Case Report

Robert's Uterus-Rare Cause of Intractable Dysmenorrhea and Chronic Pelvic Pain

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Abstract

Robert's uterus is a rare type of congenital uterine anomaly where septum divides the uterine cavity asymmetrically resulting in the formation of a noncommunicating hemiuterus. Patients may present with recurrent pain, dysmenorrhea, recurrent abortions, and infertility. They are often misdiagnosed as unicornuate uterus with a noncommunicating horn. Thirty-year nulliparous female with history of severe dysmenorrhea since her menarche for which she had been operated thrice with no relief, presented with severe abdominal pain. Magnetic resonance imaging findings were suggestive of possibility of accessory horn with left hematosalpinx and hemorrhagic fluid in pelvis. Intraoperatively, it was found to be thick uterine septum completely dividing the uterine cavity asymmetrically with no communication with cervix indicative of Robert's uterus.

Keywords: *Dysmenorrhea, hemiuterus, infertility, Robert's uterus, unicornuate*

Introduction

Mullerian duct anomalies result from the defective fusion of Mullerian ducts. The defects can be segmental or can affect the whole length. The American Fertility Society, in 1998, has classified the Mullerian abnormalities into seven main classes-segmental hypoplasia or agenesis, unicornuate uterus, uterine didelphys, bicornuate uterus, septate uterus, arcuate uterus, and diethylstilbestrol-related anomalies. The septate uterus is further classified into complete septum or partial septum. Although the prevalence of septate uterus in the general population is 2.3%, the incidence of Robert's uterus is very rare with only handful of cases in published literature. Robert's uterus is a rare type of complete septate uterus (Class V(a) of the American Society of Reproductive Medicine, and $U_{2b}C_0V_0$ by the European Society of Human Reproduction and Embryology Classification, 2013)[1] characterized by uterine septum dividing the endometrial cavity asymmetrically resulting in formation of a noncommunicating hemi uterus due to obstruction by the septum. The external uterine contour is otherwise normal. It was first described and reported by Robert in 1970[2] and it is also known as asymmetrical septate uterus.

Case Report

A 30-year-old nulliparous married woman presented with severe abdominal pain and a year old magnetic resonance imaging (MRI) report of noncommunicating cavitated uterine mass with deviated uterine cavity, with differential diagnosis of unicornuate uterus with noncommunicating horn/a cystic adenomyoma. There were also hemorrhagic deposits along the vesicouterine space, anterior lower uterine serosa, posterior bladder wall, and peritoneum suggestive of deep pelvic and bladder wall endometriosis. On examination, her body mass index was 22.9 K/m^2. Abdominal examination revealed midline scar extending from pubic symphysis to umbilicus, with tenderness in hypogastrium and right iliac fossa with vague suprapubic mass of 14 weeks deviated to right side. Her speculum examination revealed healthy cervix

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and vagina. Vaginal examination revealed extremely tender 14 weeks size uterus deviated to right side with restricted mobility.

She attained her menarche at the age of 14 years which was followed by severe dysmenorrhea, for which laparotomy was done due to an episode of the acute abdomen just 5 months after her menarche. No documentation was available regarding the diagnosis or the procedure that time. The dysmenorrhea persisted and increased in intensity gradually. She had a history of a spontaneous first trimester miscarriage, followed by secondary infertility for 11 years. She also had severe deep dyspareunia with gradually decreasing coital frequency. Ten years after the first surgery, she underwent laparohysteroscopy which was inconclusive and converted into laparotomy in view of dense adhesions and bowel injury. According to the operative notes available, ovarian cystectomy was done but her symptoms persisted. She had undergone another laparotomy year later for which documents were not available. Since her menarche she was taking regular pain killers (oral and parenteral) and the frequency of same has increased in the last few years with repeated hospital admissions.

The patient and attendants requested for hysterectomy in view of her intractable pain, moribund personal and social life. Repeat MRI was done which revealed bulky uterus with lobulated outline [Figure 1a-c]. There was a thick-walled structure seen in the region of right cornu of uterus which showed cystic cavity within. The cystic area showed T1 hyperintense contents with T2 shading suggesting hemorrhagic contents. The lesion measured approximately 4.8 cm × 4.2 cm in size. The right cornu was not discernible separately. An endometrial cavity was seen displaced to the left side smoothly ending at left cornu. No definite communication was seen with the cavitated mass. There was a tortuously dilated structure in pelvis around the left ovary and pouch of Douglas showing partial septations and T1 hyperintense contents-suggesting hematosalpinx [Figure 1d]. The endocervix and the vagina showed normal signal intensities and appeared normal. Both the ovaries are well seen and normal in appearance. Differential diagnoses of cystic adenoma or an obstructed rudimentary horn were given.

