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Simultaneous development of ulcerative colitis in the colon and sigmoid neovagina

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Abstract Vaginoplasty using sigmoid colon is a common technique for creation of a neovagina. However, special consideration must be given to potential long term consequences of using a colonic conduit for vaginal replacement. We report on the youngest described case in which a patient developed ulcerative colitis refractory to medical therapy with simultaneous involvement of a sigmoid neovagina requiring total proctocolectomy and neovaginectomy. A 17 year old XY female with a history of gonadal dysgenesis and sigmoid graft vaginoplasty presented with a history of bloody, mucoid vaginal discharge, abdominal pain, bloody diarrhea and weight loss. Colonic and neovaginal biopsies demonstrated active colitis with diffuse ulcerations, consistent with ulcerative colitis. Despite aggressive immunosuppressive treatment she had persistent neovaginal and colonic bleeding requiring multiple transfusions, subtotal colectomy and ultimately completion proctectomy and neovaginectomy. It is imperative to recognize that colectomy alone may be an inadequate surgical intervention in patients with ulcerative colitis and a colonic neovaginal graft and that a concomitant neovaginectomy may be integral in providing appropriate treatment.

Vaginoplasty using the sigmoid colon is a common technique for creation of a neovagina given the advantage of spontaneous mucous production, similarity to actual vaginal tissue, and appropriately sized diameter and length [1]. Ulcerative colitis (UC) affecting the neovagina is a rare phenomenon previously reported in only four adult patients [1–4]. Of these reports, resolution of symptoms in the diseased vaginoplasty was attained with oral sulfasalazine [2], topical therapy [1,4] or no treatment at all [3]. Half of the patients who had inflammation of the neovagina and colon

Abbreviations: UC, ulcerative colitis; INR, international normalized ratio; PCR, polymerase chain reaction; AST, aspartate aminotransferase; ALT, alanine transaminase; AP, alkaline phosphatase; GGT, gamma-glutamyl transpeptidase; IBD, inflammatory bowel disease.

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required subtotal or total colectomy; [1,3] yet, none required a surgical intervention for the neovagina.

1. Case Report

An XY female with gonadal dysgenesis who was born with ambiguous genitalia consisting of bilateral descended testicles contained in the labioscrotal folds, a persistent urogenital sinus, and a blind short vaginal pouch underwent vaginal replacement at four months of life using a 12 cm segment of sigmoid colon. Aside from expected primary amenorrhea, she was otherwise healthy until she presented at 17 years of age with a two month history of progressively worsening bloody, mucoid vaginal discharge, two weeks of abdominal pain with bloody diarrhea, and nonbilious, nonbloody emesis. Two weeks prior to this presentation, she had visited the emergency department with complaints of intermittent, crampy abdominal pain and increased stool frequency. There were no reports of blood or mucous in her stool or change in stool consistency. At that time she had an unremarkable laboratory evaluation, including complete blood count with differential and complete metabolic panel. An abdominal and pelvic CT scan obtained at that visit revealed thickening of the ascending colon. Physical examination the week prior to admission during an outpatient visit was significant for mild tenderness to palpation of the lower abdomen and two symmetrically located, erythematous ulcerations of her perineum. The lesions were approximately 1 cm in diameter and located 2 cm inferior and 3 cm lateral to her introitus.

