

Case report

Managing the triad of triple mullerian anomaly, endometriosis and adenomyosis – A case report

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ABSTRACT

Introduction and importance: Report of an extremely rare case of triple Mullerian anomaly consisting of cervical agenesis, partial vaginal agenesis and complete bicorporeal uterus with functioning endometrium associated with adenomyosis and pelvic endometriosis in a young girl managed with hysterectomy of both uterine horns and excision of pelvic endometriosis.

Presentation of case: A 20-year-old young woman presented with primary amenorrhoea and severe cyclical pain abdomen. She was diagnosed with a rare triple Mullerian anomaly consisting of cervical agenesis, partial vaginal agenesis and complete bicorporeal uterus with functioning endometrium associated with adenomyosis and pelvic endometriosis. She had undergone laparoscopic hematosalpinx drainage in an outside setting which provided her a temporary relief from symptoms. After an accurate pre-operative diagnosis of her condition, she underwent hysterectomy of both uterine horns as both the horns were grossly adenomyotic with hematometra.

Discussion: This case is unique as two different developmental anomalies agenesis and lateral fusion defect were found together leading to a triple Mullerian anomaly with co-existing adenomyosis and endometriosis. Conservative surgery in this particular case had high probability for developing obstruction, sepsis or pelvic abscess later or repeat procedures/surgeries leading to increased morbidity. She was given the option for oocyte freezing along with gestational surrogacy if she desired fertility in future.

Conclusion: Early diagnosis and tailored intervention of Mullerian anomalies is essential to improve patients' quality of life. Definitive surgery in the form of hysterectomy should be considered if there are associated pathologies such as adenomyosis and endometriosis and findings such as hematometra.

1. Introduction

The major part of the female reproductive tract including the paired fallopian tubes, uterus, cervix and upper two thirds of the vagina develop from paired paramesonephric ducts. Mullerian duct anomalies develop from a set of structural malformations resulting from failure in organogenesis, fusion or reabsorption of these paired ducts. The spectrum of possible difficulties with these anomalies ranges from primary amenorrhoea, dysmenorrhea, chronic pelvic pain, coital difficulty, infertility, recurrent miscarriages, inability to carry pregnancy till term and low self-esteem. The prevalence of congenital uterine anomalies was reported to be 6.7 % in general population, 16.7 % in women with recurrent miscarriage and 7.3 % in infertile women in a review by Saravelos et al. [1].

Here we describe a case of triple mullerian anomaly consisting of

cervical agenesis, partial vaginal agenesis and complete bicorporeal uterus with functioning endometrium. This was associated with adenomyosis and pelvic endometriosis. Patient was managed with hysterectomy of both uterine horns and pelvic endometriosis excision to improve her quality of life. This case is being reported in accordance with SCARE guidelines [2].

2. Presentation of case

We report the case of a 20-year-old unmarried female presented with a history of primary amenorrhea and cyclic pelvic pain of seven years. The characteristic of the pain had changed to severe constant pain in the last 6 months which required parenteral analgesics every few days and emergency visits for pain relief. Four years before presenting to us she had undergone laparoscopy and drainage of hematometra.

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abdominal pain in a peripheral hospital. Following this, her pain reduced for four months and then recurred.

Physical examination revealed age-appropriate secondary sexual characters, normal female external genitalia with a blind vaginal pouch. Abdominal examination revealed an abdomino-pelvic mass reaching up to the umbilicus which was tense and tender with a smooth contour. Her ultrasound demonstrated bilateral hematometra, hematosalpinges and bilateral endometriomas with aplasia of cervix and vagina representing obstructed mullerian anomaly (Figs. 1 and 2). Renal system and karyotype were normal. There were no associated cardiac and vertebral anomalies.

A decision for laparoscopy was taken with the objective to perform pelvic endometriosis excision and uterovaginal anastomosis, if possible, with one horn which is closer to the vaginal vault after removal of the other horn. This was explained to the patient and her parents, and written consent was obtained for possible hysterectomy if anastomosis is not feasible or if the uterus appeared grossly adenomyotic.