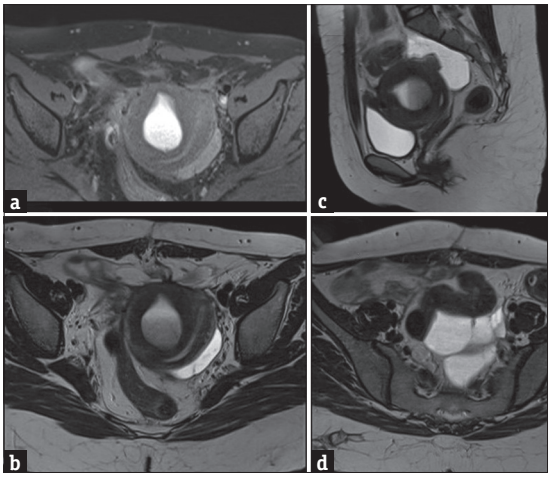

Figure 1: (a) T1W magnetic resonance imaging (axial view) showing thick walled structure seen in the region of right cornu of uterus which showed cystic cavity within. The cystic area showed T1 hyperintense contents with T2 shading suggesting haemorrhagic contents. (b): T2W magnetic resonance imaging (axial view) of Figure 1a. (c): T2W magnetic resonance imaging sagittal view: The lesion measured approximately 4.8 cm × 4.2 cm in size. Right cornu was not discernible separately. An endometrial cavity was seen displaced to the left side smoothly ending at left cornu. No definite communication was seen with the cavitated mass. (d): Dilated structure in pelvis around the left ovary and postoperative day showing partial septations and T1 hyperintense contents-suggesting hematosalpinx. The endocervix and the vagina show normal signal intensities and appear normal

Informed written consent was taken from the patient for hysteroscopy and laparoscopy accordingly with the consent of hysterectomy if needed. On laparoscopic examination, dense adhesions were seen between the uterus and the anterior abdominal wall, bowel and omentum were also adherent to it [Figure 2]. Decision for laparotomy was taken. After careful adhesiolysis, it was seen that there is a single fundus. Needle was inserted toward the right side of the fundus which revealed old tarry blood. Injection vasopressin was injected into the myometrium and incision was made over this area which drained old blood showing no communication with the rest of the uterine cavity nor with the cervical canal. It revealed oblique septum running from the fundus of the uterus toward the right side of the cavity [Figure 3]. This was confirmed by putting the dilator from below which could be felt through this long oblique septa of around 5 cm. Attendant were asked intraoperatively regarding the unification of the uterus but they refused to consent in view of previous three laparotomies. The right hemi uterus was resected along with complete shaving of

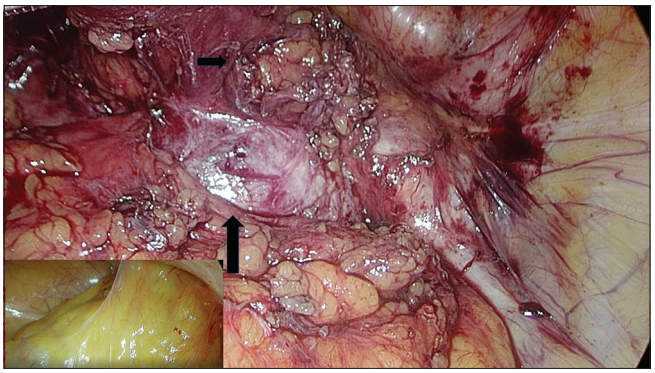

Figure 2: Laparoscopic view of uterine fundus embedded under dense adhesions

the endometrium. Myometrium over the raw area was sutured. The patient had her first menstrual bleeding 37 days after the surgery. According to her, she was significantly relieved of her dysmenorrhea and chronic pelvic pain after 3 months of surgery and satisfied with the improvement in her quality of life.

Discussion

Patients with Robert's uterus usually present with severe dysmenorrhea, recurrent abortions, infertility. Due to the obstruction by the septum, menstrual blood in the noncommunicating hemi uterus gets collected resulting in hematometra which causes severe dysmenorrhea.[1] The patients can also present with acute pain of the abdomen as an emergency.[3] Partial reflux of menstrual blood can cause hematosalpinx and even endometriosis as in this case.[4] Very few cases of pregnancy in the noncommunicating half of the uterus have also been reported.[5,6]

