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Case Report

A rare case of cervical agenesis with agenesis of the upper 2/3rd vagina with hematometra

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ABSTRACT

Cervical agenesis is an extremely rare form of congenital Mullerian anomaly. Due to the rarity of the cases with different presentations ranging from cervical agenesis to dysgenesis (fragmentation, fibrous cord, obstruction of external os), along with the functionality of the uterus, there are numerous conservative surgical options involving uterovaginal anastomosis, cervical reconstruction, and cervical canalization, and total hysterectomy in cases where conservative surgical procedures fail or not feasible. In our case, the patient was a 32-year-old female with primary amenorrhea with, a history of marriage for 6 years and a history of surgery for primary amenorrhea during adolescence. During the present visit, she had severe abdominal pain due to massive hematometra with a short blind vagina. She was planned for laparoscopic Uterovaginal anastomosis but converted to abdominal hysterectomy due to the large size of uterus, lack of uterine supports, and high length of the defect between the lower end of the uterus and vaginal end. Due to the lack of proper guidelines and variations in clinical presentation, a case-based approach is required.

Keywords: Cervical agenesis, Uterovaginal anastomosis, Hysterectomy, Hematometra, Blind vagina

INTRODUCTION

Congenital cervical agenesis is a rare condition that may or may not be associated with agenesis of the vagina. It may present with renal anomalies. Historically, the American fertility society (AFS) classification has been widely used for its simplicity and ease of utilisation and it classifies cervical agenesis as Ib.[1] But due to the lack of vaginal or uterine anomaly in this classification and the lack of specific criteria for diagnosis, other classifications were advised. Recently, ASRM Mullerian anomalies classification was formulated by a task Force that looks into all the points missed in AFS classification and classifies Mullerian anomalies into nine categories.[2]

Early diagnosis is beneficial for the reconstruction, and also MRI helps in accurate diagnosis with the association of vaginal agenesis, which would help to plan the surgical outcome.[3]

CASE REPORT

We report a case of a 32-year-old female who presented to us with severe abdominal pain with a history of primary amenorrhea and married life of 6 years. In addition, the patient had a history of abdominal surgery done to evaluate Primary amenorrhea during her adolescent period, details of which were unavailable.

During the present visit, upon evaluation in ultrasound, a Bulky uterus with the collection was noted in the endometrial cavity, with both ovaries and kidneys normal. The patient was evaluated further for MRI Pelvis, which was suggestive of a Bulky uterus 94×77×69 mm sized with moderate hematometra 65×45×43 mm sized collection with non-visualization of the lower uterine segment and cervix suggestive of severely hypoplastic/agenesis of the cervix. The upper 1/3rd vagina was not properly visualized, possibly hypoplastic. Both ovaries were normal on MRI.

The secondary sexual characteristics were found to be normal. The hormonal evaluation was also done with the following values: S. AMH 6.41, S. FSH 6.85, S. LH 10.76, and S. TSH 0.48. In addition, 2D-ECHO was also done, which was found to be normal.

Upon local examination, the patient had a tenderness of the lower abdomen with a vertical scar of previous surgery and a normal vaginal opening with a short vagina with a 2 cm in length.

