

A rare case report on complete cervical agenesis with vaginal atresia and suspended didelphys uterus with hematometra and left haematosalpinx

Divya Dwivedi¹, Madhu Jain^{2*}, Shuchi Jain², Shivi Jain³

¹Department of Obstetrics and Gynecology, Trauma Centre, IMS, Banaras Hindu University, Varanasi, Uttar Pradesh, India

²Department of Obstetrics and Gynecology, IMS, Banaras Hindu University, Varanasi, Uttar Pradesh, India

³Department of Radiology, IMS, Banaras Hindu University, Varanasi, Uttar Pradesh, India

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***Correspondence:**

Dr. Madhu Jain,

E-mail: drmadhujainbhu@gmail.com

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ABSTRACT

Congenital uterine malformations are deviations from normal anatomy resulting due to defective fusion of Mullerian ducts or the paramesonephric ducts in the developing embryo. These anomalies may be isolated or in combination with urological abnormalities. The mean prevalence of female congenital malformations in general population is up to 7%. Patients with these anomalies usually present during pubertal age due to absence of onset of menses, cyclical abdominal pain, or in reproductive age group as infertility or recurrent pregnancy loss depending upon the degree of malformation. Cervical agenesis is a rare Mullerian anomaly with an incidence of 1 in 80,000 females. It represents 3% of all uterine anomalies. It is rarely associated with a functioning uterus (4.8%). Cervical agenesis is often associated with vaginal atresia (less than 50%). It is important to classify these anomalies for easy diagnosis and plan appropriate preoperative treatment.

Keywords: Cervical agenesis, CONUTA classifications, Suspensory uterus, Vaginal atresia

INTRODUCTION

Mullerian duct anomalies (MDA) occur due to the nondevelopment or defective fusion or failure of resorption of the paramesonephric (Mullerian) ducts. The prevalence of female genital tract anomalies is 4%-7% in general population and up to 8%-10% in women who have recurrent pregnancy loss.^{1,2,4} Incidence of Mullerian duct anomaly is approximately 1% in general population and it is approximately 3% in patients with infertility. Cervical agenesis or atresia or dysgenesis is an extremely rare congenital anomaly with an occurrence of about 1 in 80,000 to 100,000 births. It may be isolated or associated

with partial or complete agenesis of vagina. Diagnosis of cervical atresia is a difficult entity but the possibility of making a correct diagnosis prior to surgery do exists with the help of ultrasound and MRI.

MRI is the investigation of choice for evaluation of Mullerian duct anomaly due to its high accuracy and detailed delineation of uterovaginal anatomy. MRI has a reported accuracy of up to 100% in the evaluation of MDA.³⁻⁵

It is non-invasive and nonionizing investigation is an added advantage. Early diagnosis is advantageous in-

patient care because it offers effective presurgical planning and preparation. We have a case report of complete cervical atresia with suspended didelphys uterus diagnosed using ultrasound which were later confirmed with MRI.

CASE REPORT

An 18-year-old girl presented to us in our outpatient department with the chief complaints of on and off cyclical abdominal pain for last 6 years and absence of onset of menses. The patient was referred to us from some private hospital with history of hymenectomy performed there. The girl had normal secondary sexual characteristics and on pelvic examination a blind short vaginal pouch was found. On per rectal examination uterus was deviated towards right side and a (6 × 6) cm cystic lump separated by a groove from uterus was felt on left side.

Transabdominal pelvic ultrasound (Figure 1A) revealed the uterus with internal endometrial collection showing low level internal echoes with non-visualization of the cervix and vaginal canal. Left showed a tubular cystic lesion with internal echoes and incomplete septations suggestive of left hydrosalpinx.

Bilateral ovaries appeared normal in size, shape and location. The trans perineal ultrasonography (Figure 1B) showed presence of atretic canal measuring 4.7 cm between urinary bladder anteriorly and anorectal canal posteriorly.

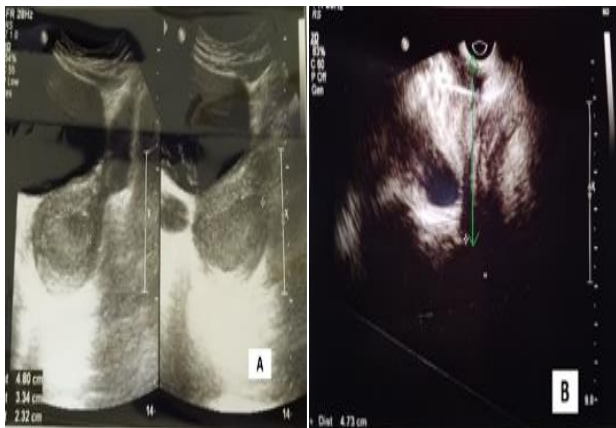


Figure 1: Transabdominal pelvic ultrasound of patient showing hematometra with non-visualisation of cervix (1A); Transperineal ultrasound showing atretic vaginal canal (1B).

The MRI of this patient revealed the same findings as on ultrasonography showing left sided (7 × 7) cm hematosalpinx with hematometra and cervical agenesis along with vaginal atresia. There was presence of a small 3x3 cm right side simple ovarian cyst, and bilateral normal ovaries (Figure 2 A-D).

