

Uterine conserving surgery in a case of cervicovaginal agenesis with unicornuate uterus

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ABSTRACT

The presence of cervicovaginal agenesis with unicornuate uterus is a very rare mullerian anomaly. Its true incidence is still unknown. The presence of functioning unicornuate uterus poses a great challenge for a gynecologist because a successful repair could restore normal menses and may preserve a patient's fertility. Hence, we report a case of 16-year-old unmarried female who presented with chief complaints of primary amenorrhea with cyclical lower abdominal pain. On clinical and radiological evaluation, she was diagnosed with complete cervicovaginal agenesis with right unicornuate uterus (hematometra and hematosalpinx). She underwent vaginoplasty (McIndoes method) along with uterovaginal anastomosis by neocervix formation, in order to preserve her uterus. On follow-up, her vagina was completely healed, and she was menstruating normally.

KEY WORDS: Hematometra, hematosalpinx, vaginal agenesis, vaginoplasty

INTRODUCTION

Cervicovaginal agenesis in the presence of uterus is a very rare condition. Its true incidence is still unknown.^[1] The prevalence of vaginal agenesis is 1 in 4000–5000 live female births,^[2] whereas that of cervical agenesis is 1 in 80,000–10,000 live births.^[3] These patients are phenotypically females with normal female genotypes and normal endocrine status. Most of these patients have small rudimentary uterus without any endometrial cavity, but very rarely unilateral cornua may present with functioning endometrial cavity, with opposite side rudimentary cornua. The successful surgical approach till date for this condition is neovagina creation, followed by transabdominal approach to create a communication between uterus and vagina by the application of stents.

CASE REPORT

A 16-year-old unmarried female was admitted with chief complaints of primary amenorrhea with cyclical lower abdominal pain, generalized weakness, and facial puffiness. She was a known case of nephrotic syndrome since 2 years and was in remission. There was no history of any surgery. On

admission, her vitals were stable with pulse 80 beats/min and blood pressure 120/80 mmHg on the right arm. Her skeletal, cardiovascular, and respiratory system examinations were normal. Her secondary sexual characteristics were appropriate for age with breast tanner stage IV and pubic hairs tanner stage IV. On abdominal examination, no mass was palpable but deep tenderness was present. Pelvic examination revealed vaginal dimple without any vaginal orifice. On rectal examination, no mass was palpable. Her complete blood counts, serum sodium, serum potassium, and random blood sugar were within normal limits. Her serum creatinine was 1.14 mg/dl. Her urine routine microscopy examination revealed the presence of proteinuria (500 md/dl). Her abdomen and pelvis sonography revealed 41 mm × 21 mm endometrial collection

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suggestive of hematometra, along with the presence of 46 mm × 43 mm complex cystic lesion in the right adnexa suggestive of hematosalpinx [Figure 1]. Her left adnexa was normal. Her right kidney was small (31 mm × 16 mm size) with loss of cortico-medullary differentiation. Her magnetic resonance imaging (MRI) abdomen-pelvis also revealed similar findings with the presence of 38 mm × 23 mm hematometra with blind-ended uterus and absence of cervix and vagina. Her right fallopian tube was distended with largest diameter of 24 mm, findings suggestive of hematosalpinx [Figure 2]. Both her ovaries were normal and were at the level of pelvic brim. Her right kidney was small, with dimensions of 34 mm × 15 mm [Figure 3]. Her left kidney was normal. Her karyotyping was normal with 46 (XX) chromosomes.

Patient's guardians were duly counseled regarding the surgical methods, possible complications, and future fertility aspects, following which a decision of neovaginal creation

and uterovaginal anastomosis was taken. Neovagina was created by McIndoes method using partial thickness skin graft, taken from the left thigh. On laparotomy, there was right-side unicornuate uterus with right-sided hematosalpinx and left-side rudimentary horn with normal left fallopian tube. Bilateral ovaries were normal. A stab incision was given on the uterus, and hemorrhagic collection was drained [Figure 4]. Hysteroscope was introduced through the same stab incision. A nick was given from below vaginally, under the guidance of hysteroscopic light. After keeping the nick, scope was removed and Kocher's forceps was inserted from above through the uterine incision. Malecot catheter of vaginal mold under continuous suction was caught hold and was inserted by rail road method [Figure 5]. Vagina was closed over mold. Uterine incision was closed, and abdomen was closed in layers. The placement of Malecot catheter in this way served as a communication channel between uterus and vagina. On the 8th postoperative day, vaginal mold was removed,