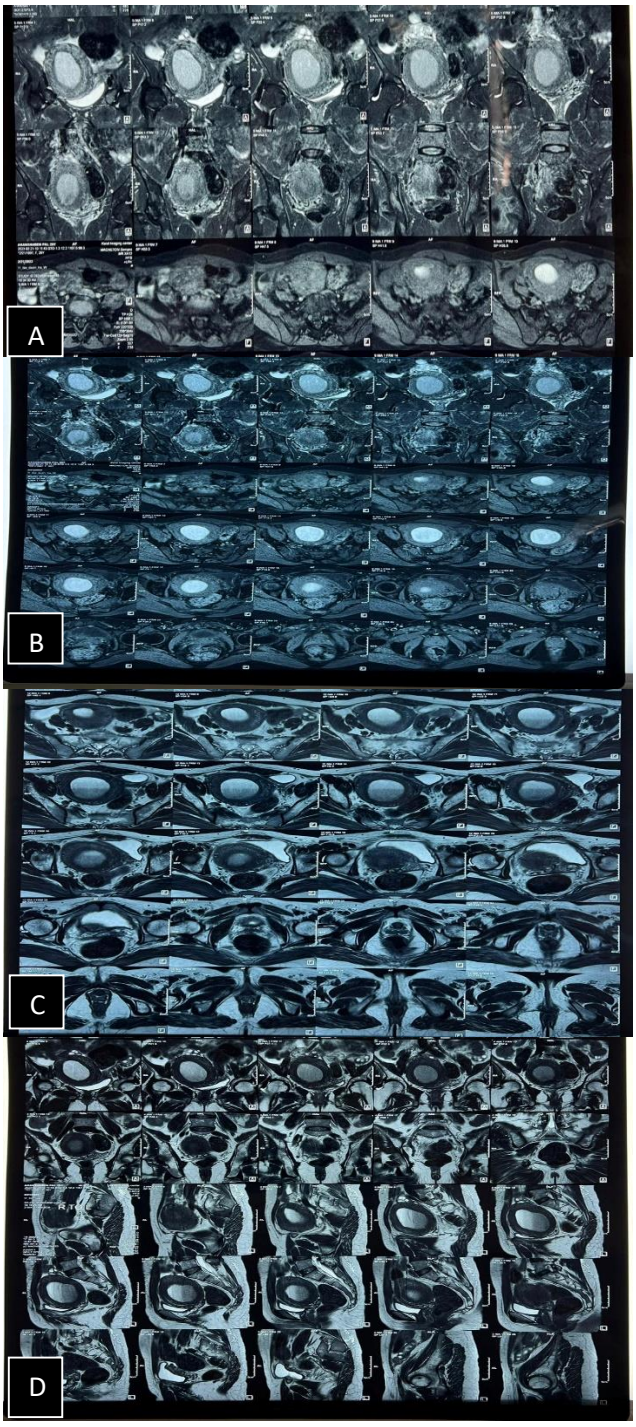

Figure 1 (A-D): MRI images: MRI pelvis of hematometra with cervical agenesis.

Operative steps done as follows with plan for conserving uterus and achieving uterovaginal anastomosis:

On exploration in laparoscopy, a bulky uterus was seen with a left ovary and fallopian tube adherent to the left lateral wall. Small bowel seen adherent over the fundus of the uterus, with right ovarian pedicle attached to posterior surface of body of uterus and adherent to retroperitoneum. Round ligaments over both sides were found to be absent.

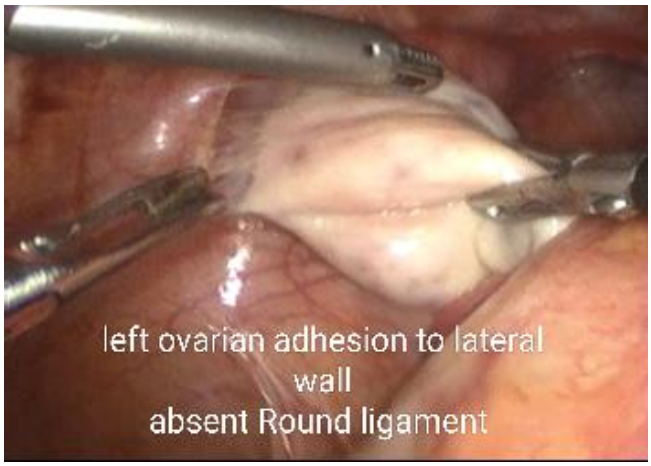

Figure 2: Adhesion of left ovary to lateral wall and absent round ligament.

Uterus was manipulated by using a Vicryl stay suture over the fundus of the uterus after injecting vasopressin diluted as 20 units in 200 ml NS.

Bowel adhesiolysis was done and dissected away from the fundus and right ovarian surface.

The right pararectal space was opened, and the right ovary was free from adhesion to the retroperitoneum. In addition, the hypogastric nerve plexus was visualized and preserved.

Bladder dissection was done, and a vertical incision was kept over the fundus and extended to the anterior wall to suction out the hematometra. Uterine walls were thick with changes of adenomyosis. The bladder was dissected further away from the level of the internal os.

On further dissection, it was seen that there was complete cervical agenesis with the absence of uterosacral attachments. Also, it was observed that there was a significant defect between the lower end of the uterus to the blind vaginal pouch.

Since there was a lack of uterine supports with the presence of a bulky adenomyotic uterus coupled with a significant defect between the lower uterine end and vaginal pouch, and the patient's insistence on a symptom-free and recurrence free postoperative period due to prior surgery and severe abdominal pain, a decision for hysterectomy with abdominal extraction of the specimen was taken.