Vaginotomy revealed blind vagina approximately 3 cm in length. The findings on laparoscopy were as follows: double uterus, both uterine horns were found to be swollen and bulky with hematometra, absent cervix and upper vagina, bilateral hematosalpinges right measuring approximately 6 × 5 cm and left measuring approximately 14 × 7 cm (Fig. 3), bilateral ovaries were higher up than the usual position with approximately 2 cm and 4 cm endometriomas in right and left ovary respectively, and dense omental adhesions with both the uterine horns and the hematosalpinges.

Omental adhesiolysis was done. Chocolate coloured fluid was drained from bilateral hematosalpinges. As both the fallopian tubes were grossly dilated and appeared non-functional, bilateral

salpingectomy was performed. Bilateral endometrioma excision was done. Both the uterine horns had hematometra and appeared grossly adenomyotic. Hence decision was taken to perform hysterectomy and both uterine horns were removed (Fig. 4). Bilateral oophorectomy was done after bringing down and fixing both the ovaries to the lateral pelvic wall below the pelvic brim. This was performed to improve the feasibility of oocyte pick up if she desires assisted reproduction in future. The patient had an uneventful recovery in the post-operative period. Histopathology confirmed adenomyosis in both horns, both cyst walls showed endometriosis and both fallopian tubes showed features of hematosalpinx and salpingitis.

Future fertility prospects were discussed with the patient and parents. The option of oocyte freezing was offered as there are chances of diminishing ovarian reserve following the endometrioma excision surgery or due to recurrent nature of the disease. Patient had a 3 cm vagina and was not sexually active, vaginoplasty was not done in the same sitting. She was advised to plan vaginoplasty if she faces coital difficulty in the future. She would require gestational surrogacy which was explained.

Patient was free from pain which she was suffering for last 7 years on follow up at four months. She had a good physical, emotional and psychological recovery after surgery.

3. Discussion

This case is unique as two different developmental anomalies agenesis and lateral fusion defect were found together leading to a triple Mullerian anomaly of cervical agenesis, partial vaginal agenesis and complete bicorporeal uterus with co-existing adenomyosis and endometriosis. Defect in organogenesis of the distal Mullerian ducts leads to cervico-vaginal agenesis and defect in lateral fusion of the paired Mullerian ducts lead to complete bicorporeal uterus [3]. The lower uterine segment narrows to terminate in a peritoneal sleeve at a point well above the normal communication with the vaginal apex. Less than 200 cases of cervico-vaginal agenesis have been reported in the literature since 1942 [4]; out of which only 7 % of the cases had functional endometrium [5]. According to the ESHRE/ESGE classification of female genital tract anomalies popularly known as "CONUTA classification" the mullerian anomaly in this patient can be classified as U3b C4 V4 [6].

Patients typically present with hematometra, disabling pelvic pain in the presence of normal secondary sexual characteristics and normal karyotype [7]. Obstruction of the menstrual flow results in development of pelvic endometriosis and adenomyosis [8]. These women may have a shortened blind vaginal pouch as the lower one third of the vagina arises from the urogenital sinus. Thus, blind vagina with abdominopelvic mass in the background of primary amenorrhoea with or without cyclical abdomino-pelvic pain should raise the possibility of either a transverse vaginal septum or vaginal/cervico-vaginal atresia; which cannot be distinguished only on clinical examination.

In these women accurate diagnosis is paramount for effective pre-surgical planning and preparation. Preoperative 3-D ultrasound or MRI aids in the diagnosis [8]. For this patient we did a 3D trans-abdominal and trans-rectal ultrasound. Nevertheless, surgical exploration confirms the final diagnosis.

These anomalies are closely related to associated malformations of the genitourinary tract in 29 % of the times necessitating renal system evaluation [3,5]. An effective psychological preparation of the patient is crucial as most of the times these are young adolescent girls who may face difficulty in having a normal sexual and reproductive life and may need corrective surgeries at different stages of their life.

