Title: Unexplained Abdominal Pain and Sepsis in Herlyn-Werner-Wunderlich Syndrome: A Diagnostic Challenge

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Background/Synopsis: Herlyn-Werner-Wunderlich (HWW) syndrome is a rare condition characterized by a Mullerian duct anomaly. The triad of uterine didelphys, obstructed hemivagina by a longitudinal vaginal septum, and ipsilateral absent or severely malformed kidney defines the syndrome. Patients often present with dysmenorrhea and cyclic pelvic pain secondary to hematometra and hematocolpos. Secondary infections, pelvic adhesions, endometriosis, infertility, spontaneous abortion, preterm delivery, fetal malpresentation, and renal impairment are all potential complications if left untreated. Magnetic Resonance Imaging (MRI) is the gold standard for pre-operative diagnosis, typically followed by surgical resection of the obstructing vaginal septum to restore physiological menstrual flow.

Objective/Purpose: To report a case of HWW syndrome that presented with several complications, which led to sepsis, a complex hospital course, and an unexpected surgical diagnosis.

Methods: Case Report

Results: A 21-year-old female G1P0010 with a known history of HWW syndrome presented with severe left lower quadrant abdominal pain for three days. During her first year of life, she underwent a left nephrectomy for a poorly functioning polycystic kidney. Also, she was treated one year prior for a chlamydia infection. She had previously been evaluated for similar complaints of left lower quadrant abdominal pain twice in the past four months. Unfortunately, due to insurance issues, she had not been able to obtain vaginal septum surgery. On physical exam, the rupture site revealed copious purulent, foul-smelling discharge and scant vaginal bleeding. MRI revealed HWW anatomy, left pelvic abscess 12cm in size, and likely reactive lymph nodes. She was admitted for a tubo-ovarian abscess (TOA) and started on ceftriaxone, doxycycline, and metronidazole. The following day, she underwent incision and drainage of the left hemivagina and pyometrocolpos, with partial vaginal septum resection. The surgery led to some immediate pain improvement. On post-operative day 1, she acutely decompensated with persistent tachycardia to 130 beats per minute, tachypnea, fever (38.5 °C), new oxygen requirement with pleuritic chest pain. Computerized tomography (CT) angiography and echocardiogram were negative. Internal medicine recommended a transition to broader antibiotics (vancomycin and piperacillin-tazobactam). CT abdomen and pelvis showed a persistent 12cm pelvic abscess, which was subsequently drained by interventional radiology, yielding 110mL of purulent fluid. The pelvic abscess fluid grew *Staphylococcus epidermidis*. After some improvement over the next three days, her condition worsened, accompanied by a new fever, leukocytosis (WBC 24,000 cells/µL), and a platelet count of 953,000/µL. The patient's abdominal pain worsened, with concerns for peritonitis. An abdominal x-ray was non-diagnostic. Due to suspected peritonitis, persistent fluid collections on imaging, and failure to

improve on antibiotic therapy, a second surgery was recommended two weeks following the first surgery with Gynecologic Oncology staffing. The patient underwent a diagnostic laparoscopy that was converted to an exploratory laparotomy, ileocecectomy (10 cm removed by general surgery), left salphingo-oophorectomy, and lysis of adhesions. Intra-operative findings included a perforated appendicitis, pelvic abscess, TOA, uterine didelphys, and severe adhesions throughout and extending to the liver capsule—infectious disease consult post-operatively suspected Fitz-Hugh-Curtis syndrome. The patient did well and was discharged five days after surgery. Two months later, the patient noted resolved abdominal pain, regular cycles on oral contraceptive pills, and denied vaginal discharge or other symptoms.

Conclusion: Appendicitis can overlap in presentation with several conditions, especially if pain is in an atypical location and there are co-existing gynecologic conditions. Managing complex cases like this one requires collaboration between different specialties (e.g., infectious disease, internal medicine, general surgery, gynecology, gynecologic oncology) for optimal care. Emphasis on the importance of routine gynecologic care (e.g., pap smears of both cervices, sexually transmitted infection screening and treatment) is crucial in patients with HWW.

Funding: No disclosures.