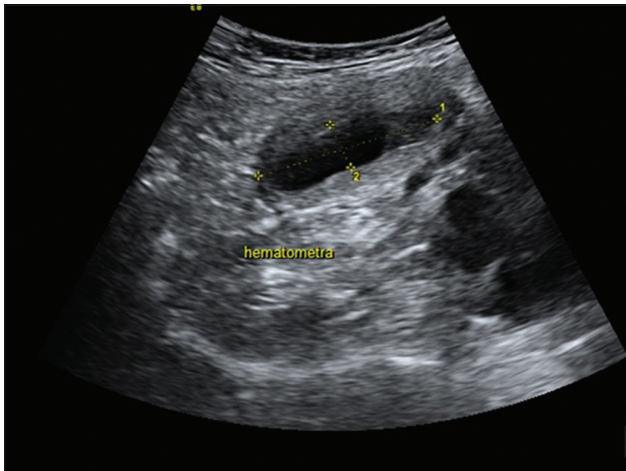


Figure 1: Sonography suggestive of hematometra along with the presence of 46 mm × 43 mm complex cystic lesion in the right adnexa suggestive of hematosalpinx

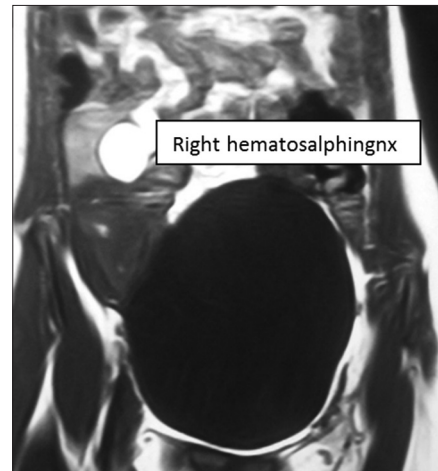


Figure 2: Magnetic resonance imaging showing the presence of hematosalpinx



Figure 3: Magnetic resonance imaging showing small right kidney

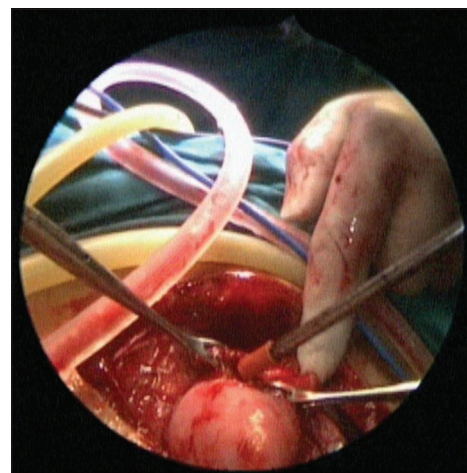


Figure 4: Drainage of hematometra after inserting stab incision over uterus

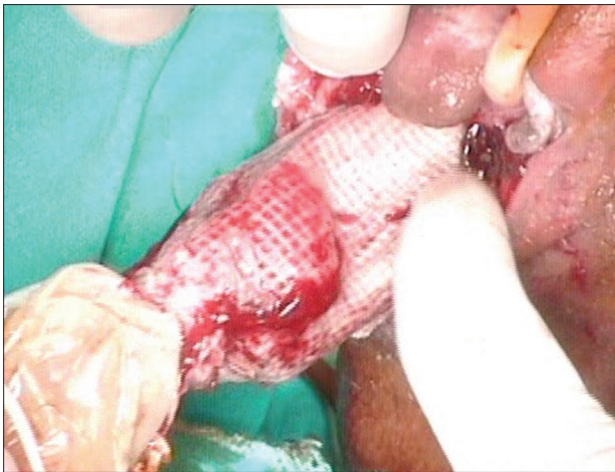


Figure 5: Placement of vaginal mold along with Malecot catheter

and diagnostic hysteroscopy was done [Figure 6]. Skin graft was healthy. A new Malecot catheter was reinserted in uterus, draining into vagina, and was fixed with silk. A new mold was placed in the vagina. The patient and her mother were educated about daily change and care of vaginal mold. She was followed up after her first menses following surgery and 6 months of surgery. Postsurgery, she was regularly menstruating with no more complaints of lower abdominal pain.