On the day of admission, the patient was having sharp abdominal pain in the epigastrium and crampy bilateral lower quadrant pain. She complained of urgency and tenesmus and was defecating approximately every 30–45 min around the clock. She had lost approximately five pounds over the past month and reported fever as high as 39 °C for the last three days with associated anorexia and fatigue. She denied a family history of gastrointestinal or autoimmune disease. On physical examination, she had kissing ulcers of the perineum with moderate neovaginal prolapse and mucoid, bloody discharge at the introitus as well as moderate epigastric and lower abdominal tenderness to palpation. Laboratory values were significant for a leukocytosis of 33.5 K/μL (3.8–10.5 K/μL) with bandemia of 32% (0%–6%), thrombocytopenia of 77,000 K/μL (150–400 K/μL) and hypoalbuminemia of 3.0 g/dL (3.3–5 g/dL). Coagulation profile revealed a prolonged prothrombin time 16.7 s (9.8–14.3 s), decreased partial thromboplastin time 23.9 s (25.2–36 s) and elevated INR 1.44 (0.87–1.24). C-reactive protein was significantly elevated, 125 mg/L (0–5 mg/L). Patient's hemoglobin 13.2 g/dL (11.5–15.5 g/dL), hematocrit 37.5% (34.5%–45%), and remaining laboratory studies were within normal limits. Due to the possibility of an infectious etiology and disseminated intravascular coagulation syndrome the patient was started on a pediatric appropriate intravenous broad spectrum antibiotic, Piperacillin–Tazobactam, and aggressively fluid resuscitated. Blood, urine and stool cultures were negative as well as a *Gonococcus–Chlamydia* Gen Probe and *Clostridium difficile* PCR.

A flexible sigmoidoscopy was performed in an unprepared bowel due to the fragility of the colon and the poor clinical status of the patient. Biopsies of the colon revealed acute inflammation. Biopsy of the prolapsed sigmoid vaginoplasty showed non-specific mild acute and chronic inflammation. Therapy with intravenous and topical intravaginal steroids was initiated. On day five of admission, the patient's liver profile was significant for a transaminitis with AST 249 U/L (0–31 U/L), ALT 192 U/L (0–31 U/L), AP 252 U/L (40–150 U/L) and GGT of 181 U/L (5–36 U/L). Serology was obtained and there was no evidence of infectious hepatitis. Concern for primary sclerosing cholangitis arose as well as for an enterocutaneous fistula given the appearance of her perineum. Magnetic resonance imaging with cholangiopancreatography was within normal limits and the patient's transaminitis and elevated GGT resolved shortly after. Magnetic resonance enterography revealed pancolitis with increased amount of pelvic free fluid, mural thickening and enhancement of the sigmoid neovagina. A fistula was not noted but perihepatic fluid and a portal venous thrombus

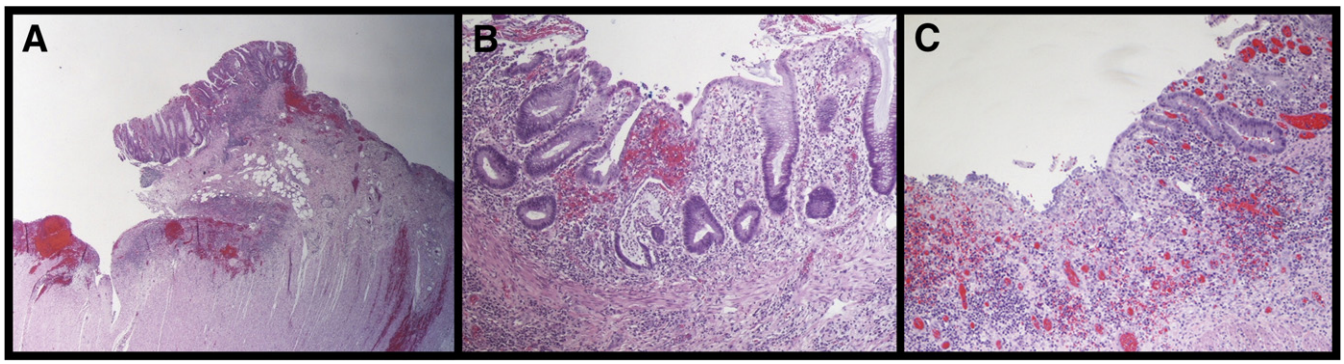

Fig. 1 (A) Neovaginal biopsy: island of intact colonic mucosa with surrounding ulceration. (B) Neovaginal biopsy: chronic active colitis. (C) Colonic biopsy: colitis with erosions, granulation tissue and extensive crypt loss.