Unlike most of the other mullerian anomalies, there is no well-established evidence-based surgical practice that gives the best outcome to patients with cervico-vaginal atresia. Co-existence of uterine anomalies such as bicorporeal uterus with functioning endometrium further adds to the surgical challenge of creating utero-vaginal

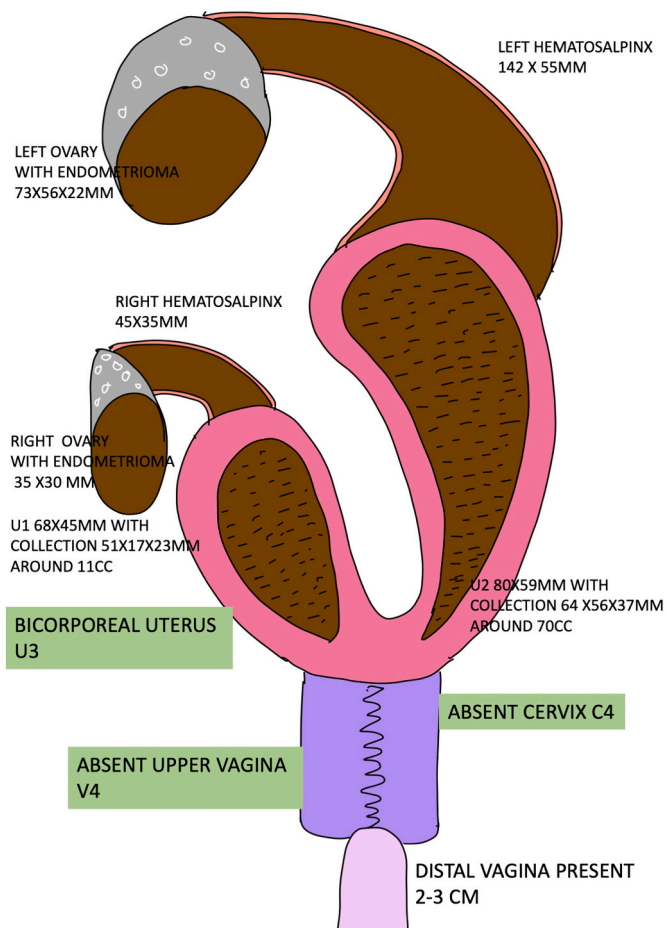


Fig. 1. Descriptive illustration of the imaging and operative findings.

