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Unilateral Atresia of Cervix Uteri and Uterus Didelphys in a Girl with Operated Congenital Pouch Colon

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ABSTRACT

This case report describes a girl with uterus didelphys with unilateral hematometra and hematosalpinx with congenital pouch colon (CPC) condition, where all or part of the colon is replaced by a pouch-like dilatation (5-15 cm in diameter), which communicates distally with the urogenital tract through a large fistula and associated with anorectal agenesis (supralevator anorectal malformation). CPC is seen in Asia and more specifically in northern India, Pakistan and Nepal. The management summary of this condition is creation of diverting colostomy at birth with or without the excision of pouch followed by pull through (abdominoposterior sagittal anorectoplasty) later on. The girl underwent similar procedure at birth and subsequent definitive surgery (pull through), to create a neoanus at 15 months of age. Subsequently since menarche, the child had severe cyclical abdominal pain, which on investigation was found due to hematometra and hematosalpinx in right-sided uterus. Laparotomy with removal of right-sided uterus and right fallopian tube was performed. Right-sided uterus had atresia of cervix uteri. This report emphasizes the need for comprehensive evaluation and a long-term management strategy for associated gynecologic anomalies in girls with CPC, especially with regard to patency of the outflow tract.

Keywords: Anorectal malformation, congenital pouch colon, cervical atresia, hematometra, uterus didelphys

Uterus didelphys with unilateral cervical atresia is an unusual mullerian duct anomaly with defect of vertical-lateral fusion. A well-known clinical association of congenital pouch colon (CPC) with genital anomalies exists.[1] At puberty, they present with incapacitating dysmenorrhea shortly after menarche. Similar findings in a girl with CPC have not been reported earlier in the literature.

CASE REPORT

A 10.5-year-old girl attended gynecology OPD with complaint of severe colicky abdominal pain since attainment of menarche. Pain was cyclical in nature occurring at monthly intervals. Periods were regular. There was history of multiple surgeries for associated CPC. On examination, there were three scars on abdomen. Available old records of child revealed that she was a diagnosed case of CPC, for which an emergency laparotomy with ileostomy was done at age of 15 days, followed by pull through to create a neoanus operation at age of 18 months and intraoperative findings suggested a type III CPC. Closure of colostomy was done at 2 years of age. Menarche was attained at the age of 10 years and she started having cyclical pain lower abdomen, unrelieved with medications. She was subsequently imaged in the private sector.

Ultrasound (whole abdominal and pelvis) showed both kidneys, uterus as normal and cystic mass in midline and towards right, 7.7 × 4.3 cm with internal echoes with a volume of 75 cc. Left ovary was normal. Contrast-enhanced computed tomography (CECT) abdomen showed a normal uterus with tubo-ovarian mass in right adnexa probably of infective etiology. CT urography and micturating cystourethrogram showed normal urethra and urinary bladder.

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Local examination showed normal perineum with patulous vagina and normally placed neoanus with visible rim of anal mucosa.

Magnetic resonance imaging (MRI) at our institute showed uterus didelphys with obstructing hemivaginal septum with right hematometra and hematosalpinx (Fig. 1). Left uterine cervix was identified while cervix could not be identified in right uterus. Right ovary was not visualized separately. Clinical diagnosis of mullerian duct anomaly, Class IV American Fertility Society (AFS) classification of uterovaginal anomalies, unusual configuration of vertical-lateral fusion was made. Lateral fusion defect of mullerian ducts may be the cause of uterus didelphys and vertical fusion disorder might have resulted in right cervical agenesis.

Examination under anesthesia showed a normal urethral orifice, patulous vagina and normally placed patulous neoanus. Per speculum examination showed normal vagina except a partial vaginal septum in the upper third of vagina. Normal well-developed cervix was present on left side. Menstrual blood was coming through the os. Per vaginum examination showed small-sized uterus in continuation with cervix on left side. On right side, no cervix could be felt, a firm mass of 5 × 4 cm was felt. Uterine sound was put through left cervical os into uterine cavity. Left uterocervical length was 6 cm.

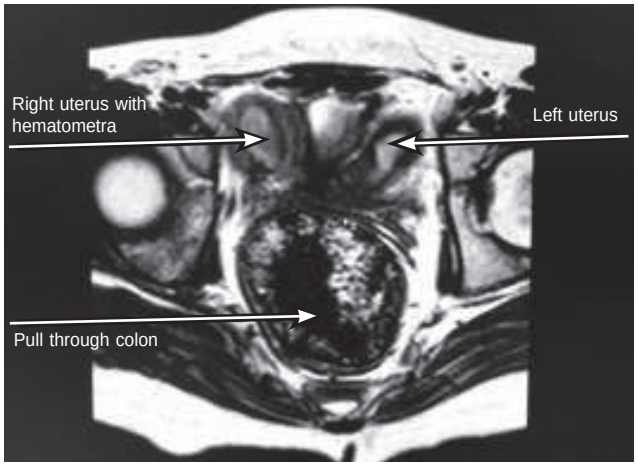

Figure 1. MRI pelvis showing uterus didelphys with obstructing hemivaginal septum with right hematometra and hematosalpinx.

