrhea several years after menarche due to the accumulation of menstrual material in an obstructed hemivagina.[1] In our case, this obstruction occurred at the stenosed right cervix. The cervical os served as an opening through which pathogenic vaginal bacteria could ascend and infect the collection of blood in the cervix and uterus, a process that typically occurs in the hemivagina behind a perforated vaginal septum.[3,4] In our patient, this resulted in pyometra and acute sepsis.

In most cases of obstructive uterus didelphys, surgical management involves vaginal septum excision and hematocolpos drainage.[2] In our case, however, with no vaginal septum to excise, a modified technique was required. The stenosed right cervical os was extended and its edges marsupialized to allow drainage of the purulent fluid collection and to promote prolonged patency of the opening. While marsupialization is often used for drainage of cysts or abscesses, it has not been described in cases like this and it is thus difficult to predict its expected success rate. However, it is known that in cases of uterus didelphys with thicker septa or incomplete septal excision, stenosis with repeat obstruction is more likely to occur.[2] With the thick tissue of a stenosed cervix in place of a vaginal septum, our patient likely fell into this category of cases at higher risk for repeat obstruction. She did in fact begin reexperiencing obstructive symptoms despite months of menstrual suppression with Depo-Provera which has, admittedly, only been used for symptom relief while awaiting surgical intervention and is not currently considered a long-term treatment option.[2]

In cases with recurrent obstruction or infection such

as ours, hemihysterectomy is often the most successful definitive treatment.[6,7,8] However, care must be taken in patients with a history of pelvic infection and chronic inflammation as this can cause difficulty identifying structures, lysing adhesions without causing organ damage, achieving adequate hemostasis, and especially dissecting the diseased uterus from remaining structures to preserve fertility.6 Indeed, numerous filmy adhesions likely resulting from our patient's history of pyometra covered her pelvic and lower abdominal structures and required lysis. Precautions were also taken while dissecting the connection between her uterine horns and closely approximated cervices to ensure no residual damage to her remaining reproductive organs and thus maximize the potential for successful future childbearing. Our hope is that these surgical precautions and the removal of her right uterine horn will prevent future complications in our patient and preserve future fertility.

CONCLUSION

While obstructive uterus didelphys is typically treated definitively with vaginal septum excision and hematocolpos drainage, anatomical variations in this anomaly along with its relatively low incidence make it difficult to streamline surgical management and compare treatment outcomes. Even in cases with an excisable vaginal septum, certain aspects of the anatomy may cause stenosis and repeat obstruction after the initial procedure and necessitate hemihysterectomy. In our case, marsupialization of the patient's stenosed right cervix was able to relieve her symptoms and concurrent pyometra acutely. However, she eventually experienced a recurrence of her symptoms and required a right hemihysterectomy to prevent further complications and preserve fertility. To date, this specific anatomy has not been described in the literature on uterus didelphys and the surgical technique used in the case has likewise not been evaluated for its efficacy in this situation. It is therefore difficult to predict if outcomes may vary among patients with different clinical courses and anatomical variations. We hope that with the continued advancement of diagnostic imaging, additional patients with this anatomy may be identified and their clinicians will be able to reference this case when discerning treatment options. With early diagnosis and intervention, patients with Mullerian anomalies will hopefully avoid unfavorable and even fatal outcomes.

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