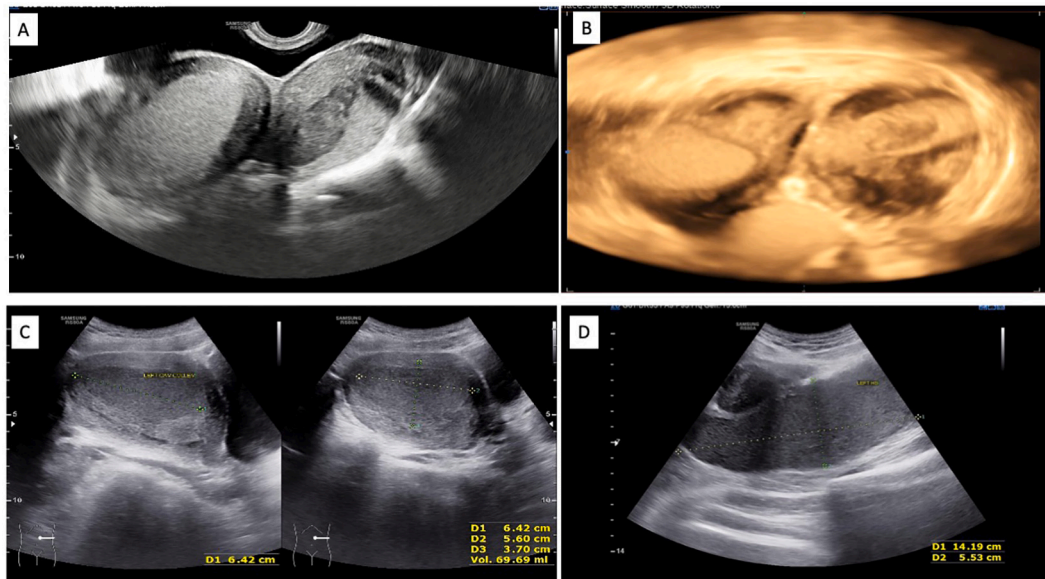


Fig. 2. Ultrasound Images of (A) Images of bicorporeal uterus with hematometra in both horns, (B) 3D image of bicorporeal uterus with hematometra in both horns, (C) Volume calculation of hematometra, (D) Left grossly dilated fallopian tube with hematosalpinx.

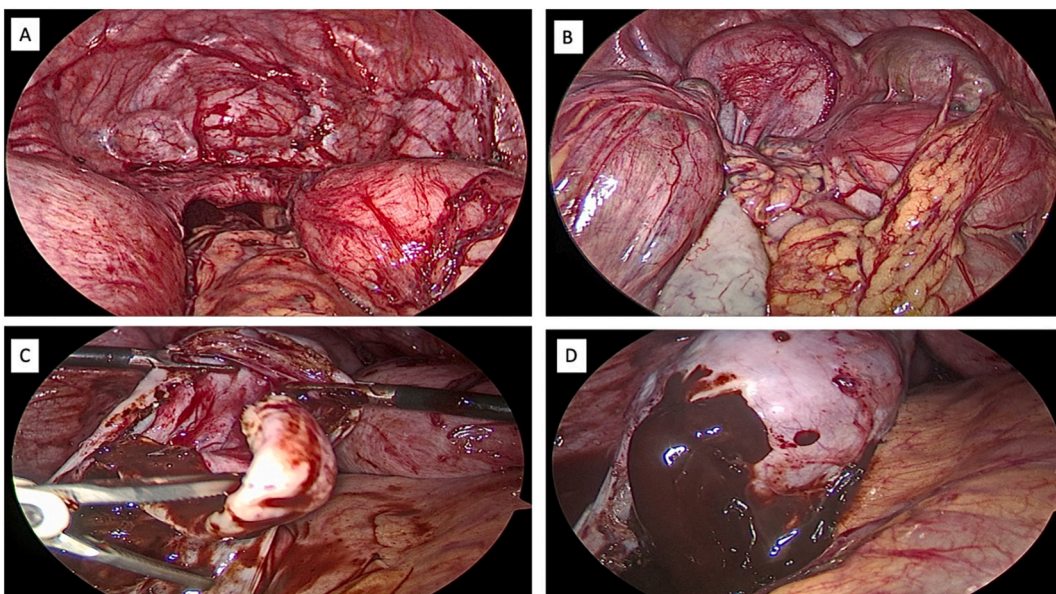


Fig. 3. Laparoscopic images of (A) Bicorporeal uterus with absent cervix, (B) Bicorporeal uterus with bilateral grossly enlarged hematosalpinges, (C) Left Endometrioma, (D) Right Endometrioma.

anastomosis. Conservative surgical management have been proposed for restoring the continuity of the genital tract such as direct uterovaginal anastomosis/canalisation, creating neocervix using small intestinal submucosal/skin graft/peritoneal flap over Foley's catheter as a plastic stent [9–13]. The aim of conservative surgeries is to restore menstruation, sexual activity, fertility and allow pregnancy till term. Small number of successful pregnancies following these procedures have been reported in the literature [7]. Although reconstructive surgeries seem ideal, significant complications are known to occur such as sepsis, pelvic inflammatory disease, chronic pelvic pain, bowel and bladder injury, re-obstruction and stenosis [10]. Successful laparoscopically assisted uterovaginal/vestibular anastomosis in patients with cervical atresia associated with partial or complete vaginal agenesis has been reported by Fedele et al. [14]. However, anastomosis was done by the authors only when normal uterine morphology was found intra-operatively and

