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Case Report

A rare case of cervical dysgenesis presented with endometriosis and ectopic kidney

Shivani Gupta*, Garima Yadav, Pratibha Singh, Shashank Shekhar

Department of Obstetrics and Gynecology, AIIMS Jodhpur, Rajasthan, India

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***Correspondence:**
Dr. Shivani Gupta,
E-mail: dr.shivani2686@gmail

ABSTRACT

Cervical dysgenesis is a rare cause of obstructive amenorrhoea. We are presenting a case of a 15-year-old girl presented with primary amenorrhoea diagnosed with cervical dysgenesis with left ectopic kidney and endometriosis. The patient underwent laparoscopy followed by exploratory laparotomy with total abdominal hysterectomy with left salpingectomy followed by neovaginal creation and right ureteric D-J stenting and vaginal mould insertion. Cervical agenesis treatment should aim at the re-establishment of patency of the outflow tract. A fistulous tract with stent placement has been done in some cases but eventually, hysterectomy is required in view of recurrent and severe pelvic infection is a common problem. Ovarian function can be preserved if a hysterectomy is performed timely.

Keywords: Cervical dysgenesis, Endometriosis, Hysterectomy

INTRODUCTION

Primary amenorrhoea is defined as the absence of menstruation in women aged 14 years without the development of secondary sexual characteristics or a female who never menstruates by the age of 16 years, with the presence of secondary sexual characteristics. Primary amenorrhoea can occur due to outflow obstruction. The most common cause of outflow obstruction is the imperforate hymen, the incidence being 1/1000.[1] It results from failure of perforation of the membrane between the urogenital sinus and vaginal cavity. The transverse vaginal septum can be a reason for obstruction which is a residual vaginal plate composed of the Müllerian duct and urogenital sinus.

Cervical dysgenesis is a rare cause of obstructive amenorrhoea. The incidence of cervical agenesis is around 1 in 80,000 to 100,000 births and <50 cases have been reported in world literature.[2,3] American society of reproductive medicine (formerly American fertility society) has classified cervical agenesis as a type Ib Mullerian anomaly. Cervical dysgenesis can be classified into four subtypes as a well-formed cervix with obstruction of the endocervical lumen or as a cervical cord observed with a completely obstructed endocervical canal or as fragmentation of the cervix with certain palpable areas of the cervix or the midportion of the cervix is hypoplastic with a bulbous tip.

CASE REPORT

We are presenting a case of a 15 years old, unmarried female who presented with primary amenorrhea with hematometra with deep pelvic endometriosis and ovarian endometrioma with vaginal agenesis of the mid and lower vagina and left ectopic kidney. The patient came with complaints of cyclical abdominal pain for 2.5 years, pain in the left lower abdomen for 2 years, and primary amenorrhoea. The pain was intermittent, initially, cyclical pain occurs for 2-3 days every month gradually progressing to continuous pain in the lower abdomen associated with vomiting. The patient was diagnosed with left ectopic kidney and imperforate hymen. The patient

underwent a hymenectomy 8months back for hematocolpos, in a private hospital. Menarche was not achieved even after hymenectomy.

On examination, general condition was fair, vitals were stable, pallor was present. Secondary sexual characters were normally developed. Bilateral breasts were normally developed tanner stage 3. Per abdominal findings suggestive of nontender, the regular abdominopelvic mass of ~14-16 weeks size uterus. A blind vagina was present.

The patient was admitted. Evaluation is done by laboratory and radiological investigations. Complete blood count suggestive of hemoglobin 7.5 gm%, total leucocyte count of 17830/cumm, and platelets count of 2.29 lac/cumm. Liver function tests, kidney function tests, coagulation profile, thyroid profile were all within normal limits. FSH and LH levels were 1.79 mIU/ml and 0.17 mIU/ml respectively. Ultrasonography was suggestive of the normal-sized uterus and distended vagina with the presence of fluid in the vaginal canal, suggestive of hematocolpos with imperforate hymen/distal vaginal atresia. There was a complex cystic area with internal echoes, fluid debris level is seen in left adnexa most likely hematosalpinx, ectopic left kidney was also present. MRI was suggestive of multifocal diffuse adenomyosis in the lower uterine segment and cervix with blurred and thickened junctional zone with resultant possible cervical stenosis with mild hematometra. Left ovarian endometrioma was present with deep pelvic endometriosis involving anterior aspect of uterosacral ligament, recto-uterine pouch, posterior wall of the urinary bladder (adjacent to right VUJ), and along the sub-serosal margin of the uterus. Left simple ovarian cyst with a pelvic left ectopic kidney was seen.

The patient was planned for examination under general anesthesia. Per abdomen findings suggestive of a ~16 weeks size firm abdominopelvic mass felt slightly deviated to the left with restricted mobility. Per peculum, findings revealed a 1 cm blind vaginal pouch. No lump felt through the vaginal pouch on per vaginal examination. Per Rectal examination was suggestive of a firm mass felt most likely dilated cervix with nodularity felt in POD, uterus deviated to left and is dilated. Final Impression was of hematometra with endometriosis with most likely cervical stenosis or high transverse vaginal septum with vaginal agenesis of mid and lower vagina.

Laparoscopy f/b exploratory laparotomy followed by total abdominal hysterectomy with left salpingectomy followed by neovaginal creation and right ureteric D-J stenting and vaginal mould insertion was done. Intraoperatively, uterus was distended with bilateral tubes, and ovaries were visualized. An endometriotic cyst around 4×4 cm was seen near bilateral ovaries. Adhesions were present between the anterior wall of the uterus and urinary bladder, adhesiolysis was done. The bladder wall was pushed down the lower end of the uterus visualized; cervix not visualized.

