INTRODUCTION

Genetic errors and teratogenic events during embryonic development can lead to congenital abnormalities of the female reproductive tract. Many patients are asymptomatic while some major abnormalities can cause severe impairment of menstrual and reproductive functions. This case report focuses on Müllerian anomaly class III, uterine didelphys, along with obstructed hemivagina and ipsilateral renal agenesis (Herlyn-Werner-Wunderlich syndrome).

CASE

The patient presented with cyclic pelvic pain due to the hematometra and hematocolpos. Prior work-up for recurrent urinary tract infection showed an absent right kidney. Further imaging also revealed two uteri consistent with uterine didelphys (Figure 1). She was diagnosed with obstructed hemivagina ipsilateral renal agenesis (OHVIRA) and underwent resection of vaginal septum. However, the patient had recurrence of obstructive symptoms more than a year after the surgery was performed. She was initially planned to have re-excision of the vaginal septum. But upon vaginal exploration, the right cervix was not identified (Figure 2). This prompted further evaluation with magnetic resonance imaging revealing cystic dilatation of the right uterine horn with hypoplastic right vagina appearing to end blindly and no intravaginal septum formation noted. Patient subsequently underwent hemihysterectomy. The right hemiuterus was noted to have a depression on its thickened inferior aspect but no cervix was identified (Figure 3).

CONCLUSION

This case is a variant of the classic HWW syndrome as there was noted cervical atresia on the right uterine corpus. A thorough preoperative evaluation and accurate intraoperative assessment of patients with müllerian anomalies can decrease misdiagnoses, guide appropriate intervention, and decrease risk of future reproductive complications.

REFERENCES