The accurate diagnosis of Robert's uterus is difficult. It is mostly misdiagnosed as unicornuate uterus with a noncommunicating horn as in our case three times. Ultrasound and hysterosalpingography are the initial investigations done but are less informative as ultrasound is often operator dependent, and hysterosalpingography will not show the external uterine appearance and also the noncommunicating part. MRI is the most informative noninvasive diagnostic modality in patients suspected of uterine anomalies. It will demonstrate the uterine septum as well as the normal external uterine contour which is necessary for differentiating the septate uterus from unicornuate uterus. The coronal T2W images of MRI are ideal for demonstrating the uterine septum dividing the endometrial cavity asymmetrically along with the blind ending cavity and hematometra.[7] The T1W images can show the presence of hematometra, hematosalpinx, if any. Retrospectively, MRI was re-evaluated and depiction of various structures is shown in Figure 4. In our case, the uterus appeared bulky and lobulated. There was a noncommunicating cavity on the right side of the uterus with T1 hyperintense contents and T2 shading suggestive of hemorrhagic contents. Hence, the diagnosis of obstructed rudimentary horn of the unicornuate uterus was given. To identify Robert uterus, one must look for single fundus, thick myometrium, presence of hematometra with oblique orientation of the septum joining the lateral uterine wall above internal os. Combined hysteroscopy and laparoscopy can accurately diagnose a Robert's uterus as well as differentiate it from the unicornuate uterus. The fundal contour will be normal in Robert's uterus, while there will be a fundal cleft >1 cm in unicornuate uterus.[8]

There is not a single best modality of treatment of Robert's uterus. The surgery can be approached laparoscopically, hysteroscopically, or through a laparotomy. Total resection of the blinded cavity can be performed,[9] which was done in this case as attendant were not ready for any surgery having a chance of failure. A more conservative surgery in the form of excision of the septum with unification of the endometrial cavity can also be done.[9] Few cases of 3D ultrasound-guided hysteroscopic resection of the septum have also been reported.[10,11]

Robert's uterus should be kept as a possibility while dealing with suspected unicornuate uterus with hematometra. Primary surgery has the best chance and outcome in Mullerian anomalies, so should be dealt by an expert. Resection of hemiuterus necessitates complete

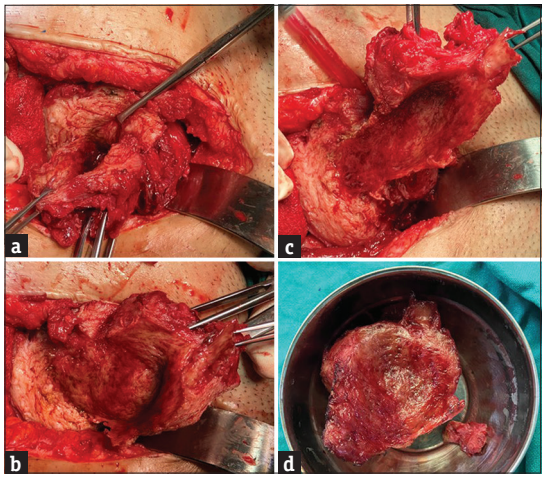

Figure 3: Peroperative images of procedure. (a): Uterine incision over the right hemiuterus showing well cavitated uterus, (b): Complete incision over the right hemiuterus showing broad medial surface of the right hemiuterus (indicative of septum confirmed by feeling the dilator on the other side). (c): Resection of the right hemiuterus with complete shaving of the endometrium from septal area to avoid any recurrence. (d): Resected specimen of right hemiuterus

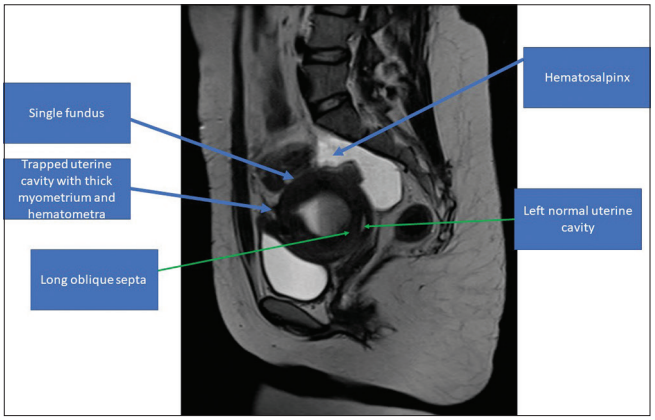

Figure 4: Sagittal T2 weighted magnetic resonance image showing the oblique septa with 2 cavities. Anterior (towards right) hemiuterus shows fluid filled levels and thick myometrial lining suggesting hematometra. The true cavity connected with vagina is seen posteriorly with thin endometrium

shaving of the endometrium from the medial aspect to avoid the chance of recurrence. The reproductive outcome will be similar to the unicornuate uterus and elective cesarean section is advisable in view of scarred uterus and reduced volume.

Conclusion

Robert's uterus is a rare anomaly and young women with Mullerian anomalies with intractable dysmenorrhea should be evaluated carefully to achieve a good reproductive outcome.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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