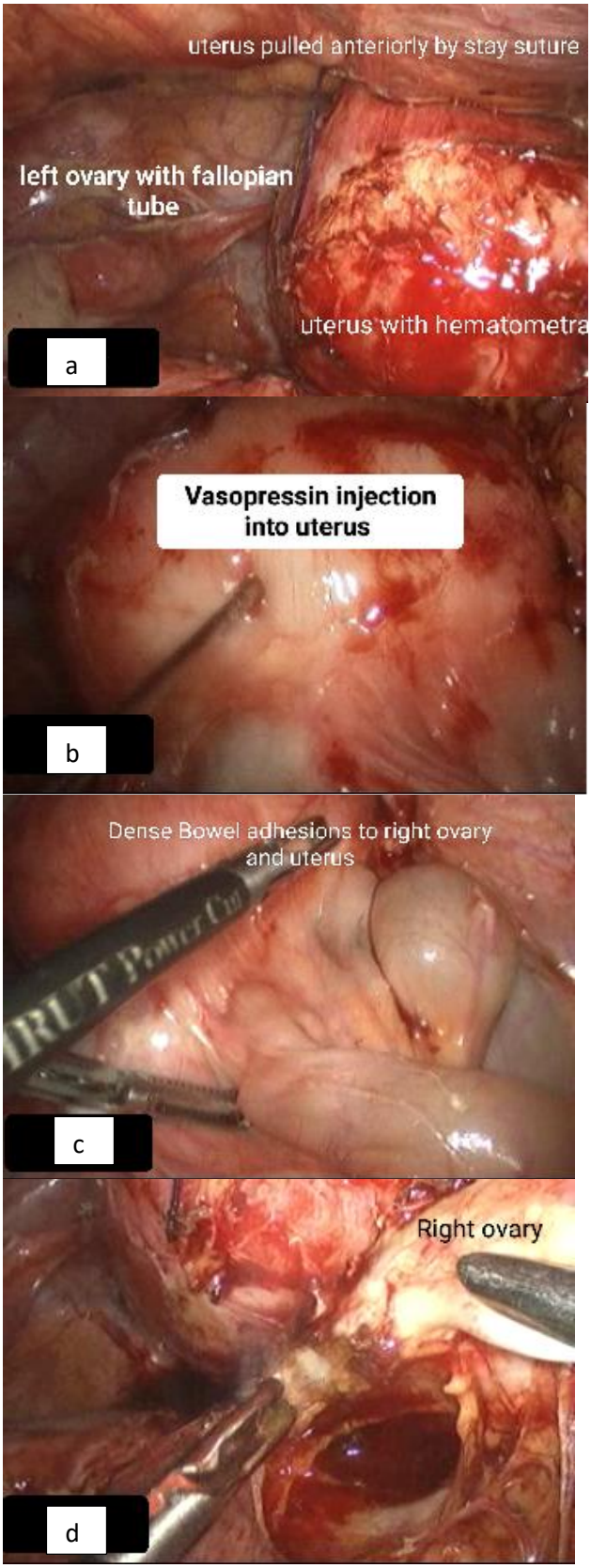

Figure 3 (a-d): Uterus with hematometra with left ovary with left fallopian tube, injecting vasopressin into uterus, right ovary densely adherent to bowel and right ovary after adhesiolysis.

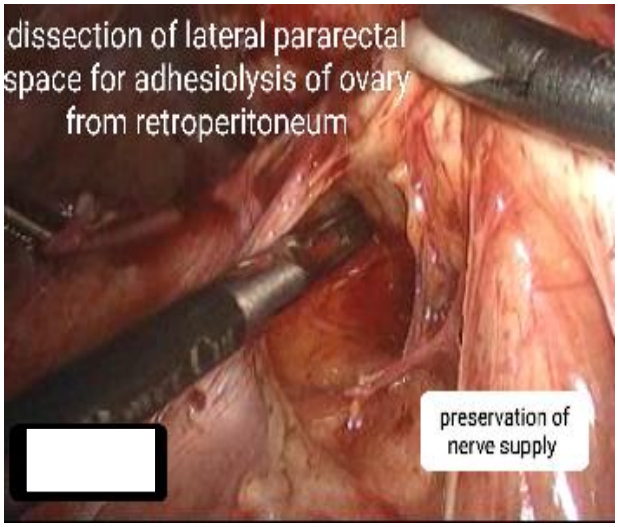

Figure 4: Lateral pararectal space dissection for removal of adhesion of right ovary.

