Office treatment of didelphys uterus and obstructed hemi-vagina with renal agenesis: presentation and management of four cases of Herlyn-Werner-Wunderlich Syndrome

Fascilla FD , Cramarossa P , De Palma D , Cannone R , Olivieri C , Murgia F, Fumarulo VV , Cicinelli E , Bettocchi S.

Dipartimento di scienze biomediche ed oncologia umana, Università degli studi di Bari "A. Moro", II U.O. di Ginecologia ed Ostetricia, policlinico di Bari, Bari, Italia

Abstract

Objectives:
Purpose of this study is to describe clinical presentations, diagnostic process and office treatment of didelphys uterus and obstructed hemi-vagina with ipsilateral renal agenesis (also known as Herlyn Werner Wunderlich syndrome), and to suggest a proper diagnostic and therapeutic management based on our office experience.

Methods:
Four cases of Herlyn Werner Wunderlich Syndrome treated in office hysteroscopy. **Case 1 and 2**: Two sisters, not twins, aged 26 and 21 years, both with dysmenorrhea, dyspareunia, vaginal discharge, methrorragia and pelvic pain were diagnosed for didelphys uterus and obstructed hemi-vagina after bi-dimensional (2D), three-dimensional (3D) transvaginal sonography (TV-US), and office hysteroscopy (OH). Transabdominal sonography (TA-US) and magnetic resonance (MR) confirmed renal agenesis ipsilateral to obstructed hemi-vagina. Diagnosis of Herlyn Werner Wunderlich was settled down. Malformation in one patient was specular to the sister. OH was diagnostic and therapeutic in both cases. **CASE 3:** 12 years, virgo, with acute abdominopelvic pain and cryptomenorrhea. She underwent 2D and 3D transabdominal and transrectal sonography to get a diagnosis of didelphys uterus. Bimanual palpation and hysteroscopy revealed a vaginal septum occluding one hemi-vagina. MR revealed renal agenesis. Vaginal malformation was removed by office hysteroscopy. **CASE 4:** 16 years old young woman with dysmenorrhea, methrorragia and persistent pelvic pain. Bimanual palpation, TA and 2D/3D TV sonography were performed. A didelphys uterus and a vaginal septum occluding one hemi-vagina was revealed and renal agenesis were suspected. MR confirmed Mullerian duct anomalies and malformations. Office hysteroscopy allowed treatment of vaginal malformation.
Treatment consisted in office operative vaginoscopy: 1) opening of vaginal recess and drainage of hoematocolpos when present, 2) visualization of both external uterine os, 3) removal of abundant vaginal by using Twizzle bipolar electrode and 4) superficial coagulation of the vaginal wall to retract the mucosa by using Spring bipolar electrode.
One month later the Office procedure the patients underwent follow-up with removal of residual vaginal exceeding tissue, vaginoscopically. The vagina appears larger than usual, but within normal ranges. In Case 2, hysteroscopic examination has been performed with laparoscopic control that revealed the progression of the hysteroscope through the hemivagina till the external iliac vessels, via a narrow canal. It suggest that the vaginal malformation consists in an ureteral recess.

Results:
All patients were treated in an office setting, with minimal discomfort, time waste, sequel and costs. In cases 1, one of the sisters become pregnant, successfully, 7 years later in the hemi-uterus ipsilateral to malformation; in case 2 and all the other complete or almost complete relief of symptoms was reached. Subsequent follow up, settled down with vaginal-hysteroscopy and 2D/3D transabdominal and transvaginal sonography was optimal.

Conclusions:
Conservative treatment of Herlyn Werner Wunderlich syndrome, performed in an office setting by expert hysteroscopists, seems to be the best management, according to our experience. Physiologic pregnancy is possible even in the hemi-uterus ipsilateral to malformation and renal agenesis.