Management of a late-presenting complex - an unclassified uterine anomaly in the presence of large leiomyomas

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Summary

This is a case report of a unique, late-presenting, Müllerian anomaly in an infertile patient. The authors discuss the diagnostic challenges of characterizing distorted gynecological anatomy by Müllerian anomalies in the presence of sizeable coexisting fibroids. This case report adds new insight to the already-existing understanding of Müllerian anomalies by demonstrating how a symptomatic and benign uterine pathology can complicate the diagnosis and management of patients with Müllerian defects.

Key words: Complex Müllerian anomaly; Leiomyomata; Surgical management of Müllerian anomalies; Diagnostic imaging of Müllerian anomalies.

Introduction

Müllerian anomalies are rare defects of the internal female reproductive system with an estimated prevalence of 6.7% in the general population and 7.3% in the infertile population [1]. The American Fertility Society anatomically classifies Müllerian anomalies into six different classes based on availability of treatment and fertility prognosis [2]. Cervical agenesis and dysgenesis have been organized into four categories: 1) cervical agenesis, 2) cervical fragmentation, 3) cervical fibrous cord, and 4) cervical obstruction [3]. Very rare and unclassified complex anomalies have been described [4-6]. The authors present a case of congenital absence of the cervix and a vertical uterine fusion defect. This as of yet described Müllerian anomaly has features of both class I (agenesis) and class III (non-fusion) defects.

Additionally, Müllerian anomalies are commonly associated with defects of the urinary system. Preoperative evaluation of the collecting system is imperative. The surgical management of Müllerian anomalies remains controversial given the numerous variants, their associated urologic anomalies, as well as their uncertain prognosis [4]. Because of its rare incidence, the management of cervical agenesis has been minimally discussed in the literature. Classically, patients have been managed by abdominal hysterectomy or by restoring the menstrual outflow tract via uterovaginal anastamosis [3-9].

The authors describe a complex, unclassified Müllerian anomaly with a large uterine leiomyoma in one uterine horn and duplication of ureters. A review of the pertinent literature, as well as the diagnostic challenges and logic supporting the present surgical management, are discussed in this case report.

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Case Report

A 39-year-old G0 presented with intermittent abdominal pressure and pain for six months. Her history was significant for primary amenorrhea and a history of diagnostic laparoscopy 20 years ago.

Speculum exam revealed a seven cm vagina and absent cervix. Her external genitalia were normal, and she had Tanner stage 5 breasts and pubic hair. Pelvic exam revealed an irregularly-shaped, non-tender, mobile mass extending to the level of the umbilicus.

A second, small, non-tender left-sided mass was palpated, which was thought to be either an ovary or a pedunculated fibroid.

Transvaginal ultrasound (TVUS) demonstrated a midline pelvic mass suggesting multiple myomas, the largest of which measured 12 cm. A computed tomography (CT) scan revealed a 18 x 15 cm complex pelvic mass arising from a right pelvis, a second 6 x 6 cm mass arising from the left (Figure 1A), a duplex left kidney with duplicated left renal collecting system (Figure 1B), and a normal right kidney and ureter.

The surgical management of symptomatic, large pelvic mass was planned via exploratory laparotomy with pelvic mass removal. Due to the identification of a complex Müllerian anomaly along with a duplicated left collecting system, the patient underwent preoperative cystoscopy and bilateral ureteral stent placement.

Intraoperative findings at time of exploratory laparotomy were significant for a duplicated left ureter and uterine didelphys with cervical agenesis. The right uterus had multiple leiomyomas, the largest of which measured 18 cm (Figure 2A). Neither uterus had an identifiable endometrial cavity. No overt cervical tissue was palpable; however, a 0.5 cm fibrotic area was noted between the vagina and base of the uteri. Ovaries were bilaterally normal.

A hysterectomy of the didelphys uterus with cervical agenesis and associated multiple, large leiomyomas was performed (Figure 2B). The patient's post operative course was uneventful. She was discharged home on post-operative day 2.