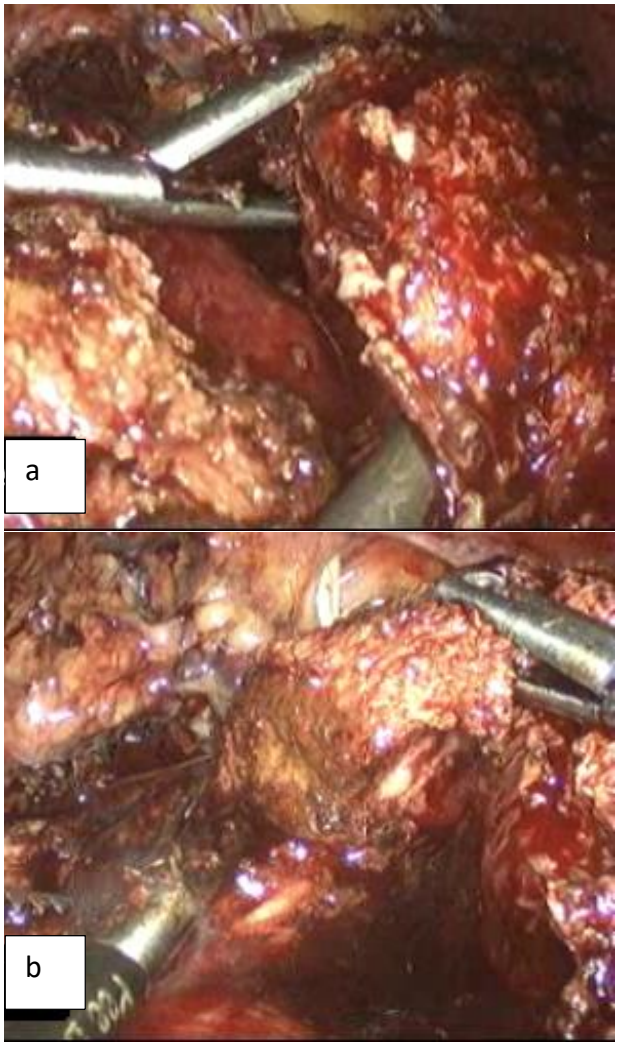

Figure 5 (a and b): Hematometra drainage by vertical incision on uterus extending from just below fundus.

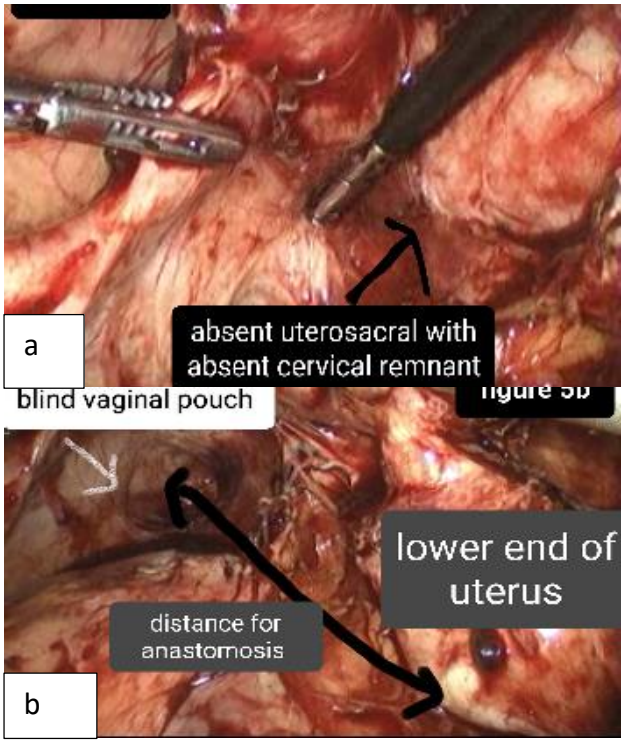

Figure 6 (a and b): Absent uterosacral ligament with absent cervical remanent and arrow showing distance between blind vaginal pouch and lower end of uterus.