hematometra was excluded. Kang et al. reported 4 cases of postoperative pelvic abscess developing after a mean interval of 67.7 months after primary canalisation among 54 patients who underwent fertility sparing surgeries for cervico-vaginal atresia [15]. These women eventually required hysterectomy and pelvic abscess removal. Literature reports nearly 33 % of women undergoing conservative surgeries require to undergo hysterectomy owing to one or the other complications [16]. Two cases of death following uterovaginal anastomosis due to peritonitis and septic shock at 6 days and 7 weeks after canalization have been reported [17,18]. In our case decision for hysterectomy was taken as both uterine horns were grossly adenomyotic with hematometra along with pelvic endometriosis. Conservative surgery in this particular case had high probability for developing obstruction, sepsis or pelvic abscess later or repeat procedures/surgeries leading to increased morbidity.

This case has been reported to highlight the fact that complex

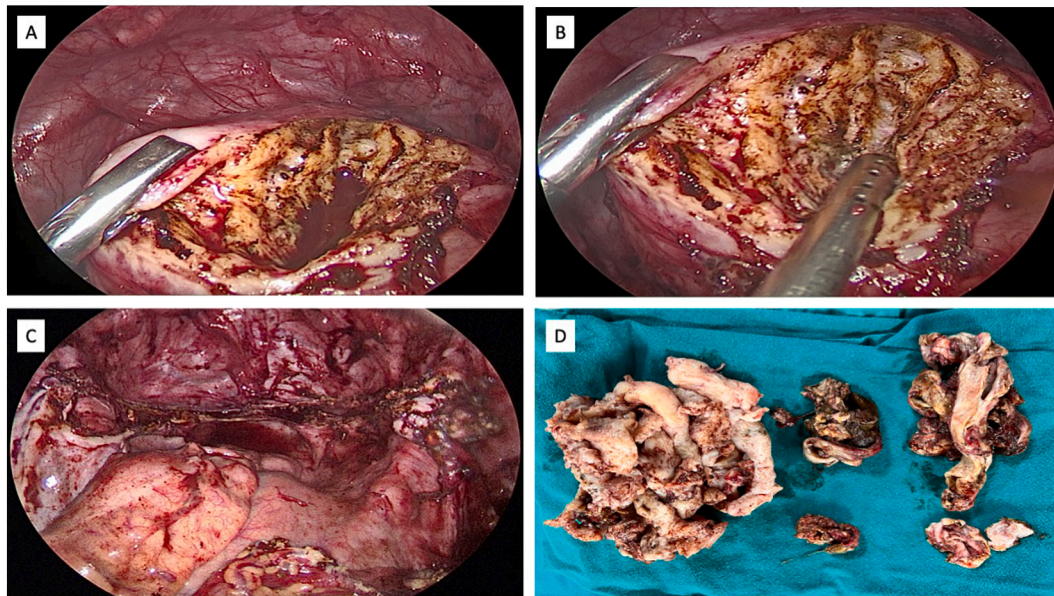


Fig. 4. (A) Hematometra, (B) Diffuse adenomyosis of uterus, (C) Final view after bicorporeal hysterectomy with ovarian preservation, (D) Specimen of bicorporeal uterus, bilateral hematosalpinges and bilateral endometriotic cyst walls.

mullerian anomalies need individualised management based on the presence of other pathologies such as adenomyosis and endometriosis.

4. Conclusions

Early diagnosis of Mullerian anomalies can improve a woman's quality of life. Conservative surgeries should not increase future morbidity for a patient and should not let them undergo repeat surgeries for complications. Thus, it is vital to tailor the management for each patient according to the diagnosis, intraoperative findings, and associated pathologies. Discussion with other colleagues pre-operatively may help in taking a proper decision for patients' benefit. Assisted reproduction can help these women to have their own offspring where conservative surgery is not an option.

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Consent

Yes.

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Guarantor

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