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# A RARE CASE OF CERVICAL ATRESIA WITH PARTIAL VAGINAL ATRESIA

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**Abstract :** Atresia of the uterine cervix is a rare Mullerian malformation which may be associated with vaginal agenesis. It is very rare in incidence, less than 60 cases is reported in literature so far. 24 Year old unmarried girl presented with primary amenorrhoea. She was already evaluated for the same in 2007, during which she was diagnosed as lower vaginal atresia and vaginoplasty has been performed then. She was advised regular vaginal dilatation with a mould. But patient had not resumed her menstruation in spite of dilatation for six months and she lost follow up further. She presented again in 2014 July with primary amenorrhoea and severe cyclical abdominal pain restricting her from activities of daily living. Per abdomen examination revealed uterus 24 weeks size. Investigation revealed bulky uterus with adenomyosis, hematometra, bilateral hematosalphinx, and bilateral endometriotic cyst. Intra op finding revealed complete cervical atresia and hysterectomy with bilateral removal of endometriotic cysts after leaving behind ovarian tissue. Mc Indo redo vaginoplasty done. To conclude, diagnosis of uterine cervical atresia requires higher clinical suspicion. Diagnosis should be made as early as possible to avoid complication that may lead to aggressive surgery such as adnexectomy or hysterectomy. Though the above patient presented to us with complications, maximum efforts had been taken to leaving behind ovarian tissue.

**Keyword :**Vaginal agenesis, Vaginoplasty, Primary amenorrhea, Cervical agenesis

**INTRODUCTION:**

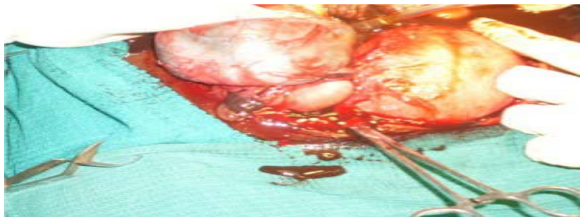
Cervical agenesis is a very rare condition often associated with atresia of vagina. Reported first by Ludwig in 1990. Less than 60 cases is reported in literature so far. Clinical diagnosis is usually difficult before surgery. Vaginal agenesis is also a rare condition that results from incomplete fusion between vaginal components of the mullerian ducts and urogenital sinus. Clinical presentation depends on whether it's partial or complete. With complete agenesis menstrual blood accumulates after puberty resulting in hematometra, hematosalphinx and ovarian endometriotic cysts. Such patients usually present with cyclical lower abdominal pain with primary amenorrhea and ultrasonographic findings of hematometra and hematosalpinx. Occasionally a lower abdominal mass (hematometra) is palpable. High suspicion is required for prompt diagnosis and management.

**CASE REPORT:**

24 Year old unmarried girl presented with complaints of primary amenorrhoea and severe abdominal pain. She was already evaluated for the same in 2007 elsewhere, where she was diagnosed as a case of lower vaginal atresia and vaginoplasty had been performed then. She was advised regular vaginal dilatation with a mould. But patient had not resumed her menstruation in spite of dilatation for six months and she lost follow up further. She presented again in 2014 July to us with primary amenorrhoea and severe cyclical abdominal pain restricting her from activities of daily living. Per abdomen examination revealed a irregularly enlarged mass of size 15 x 15 cm arising from pelvis corresponding to 24 week size of that of a gravid uterus. Ultrasonogram revealed a distended endometrial cavity with fluid with internal echoes, uterus measured 13x10x10 cm which was suggestive of haematometra. Distally vagina was imaged for a distance of 0.5 cm .Cystic lesions with internal echoes seen in both adnexa suggestive of tubo-ovarian pathology .Left sided hydronephrosis (due to pressure effect of adnexal lesion). MRI showed a similar finding along with a stenotic vagina and non visualisation of endocervical canal. Intra operative finding revealed grossly enlarged uterus, tubes and ovaries suggestive of haematometra, haematosalphinx and bilateralendometriotic cysts respectively. Cervix was totally collapsed and seen as a thin fibrous band measuring 2.8 cm along with vaginal atresia of about 3 cm. Small transverse incision was given over anterior surface of uterus and approximately 600 ml of old menstrual blood was drained. Probe passed through uterus which ended blindly. Hysterectomy was done retaining both ovaries. Vagina was atretic, dissection was done in the loose areolar tissue or space between the bladder and rectum towards the cervical band.