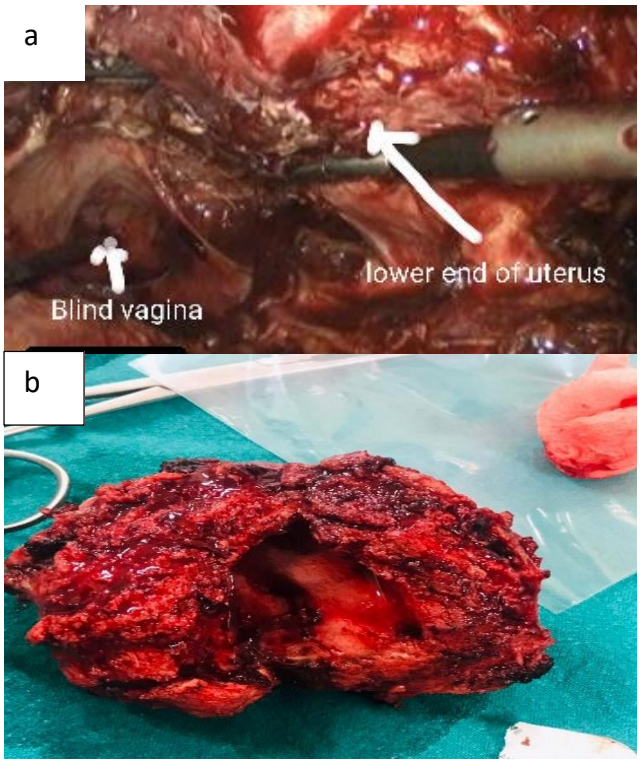

Figure 7 (a and b): Non-anastomotic distance between the blind vaginal pouch and lower end of uterus, specimen of uterus-adenomyotic uterus.

Post-operative period was uneventful, with histopathology suggestive of adenomyosis.

DISCUSSION

Cases with hematometra due to cervical atresia/agenesis have a controversial management plan due to variability in the presentation. Patients with fibrous cord/ fragmentation may have a better chance at reconstruction as compared with complete agenesis.[4] Due to complications associated with failure of anastomosis, like infection and stenosis, and also the less likelihood of attaining a viable pregnancy with cervical agenesis, hysterectomy is often preferred in cases of complete cervical agenesis.[5,6] It should be due noted that Mullerian anomalies are often associated with renal anomalies and thus should be specifically investigated during preoperative evaluation.[7]

With the advances in minimal invasive surgery, primary key goal in patients with cervical agenesis has been reconstructive surgery with uterovaginal anastomosis.[8] But due to rarity of these cases, no randomized controlled trials (RCTs) or even multicentric studies have been possible to formulate a clinical plan for operative management. Also, in the successful cases of Uterovaginal anastomosis it has been observed that the following factors play a pivotal role like early age at diagnosis, presence of a functioning endometrium, presence of uterine supports like round ligament, uterosacral ligament attached to a fragment/remnant of cervix, and the short defect allowing the creation of a tension free anastomosis.[8-10]

CONCLUSION

Following these findings, we cite the lack of early diagnosis and early surgery, an adenomyotic uterus with a lack of uterine support, and the short vaginal length that favored our case for an ultimate Hysterectomy. Thus, we conclude that now that the classification of these anomalies is incorporated with the new ASRM classification, a claim-based guideline should be developed so that it can be stressed that the need for Early diagnosis and management with the appropriate defect correction and reconstructive approach is made possible for all cases. The need for hysterectomy is limited to the very few cases that need it.

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REFERENCES

1. The American Fertility Society classifications of adnexal adhesions, distal tubal occlusion, tubal occlusion secondary to tubal ligation, tubal pregnancies, müllerian anomalies and intrauterine adhesions. Fertil Steril. 1988;49(6):944-955.

2. Pfeifer SM, Attaran M, Goldstein J. ASRM müllerian anomalies classification 2021. Fertil Steril. 2021;116(5):1238-52.
3. Markham SM, Parmley TH, Murphy AA, Huggins GR, Rock JA. Cervical agenesis combined with vaginal agenesis diagnosed by magnetic resonance imaging. Fertil Steril. 1987;48(1):143-5.
4. Acién P, Acién M, Sánchez-Ferrer M. Complex malformations of the female genital tract. New types and revision of classification. Hum Reprod. 2004;19(10):2377-84.
5. Deffarges JV, Haddad B, Musset R, Paniel BJ. Utero-vaginal anastomosis in women with uterine cervix atresia: long-term follow-up and reproductive performance. A study of 18 cases. Hum Reprod. 2001;16(8):1722-5.
6. Al-Jaroudi D, Saleh A, Al-Obaid S, Agdi M, Salih A, Khan F. Pregnancy with cervical dysgenesis. Fertil Steril. 2011;96(6):1355-6.
7. van Dam MJCM, Zegers BSHJ, Schreuder MF. Case Report: Uterine Anomalies in Girls With a Congenital Solitary Functioning Kidney. Front Pediatr. 2021;9:791499.
8. Padmawar A, Syed R, Naval S. Laparoscopic Uterovaginal Anastomosis for Cervical Agenesis: A Case Report. J Minim Invasive Gynecol. 2018;25(2):334-5.
9. Alborzi S, Momtahan M, Parsanezhad ME, Yazdani M. Successful treatment of cervical aplasia using a peritoneal graft. Int J Gynaecol Obstet. 2005;88(3):299-302.
10. Gurbuz A, Karateke A, Haliloglu B. Abdominal surgical approach to a case of complete cervical and partial vaginal agenesis. Fertil Steril. 2005;84(1):217.

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