DISCUSSION

Normal vagina develops from the fusion of mesodermal müllerian duct and the endodermal urogenital sinus. Upper 2/3 of the vagina is formed by müllerian tubercle and lower 1/3 by urogenital sinus. Müllerian agenesis can be partial or complete. Partial müllerian agenesis is rare, characterized by normal uterus and small vaginal pouch distal to the cervix,^[4] whereas complete müllerian agenesis (MRKH syndrome) is the most common variant encountered. Vaginal agenesis is most commonly associated with MRKH syndrome.^[5] It is characterized with normal female genotype, phenotype, and normal endocrine status. These patients may have a rudimentary horn or total absence of uterus. Their ovaries are normal, and the secondary sexual characteristics develop normally.

Isolated uterovaginal aplasia is referred to as Rokitansky sequence or type 1 (isolated) MRKH syndrome. Incomplete aplasia associated with other malformations is generally referred to as MURCS association (or type 2 MRKH syndrome). Type 1 MRKH is less frequent than MURCS association. Type 2 MRKH is often associated with anomalies of the renal (40%–60%) system, skeletal (20%) systems, and cervico-thoracic somite dysplasia (MURCS).^[6] Renal anomalies such as unilateral agenesis of kidney, ectopic kidney, and horseshoe kidney may be present. Skeletal anomalies may include fused vertebrae or other variants.



Figure 6: Hysteroscopic view of neovagina, graft being healthy

Diagnosis is usually done by typical history of primary amenorrhea with cyclic lower abdominal pain. Clinical examination easily eliminates hymenal imperforation. Transabdominal or trans-perineal ultrasonography may specify the level of obstacle and agenesis but are not very reliable for the diagnosis. MRI seems to be the most reliable examination for the diagnosis of uterovaginal malformation and other associated malformations.^[7] Intravenous pyelography and karyotyping may help rule out other malformations.

Previously, the recommended treatment of cervico-vaginal agenesis was hysterectomy because of high failure rate of canalization procedures and risk of serious ascending infection. But this approach has been replaced by conservative approach due to recent advances in minimal surgery.^[8] Basic surgical principle is to create neovagina. Once the neovagina is completely epithelized and graft taken up, it is followed by the resection of atretic cervical tissue if present and uterovaginal anastomosis^[9] as done in our case. This surgery should be done in adolescence period as to avoid longstanding complication of hematometra and hematosalpingnx, which can lead to severe endometriosis, further hampering fertility potential.^[9] However, even after such surgery, patients on long term are at risk of stenosis, necessitating further surgery. Repeated episodes of hematometra, endometriosis, and recurrent obstruction may require hysterectomy as a last option.^[10]

There are only four cases of successful pregnancies in women with either vaginal atresia or cervicovaginal atresia described in literature so far. The case reported by Singh and Devi's had first described successful pregnancy outcome in case of neovagina.^[11] In 1996, Nargund and Parsons reported pregnancy which was achieved by *in vitro* fertilization in case of neovagina.^[12] Chakravarty *et al.* described the first case of congenital cervicovaginal atresia where the patient conceived following conservative

surgery.^[13] Acien *et al.* (2008) also described a similar case where the patient had spontaneously conceived and had delivered uneventfully.^[9]

CONCLUSION

Cervicovaginal agenesis in the presence of uterus is a rare condition. Most of the patients have small rudimentary horn, but rarely some may have unilateral cornua with functioning endometrium. Preservation of the uterus should always be attempted in such patients. There is always a possibility of pregnancy either spontaneously or through assisted reproductive technology.

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Conflicts of interest

There are no conflicts of interest.

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