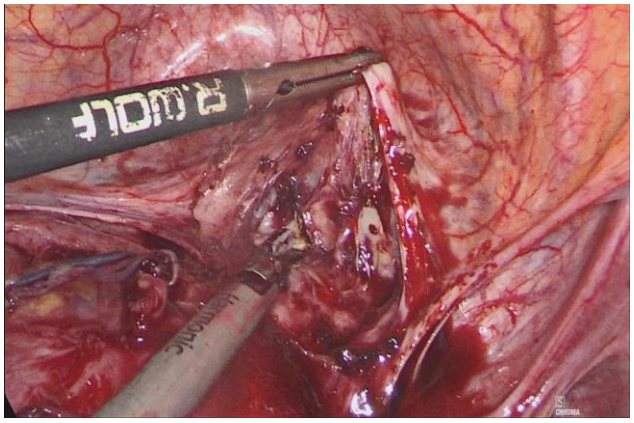

Figure 1: Adhesiolysis between bladder and uterus, bladder wall was pushed down the lower end of the uterus visualized, cervix not visualized.

Injection Vasopressin was injected intramyometrial, an incision was given over the fundus of the uterus and the uterine cavity was entered, 30 cc of old dark-colored collected blood drained→cervix was still not visualized Adenomyoma around 3×3 cm was seen over the lower uterus. Left-sided hematosalpinx and endometrioma noted→left salpingectomy and endometrioma excision done.

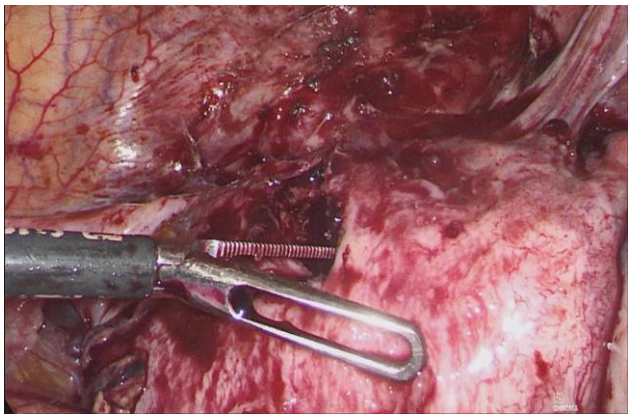

Figure 2: An incision was given over the fundus of the uterus and the uterine cavity was entered,30cc of old dark-colored collected blood drained; cervix was still not visualized.

The pelvic kidney was identified. The right adnexa was densely adherent to the uterus and lateral pelvic wall. Right side lateral peritoneum opened and right hydroureter identified, decision for laparotomy made for difficult manipulation of uterus and excessive bleeding from the incision site. Total abdominal hysterectomy with left salpingectomy done. Right DJ stenting was right-sided hydroureter, the decision for cystoscopy and right DJ stenting was taken, DJ stenting was done in the right ureter by the urologist. Dissection of the rectovaginal pouch was done to create a neovagina. Vaginal mould prepared and inserted.

Postoperatively, Injection leuprolide was given on day 7 of the surgery which was repeated after 28 days for 3 cycles. Vaginal mould reinsertion was done on POD-8 under general anesthesia.

The histopathological report was suggestive of adenomyosis with endometriosis.

DISCUSSION

Cervical agenesis is a rare Mullerian anomaly. It may be associated with the partial or complete absence of vagina. Patients with cervical agenesis may have cyclic lower abdominal pain due to retention of menstrual blood. Cervical agenesis, diagnosis before surgery is difficult but radiological imaging like MRI is helpful for preoperative diagnosis.

Cervical agenesis treatment should aim at the re-establishment of patency of the outflow tract. In cervical agenesis for passage of menstrual flow, a fistulous tract is to be obtained which is difficult to obtain. Placement of stent has been tried to keep the tract intact but later the uterovaginal tract closes by constriction by a fibrous band. Some cases report successful open pathway and normal cyclic menstruation have been reported, sperm transport is hindered due to lack of cervical mucus because of the absence of endocervical glands development. Retrograde menstruation can lead to the development of endometriosis can develop in ovaries and other pelvic tissues. Eventually, hysterectomy is required in view of recurrent and severe pelvic infection is a common problem. Ovarian function can be preserved if a hysterectomy is performed timely.

Some recent reports suggested a role of surgical uterovaginal communication in cervical agenesis which gives a chance to the patient for conception with help of advanced reproductive technologies.[8] A cerclage may prove helpful in these patients and most patients have reported delivery by cesarean section.[9] Cervical canalization can be done in patients with adequate stroma to allow a cervicovaginal anastomosis. Different surgical techniques have been recommended by various surgeons based upon the specific cervical anatomy.[10] Sometimes uterovaginal anastomosis can lead to life-threatening complications like recurrent endometritis, pelvic inflammatory disease, bowel and bladder injury, re-stenosis requiring repeat surgery.

CONCLUSION

Cervical agenesis/dysgenesis is a rare cause of cryptomenorrhea. Cervical dysgenesis imposes diagnostic challenges but is possible with current radiological imaging techniques. Patients with primary amenorrhoea should be thoroughly evaluated for this entity. Prompt and early diagnosis is important to prevent further complications mainly in the form of endometriosis due to retrograde menstruation. Corrective surgery is planned according to the type of abnormality and patient preference. The preferred treatment is hysterectomy for cervical dysgenesis.

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