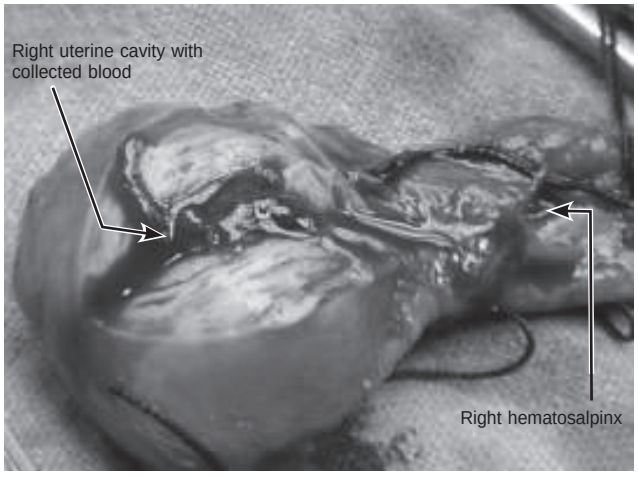

Figure 2. Cut section of removed right uterus with hematometra and hematosalpinx.

Laparotomy was done, which revealed uterus didelphys with right uterus distended with blood and a large right hematosalpinx. Right-sided ovary could not be visualized. Left-sided uterus, fallopian tube and ovary were normal. Removal of right-sided uterus and right hematosalpinx was done (Fig. 2). There was complete cervical atresia on right side. The postoperative period was uneventful.

The histopathological examination confirmed right-sided hematosalpinx and right-sided uterus with no cervix.

DISCUSSION

Mullerian ducts represent the primordial components of the female reproductive system. They differentiate into fallopian tubes, uterus, cervix and superior aspect of vagina. Overall published data suggest prevalence of uterovaginal anomalies of 1-6%.[2] Complete failure of medial fusion of two mullerian ducts can result in complete duplication, partial failure of fusion can result in a single vagina with a single or duplicate cervix and complete or partial duplication of the uterine corpus. If uterine anomaly is associated with obstruction of menstrual flow, then it causes symptoms that will come to the attention of the gynecologist shortly after menarche.[3] Early diagnosis offers significant advantages in patient care.

Incapacitating dysmenorrhea shortly after menarche with regular menstrual periods in young girls can be due to unicornuate uterus with a noncommunicating rudimentary anlagen containing functional endometrium, unilateral obstruction of a cavity of a double uterus (complete septum), a clinical syndrome consisting of a double uterus, obstruction of vagina and ipsilateral renal agenesis or unusual configuration of vertical-lateral fusion defects Class IV AFS, as in our case where there was uterus didelphys with unilateral cervical atresia.

It is important to make diagnosis as soon as possible, because if lumen of tube communicates with functional

endometrial cavity and is patent at fimbrial end, then retrograde menstruation and pelvic endometriosis can develop, destroying reproductive potential. Removal of associated fallopian tube is strongly recommended to minimize risk of an ectopic pregnancy.[3] There are very few reports of uterus didelphys, a finding that appears to be invariable in girls with types I-III CPC.[4] Chadha et al described a girl with CPC, uterus didelphys with bilateral cervical atresia. At puberty, child had primary amenorrhea with severe cyclic abdominal pain due to hematometra, hematosalpinx and endometriosis. Laparotomy with removal of both uteri was done. Both uteri had atresia of cervix uteri.[5]

In a review on the subject of gynecologic concerns in girls with anorectal malformation (ARM), Breech[6] emphasized the role of vaginoscopy before puberty, preferably at the time of definitive repair of the ARM. Vaginoscopy allows evaluation of the vaginal anatomy and can also document the appearance, development and position of the cervices in vagina and the presence or absence of mucus at the ectocervix (to infer patency). Any underdevelopment of the Müllerian structures can be detected by serial US starting soon after the onset of breast development.[6] If neglected, onset of menarche in the presence of obstruction to the outflow tract may result in hematometra and/or hematocolpos, hematosalpinx, adnexal cysts, endometriosis and chronic abdominal pain.

CONCLUSION

In conclusion, our report emphasizes the need for a comprehensive evaluation and long-term management strategy for associated gynecologic anomalies in girls with CPC, especially with regard to the patency of the outflow tract. Uterus didelphys with a septate vagina appears to be invariable in girls with types I-III CPC[4] and needs to be assessed and managed appropriately with awareness of the possibility of obstetric complications in later life.[7,8] Therefore, it is strongly recommended that the parents of such patients are duly counseled and the entire case record with operative findings are to be preserved. It is also advocated that treating pediatric surgeon should team up with gynecologist for ideal long-term management.

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AHA Recommends Dietary Screening at Routine Checkups

A new scientific statement from the American Heart Association (AHA) has recommended the inclusion of a rapid diet-screening tool into routine primary care in order to guide dietary counseling. It also recommends integration of the tool into patients' electronic health record (EHR) platforms across all healthcare settings.

The authors evaluated 15 screening tools. While no specific tool was recommended, the authors put forward the advantages and disadvantages of some of the tools. They also encouraged conversations among clinicians and other specialists to determine a tool that would be most appropriate for use in a particular healthcare setting. The authors published the statement in *Circulation: Cardiovascular Quality and Outcomes*… (*Medscape*)

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