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Case Report

Caudal duplication syndrome: imaging evaluation of a rare entity in an adult patient

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ABSTRACT

Several theories have been put forth to explain the complex yet symmetrical malformations and the myriad of clinical presentations of caudal duplication syndrome. Hereby, reported case is a 28-year-old female, gravida 2 para 2, with congenital caudal malformation who has undergone partial reconstructive surgeries in infancy to connect her 2 colons. She presented with recurrent left lower abdominal pain associated with nausea, vomiting, and subsequent feculent anal discharge. Imaging reveals duplication of the urinary bladder, urethra, and colon with with cloacal malformations and fistulae from the left-sided cloaca, uterus didelphys with separate cervices and vaginal canals, right-sided aortic arch and descending thoracic aorta, and dysraphic midline sacrococcygeal defect. Hydronephrosis of the left kidney with left hydroureter and inflammation of one of the colons were suspected to be the cause of the patient's acute complaints. She improved symptomatically over the course of her hospitalization stay with conservative treatments. The management for this syndrome is individualized and may include surgical intervention to fuse or excise the duplicated organs.

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Introduction

Caudal duplication syndrome (CDS) is a rare disease entity with prevalence of less than 1 per 100,000 at birth. It encompasses a wide spectrum of anomalies and clinical manifestations, from partial or isolated duplication of organs within the gastrointestinal (GI), genitourinary (GU), and distal neural tube system, to complete duplication. Those anomalous structures can usually be demonstrated by imaging modalities including transabdominal or transvaginal ultrasound, computed tomographic scan, and/or magnetic resonance imaging (MRI). Several hypotheses have been put forth to explain the etiology of CDS, including misexpression of homeobox (HOX) genes, an early insult to the urorectal septum, and other abnormal regression or duplication process that disrupts the embryogenesis. In most cases, reconstructive surgeries are performed to address the common complications associated with CDS including imperforate anus, fistula, and obstruction within the GI and/or GU tracts, to improve the cosmetic appearance

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in the case of genital duplication, or to help correct infertility associated with anatomic anomalies.

Case report

The patient is a 28-year-old female gravida 2 para 2 with medical history of congenital (GI) and GU system malformation who presented with acute onset of left lower quadrant pain, nausea, and vomiting for the past several days. She endorsed 3 similar episodes in the past 10 years, some of which were associated with urinary tract infections (UTI). After being examined, she started to develop continuous feculent discharge from her anus which was accompanied by some relief of her presenting pain. Surgical history is significant for serial anal dilatation for anal stenosis at birth, 2 reconstructive surgeries for anastomozing her 2 colons, and 2 caesarean sections with classical incisions.

On physical examination, her abdomen was soft with diffuse rebound tenderness but no guarding. Hyperactive bowel sounds were present diffusely across the abdomen. The patient had 2 vaginal openings with labia lateral to each vagina and a third labial fold in between. Two gluteal clefts with a fat pad in between the clefts were noted. One anal orifice was present in the right gluteal cleft; however, the anal sphincter was not appreciated on internal and external palpation. There was no perforation in the left gluteal cleft. During colonoscopy, evidence of prior surgery was seen with 2 orifices noted immediately after intubating the anus. One of them was unable to be cannulated, whereas the second one was strictured, friable, and erythematous. Fecalith of approximately 3 cm in the rectum and diffuse areas of ulcerated mucosa with pseudomembranes, particularly in the distal colon, were noted.

The patient has carried 2 pregnancies in the right uterus, and she normally uses the right vagina for intercourse. She passes urine from both the left and right vagina, although the urinary stream on the right is stronger. Prior episodes of UTI are believed to have occurred from the left urinary system. She is unable to state how she normally passes stool.

Investigations and/or imaging findings

A pelvic ultrasound was performed, followed by contrasted computed tomographic scan of the abdomen and pelvis and the pelvic MRI with and without contrast. Mullerian anomalies were demonstrated significant for uterus didelphys with 2 cervices (Figs. 1 and 2). Two urinary bladders were visualized, the left one with a trabeculated wall (Fig. 3). The left kidney demonstrated mild hydronephrosis with hydroureter which drained into the left bladder (Fig. 4A). A small nonenhancing mass just inferior to the right kidney that resembled a tiny dysplastic kidney had a subtle atrophic vascular connection to the inferior vena cava (Fig. 4B). The complete colon was also duplicated, with one colon containing normal well-formed stool. The other colon appeared distended with fluid, had wall thickening with mild surrounding inflammation, and contained a fecalith in the rectum (Fig. 4). Midline fusion defects of L5 and the sacrum were also noted (Fig. 5). The descending aorta was right sided at the diaphragmatic hiatus, and chest radiograph demonstrated a right-sided aortic arch (Fig. 6). MRI of the pelvis demonstrated cloacal malformations with fistula from the left-sided anus to the left cloaca (Fig. 7A) and additional fistula from the left cloaca to the external skin surface (Fig. 7B).

Treatment

The patient was admitted for inpatient monitoring and stabilization. Intravenous ciprofloxacin and metronidazole were administered for colitis. Her abdominal pain and anal discharge gradually resolved with the removal of the fecalith during colonoscopy and with the drainage of the colonic fluid. Over the next couple of days, she resumed normal bowel movements with formed stool. Random biopsies of the colon showed active colitis, most likely a consequence of stasis and bacterial overgrowth in the duplicated colon. The patient was provided with the recommendation from the Pediatric and Colorectal Surgery teams to have surgical intervention with removal of the duplicate colon; however, she preferred to be discharged from the hospital and be followed up as an outpatient.

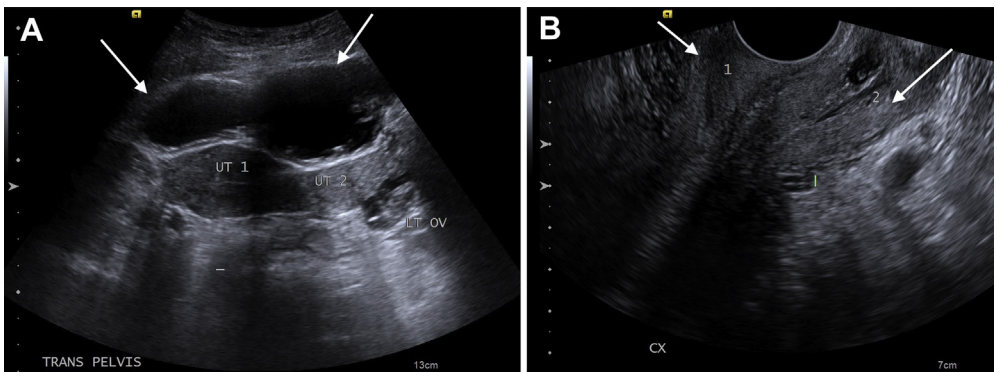

Fig. 1 – (A) Transverse transabdominal sonographic image shows duplicated uterus and bladder (arrows). (B) Transverse transvaginal sonographic image shows duplication of the cervix (arrows).

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