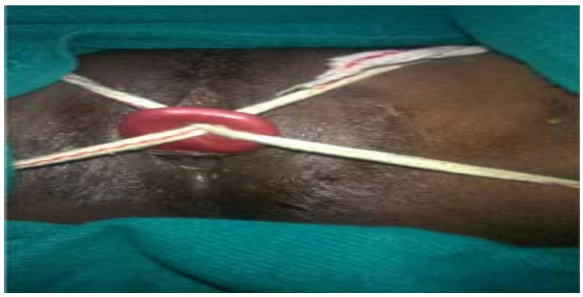
A split thickness skin graft was taken from right thigh for construction of neovagina of about 6 cm depth by Mc Indoe technique. Prosthesis was kept under aseptic precautions to maintain the depth of the neovagina. Following this period,

prosthesis was replaced. Neovagina developed with proper squamous epithelium with a depth of 6 cm in post operative period.



**Intraoperative Picture**

**Specimen Showing atretic cervix**



**Postoperative picture with vaginal mould insitu**

**DISCUSSION:**

Atresia of the uterine cervix is a very uncommon Mullerian malformation associated in 50% of the cases with a vaginal aplasia. The incidence is still unknown and less than 60 cases is reported in literature so far. Cervical agenesis arises during fetal development, during which time the paramesonephric duct fails to canalize in formation of the cervix. Any abdominal or pelvic pain which may be acute or chronic, in a pubescent girl with primary amennorhoea must evoke a diagnosis of obstructive genital syndrome. The presence of a mass inside the vagina, discovered on rectal examination, suggests blood retention above an obstacle. Clinical examination easily eliminates hymeneal imperforation or blind hemivagina, but might not differentiate cervical atresia from high vagina diaphragm.

**TYPES OF CERVICAL ATRESIA**

Type 1:
Internal os is present. Hematoma collects between the anatomical and histological os of the cervix

Type 2:
Short and Solid cervix with round end

Type 3:
Lowest region of uterus exhibited a long and solid cervix

Type 4:
Internal os absent and blind lumen

**COMPLICATIONS:**

If untreated, the accumulation of menstrual fluid in the uterus caused by cervical agenesis can lead to hematocolpos, hematosalpinx, endometriosis, endometrioma and pelvic adhesions

**INVESTIGATIONS:**

Transabdominal or transperineal ultrasonography may specify the level of the obstacle, but seems not very reliable for the diagnosis of uterine cervix atresia. Conversely, transrectal ultrasonography may help to analyse the cervix, as it provides an accurate sight of pelvic organs, and three-dimensional ultrasound may contribute to analyse the external shape of the uterus. Magnetic resonance imaging (MRI) currently appears to be the most reliable morphological examination for the diagnosis of utero-vaginal malformations with a surgical correlation. Moreover, MRI may help in the diagnosis of upper genital tract associated complications. This technique, however, is limited in case of previous surgical procedures which is very frequent in these patients. Moreover, MRI does not change radically the management of this malformation. Laparoscopic exploration has the ability to assess the type of uterine malformation, and reveals other complications of the upper genital tract that may require appropriate surgery.

**MANAGEMENT:**

Traditionally, a hysterectomy was the treatment of choice. However, the patient's desire for future pregnancy is an important consideration, as there are now options available to avoid a hysterectomy. If the complications have not set in, it can be recommended to do an utero vaginal anastomosis. The success rate of uterovaginal anastomosis is less than 50% and most patients require multiple surgeries while many develop cervical stenosis. Despite this, several pregnancies have been reported in women with cervical agenesis who underwent surgical treatment In conclusion, the diagnosis of uterine cervix atresia should be made as early as possible to avoid genital complications that may lead to aggressive surgery such as adnexectomy or hysterectomy. Utero-vaginal anastomosis appears to be feasible option; however repeated peritonitis and recanalisation for secondary stenosis may be needed. By preserving uterus, reproductive functions are conserved.

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