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Case Report

Congenital utero-cervical and vaginal atresia: a rare case report

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ABSTRACT

A genesis of cervix is a very rare developmental anomaly. It may be associated with vaginal agenesis. The usual age of presentation is teenage, with symptoms like cyclical abdominal pain and failure to attain menarche. Earlier, hysterectomy used to be the only option for such girls. Recently reconstructive surgery is gaining popularity. This is a case report presenting an eighteen-year-old girl with classical symptoms, diagnosed with both cervical and vaginal atresia, and managed by reconstructive surgery (Singapore flap method). It was a single step surgery done by abdomino-perineal approach. The girl attained menses and has been symptom free at one year follow-up. The postoperative complications were minor and managed conservatively. A patient of cervical atresia should be given the option of a reconstructive procedure even in the presence of vaginal agenesis. Utero-cervical anastomosis has shown good results in several studies. However, the reproductive potential has to be assessed by long term follow up.

Keywords: Cervical atresia, MURCS syndrome, Singapore flap

INTRODUCTION

Atresia or agenesis of the uterine cervix is a rare developmental malformation of the female genital tract. The incidence of this condition is 1 in 80000 to 1 in 100000.¹ More than half cases of cervical agenesis are associated with partial or complete vaginal agenesis.² The treatment options include conventional hysterectomy and recently, reconstructive surgery. For many decades, the mainstay of treatment of cervical malformations was hysterectomy only.³

CASE REPORT

An eighteen-year-old, unmarried girl presented with the complaints of cyclical pain in the lower abdomen and failure to attain menarche. Her past, personal and family history was uneventful. Clinical examination revealed normal secondary sexual characteristics, no thyroid swelling local examination revealed absent vaginal introitus.

Trans-abdominal 3 D ultrasound confirmed a fluid filled uterus, absent cervix and a complex cyst in right adnexa. The urinary tract was grossly normal. All other blood and hormonal work-up was within normal limits.

The patient was given the option of reconstructive surgery. With informed consent, combined cervical and vaginal reconstruction was performed using bilateral pudendal thigh flap (Singapore flap) by abdominal-perineal approach (Figure 1 and 2). Finally, the utero-vesical fold of peritoneum was incised and the bladder was reflected down. A vertical incision was made just below the fundus and the endometrial cavity was opened. A small transverse incision was given on the upper-most portion of the vaginal flap. Foley's catheter was introduced from below, made to introduce into the endometrial cavity, by-passing the atretic cervix. With the Foley's in place, the edges of uterine incision were sutured with the edges of the vaginal incision. Thus, the lower most portion of the uterus was anastomosed with the upper most part of the patent vagina.

The procedure was uneventful. Patient resumed menses 15 days after surgery. Her complaint of pain is relieved. Patient is in regular follow up since last one year. The patient had hair growth on her vagina which was managed by laser treatment (Figure 3). However, long term follow up is needed to establish this method as a first line management in case of cervical as well as the vaginal atresia.



Figure 1: Vaginal space creation.

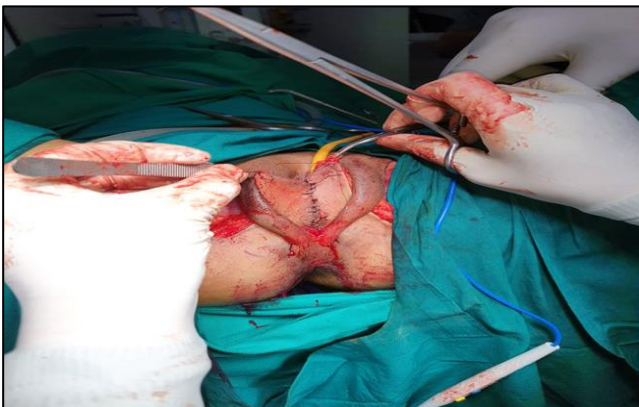


Figure 2: Flap placement.



Figure 3: Hair growth in the vagina on follow-up.

DISCUSSION

We described a rare case which has a rare mention in medical history, namely vagina atresia with cervical atresia (MURCS syndrome). Embryologically, this condition arises due to non-fusion of the lower part of Mullerian ducts. In the presence of functional endometrium, in addition to the construction of anatomically, functionally normal and sensitive neovagina and cervix, the preservation of uterus for future possible pregnancies must also be considered as the main goal in the process.⁴ In this case, combined cervical and vaginal reconstructions were successfully performed by using bilateral pudendal thigh flaps. The Singapore flap is a pedicled pudendal fascio-cutaneous flap from the thigh, first described by Wee and Joseph in 1989.⁵ The flap is raised from the tissue lateral to labia majora, based posteriorly and transposed medially. The chances of necrosis are almost negligible due to robust blood supply. There is no need for stenting, the angle of inclination of vagina is physiological and the scars of the donor site are well hidden in the groin crease. Most importantly, the sensitivity of the vagina is intact, retaining the same innervation of the erogenous zones of the perineum.⁵

In the literature, laparoscopic assisted utero-vaginal anastomosis with placement of silicone tube as stent, was done by Kriplani et al.⁶ They reported a good success rate. Similar success was reported by Fedele et al, where they followed up the patients for a period of one year.⁷ Gurlek et al reported a successful re-anastomosis by Singapore flap technique with intact vaginal sensitivity.⁴ Deffarges et al also concluded that cervico-vaginal anastomosis had good functional results.⁸

The possible complications include wound dehiscence, vaginal stenosis, infection, vulval pain or dyspareunia and very rarely, flap necrosis. However, proper case selection, expertise and early surgery minimizes the complication rate.⁹

CONCLUSION

A patient of cervical atresia should be given the option of a reconstructive procedure even in the presence of vaginal agenesis. Utero-cervical anastomosis has shown good results in several studies. However, the reproductive potential has to be assessed by long term follow up. The surgery should be performed as early as possible to avoid complications.

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