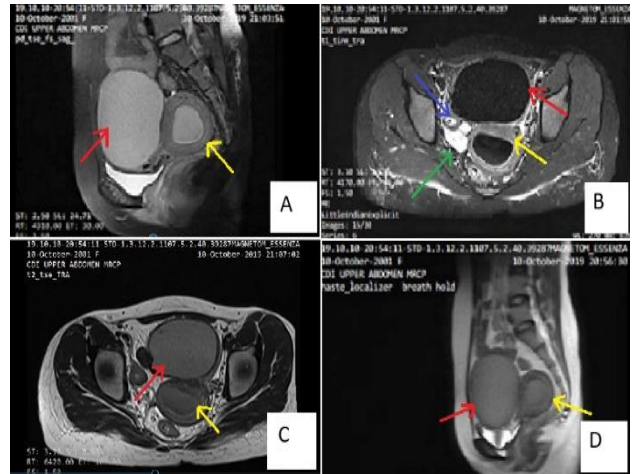


Figure 2: MRI showing presence of left sided haematosalpinx with haematometra and bilateral normal ovaries(A, B, C, D); yellow arrow points to haematometra, red arrow points to haematosalpinx, green points to right sided normal ovary (Figure 2 B), blue arrow points to rudimentary right sided uterus (Figure 2 B).

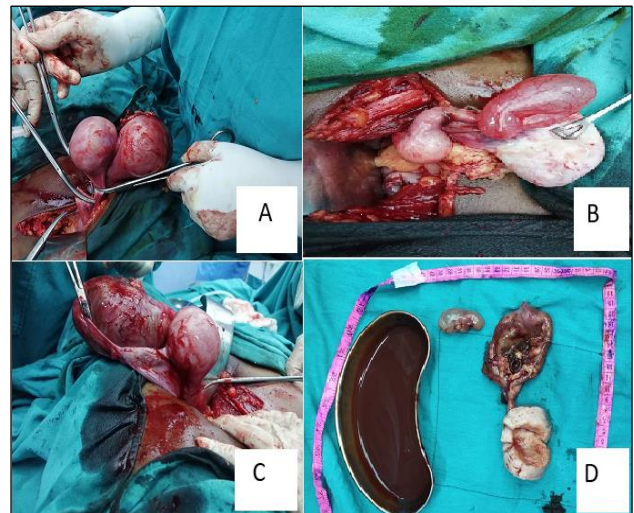


Figure 3: This is the view from anterior aspect of patient with the artery forceps pointing the left sided suspended hematometra with adhered left hematosalpinx (A); This is the right sided ovary with right sided rudimentary non-canalized uterus with right fimbrial cyst (B); This is the view from posterior aspect of the left sided suspended hematometra with adhered left ovary and left sided hematosalpinx(C); Cut section of suspensory uterus and drained hematosalpinx with chocolate content of hematosalpinx(D).

The patient was operated, and abdominoperineal repair was planned after taking proper consent and explaining the guardians of patient regarding possibility of hysterectomy. On exploratory laparotomy a large 7 × 7 cm hematosalpinx and a suspensory uterus (Figure 3A) without cervix with no connection with vagina was seen.

Left ovary was adhered to the haematosalpinx and was separated by adhesiolysis. On right side non canalised rudimentary uterus was there with fallopian tube, normal ovary and a (3 × 4) cm right fimbrial cyst (Figure 3 B).

The decision for removal of suspensory uterus was taken due to high position of uterus in pelvis and long distance between suspensory uterus and neovagina, obscuring the possibility of creation of utero-vaginal canal. Left salpingectomy was done for left sided hematosalpinx. The right sided fimbrial cyst was also removed (Figure 3 B, D).

The neovagina was created after dissecting space between urinary bladder anteriorly and anorectal canal posteriorly and an intermediate thickness split skin graft taken from patient's left thigh was grafted in vagina over soft mould. Patient was kept nil orally for 3 days and on liquid diet for 3 days and then was allowed solid diet from 6th post-operative day. The graft was taken up well by 8th post-operative day. Patient was explained to use soft mould for next 15 days followed by hard mould.

DISCUSSION

This is a case of rare uterine anomaly not reported before in the literature. The American Society of reproductive medicine classification of uterine anomalies do not include this condition. According to the ESHRE/ESGE classification of congenital uterine malformations popularly known as CONUTA classification the congenital uterine anomaly in this patient can be classified as U6 C4 V4 type of uterine malformation.⁶

Obstructive uterine anomalies occlude the normal menstrual flow resulting in primary amenorrhea. Any abdominal or pelvic pain in a pubescent girl must evoke the suspicion of possible obstructive genital anomaly. Clinically it may present with obstructive symptoms like hematometra, hematocolpos, cyclical lower abdominal pain, or may be asymptomatic depending on the functional status of the endometrium.⁷ Endometriosis can develop from retrograde menstruation in such cases.⁸ Clinical examination is helpful in identifying lower genital tract anomalies like imperforate hymen or blind vaginal pouch. However, the distinction between cervical atresia and a high vaginal transverse septum is not possible by clinical examination alone and needs MRI evaluation.¹¹

The preferred treatment modality in cervical atresia or agenesis is reconstructive surgery (uterovaginal anastomosis). The goals of reconstructive surgery are to provide a conduit for menstruation, to relieve pain and to preserve reproductive potential. Patients with atresia or cervical fragmentation are usually poor candidates for canalization and total hysterectomy is the treatment of choice.⁹ Patients with either cervical obstruction or a fibrous cord may reasonably be considered for reconstruction.^{7,10}

Controversies do exist in the treatment options and some authors describe uterovaginal anastomosis as the first line of management.^{11,12} However a complete preoperative evaluation is essential to provide useful information about the remnant cervix which serves as the basis for assessing the risks and benefits of the possible procedures to be performed. Hysterectomy was the eventual treatment for cervical agenesis because of the common complications of recanalization of the cervix and the unlikelihood of a viable pregnancy.¹³ However, hysterectomy might be necessary when the conservative treatment fails.

CONCLUSION

In our case as the functioning uterus was suspended high in pelvis with no cervix and no connection between the suspended uterus and vagina, there was no possibility of creating a utero-vaginal anastomosis, hence hysterectomy was done.

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