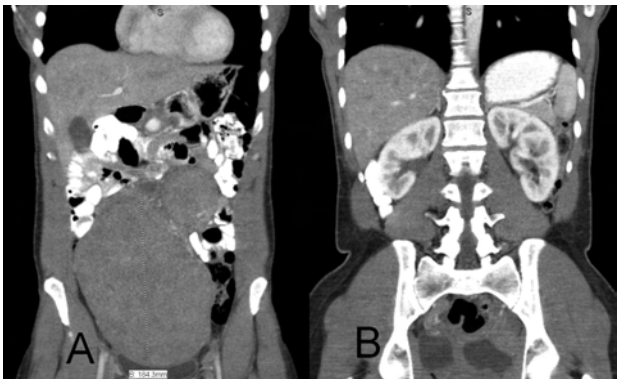

Figure 1. — CT scan revealing a complex pelvic mass. A 18 x 15 cm mass can be seen arising from the right pelvis (A) and the duplex left kidney with duplicated left renal collecting system (B).

Discussion

The management of Müllerian anomalies should be tailored to the unique aspects of the defect and/or clinical presentation. Patients with cervical agenesis and hematometra or pain may be candidates for fertility-conserving surgical management. Surgical canalization has been described resulting in normal menstrual bleeding, resolution of cyclic pelvic pain, and some potential for fertility [3-6]. Definitive surgical management is a hysterectomy.

Cervical agenesis is an extremely rare anomaly with 116 cases of transverse cervical defects described in the literature since 1900 [6]. Patients often present in late adolescence with amenorrhea and cyclical abdominal pain. Patients without an endometrium, like the present patient, may not have associated pain. It is important to evaluate patients for the presence of an endometrium. If the endometrium is absent, patients do not necessarily require surgical intervention. However, they should have routine pelvic exams to evaluate other vaginal, uterine, or ovarian pathologies.

The standard imaging modality for Müllerian defects is magnetic resonance imaging (MRI) [10, 11]. MRI allows for evaluation of uterine cavities, communication between rudimentary uterine horns, and renal or collecting system anomalies [11]. The present patient had a prior CT scan demonstrating her pelvic mass and collecting system anomaly. She also had an ultrasound confirming cervical absence and the authors felt that there was no added benefit from obtaining further imaging. The most common collecting system anomaly found with Müllerian defects is a unilateral duplicated ureter that fuses into one ureteral orifice. If managed surgically, the authors recommend placement of ureteral stents, especially if a urinary anomaly is suspected or known. Stent placement allows for rapid identification of anatomy and intraoperative recognition of ureteral injury.

The question of fertility is common in these patients. Fortunately, the present patient had normal ovaries; thus biologic fertility would be an option via in vitro fertilization (IVF) with a gestational carrier.

In summary, the authors have demonstrated the characterization of a unique, late presenting, Müllerian anomaly. They have discussed the diagnostic challenges of characterizing distorted gynecological anatomy by Müllerian anomalies in the presence of sizeable coexisting fibroids along with a surgical management option. Thus, this case report adds new insight to the already-existing understanding of Müllerian anomalies by demonstrating how a symptomatic and benign uterine pathology can complicate the diagnosis and management of patients with Müllerian defects.

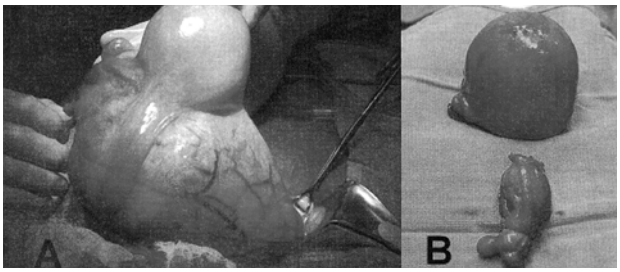

Figure 2. — Gross photographs of the anomaly. Large leiomyomas (A) and didelphys uterus (B) can be seen. The right uterus is shown above and the left uterus is shown below.

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