Vaginoplasty In Cryptomenorrhoea With Functioning Uterus

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Citation

S Ghose, V Rmya, S Samal, S Swain. *Vaginoplasty In Cryptomenorrhoea With Functioning Uterus*. The Internet Journal of Gynecology and Obstetrics. 2013 Volume 17 Number 1.

Abstract

A 16 year old girl presented with history of cyclical pain abdomen for two months. On examination she was found to have normal secondary sex characters. Radiological investigation revealed normal uterus with homogenous collection in endometrial cavity and normal renal anatomy. A diagnosis of functioning uterus with vaginal atresia made, which was treated by abdominoperineal approach with bilateral full thickness pudendal skin graft. She resumed her normal menstrual cycle following surgery.

INTRODUCTION

Vaginoplasty is a reconstructive plastic surgery for the vulvo-vaginal structures. Usually it is indicated for congenital disease like various degree of vaginal atresia or acquired causes like childbirth, physical trauma and malignancy of lower genital tract. This usually occurs as a part of Mayor- Rokitansky-Kuster-Hauser syndrome where uterus is also absent. But most of the patients with mullerian agenesis have small rudimentary uterus without any endometrial cavity, where as 7% - 8% may have functioning uterus. When vaginal atresia is associated with anatomically normal functioning uterus, it possesses challenge to the gynaecologist not only to reconstruct vagina but also to establish and maintain patent outflow tract. Although different surgical methods have been tried for women with Mayor-Rokitansky syndrome, options are limited for women who are having vaginal atresia with functioning uterus. Here we are reporting a case of vaginal atresia with functioning uterus which was managed and followed up successfully.

CASE HISTORY

A 16 year old girl attended the gynaecology OPD with complain of cyclical lower abdominal pain for two months. She had not attended menarche. No similar history in her sibling. On examination her secondary sex characters were well developed. There were no visual or auditory abnormalities. Abdominal examination revealed no mass. External examinations demonstrate normal labia with absent vaginal opening and no bulging (Fig.1).

Figure 1
Absent vaginal opening and no bulging



On recto-abdominal examination midline structure was present .Radiological investigations, including USG and MRI, revealed normal uterus, homogenous collection in endometrial cavity (Fig.2) and normal renal anatomy (Fig. 3).

Figure 2 MRI, revealed normal uterus, homogenous collection in endometrial cavity



Figure 3

Normal renal anatomy



So the case was diagnosed as functioning uterus with non-canalization of lower two third of vagina and planned for vaginoplasty with combined abdomino-perineal approach. On laparotomy the uterus and both ovaries were found to be of normal size (Fig.4) .There was cystic swelling at the region of cervix, which when incised anteriorly a dark brown coloured semiliquid material came out (Fig.5).

Figure 4
Normal uterus & B/L Ovaries



Figure 5
Dark semiliquid material



There was no communication downward. A passage was created from bellow starting at introitus above up to region near cervix using sharp and blunt dissection. With the help of surgeon, a bilateral full thickness pludendal thigh graft was raised (Fig.6) which was converted into a tubular shaped skin graft for vagina (Fig.7).

Figure 6Full thickness pudendal graft



Figure 7Tubular vaginal skin graft



This tubular graft was pulled into the space created for vagina and anchored to the anterior and posterior aspect of the cervix. A drainage tube was inserted into uterine cavity and a tight vaginal pack was kept. The primary skin graft site was stitched with proper haemostasis (Fig.8) and the abdomen was closed in layers. The postoperative period was uneventful. She was put on OCP for one cycle and advised for follow up in next cycle. After stopping OCP she got her period without lower abdominal pain. Endoscopy was planned after cessation of period and an os like opening was visualised (Fig.9). The patient is on follow up for last 6 months.

Figure 8
Immediate post operative result

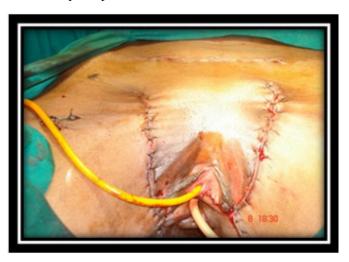


Figure 9
Vagina and cervix after one month



DISCUSSION

The vagina is a composite structure formed partly from the mullerian duct and partly from urogenital sinus. Vaginal agenesis is a congenital anomaly of the female genital tract and may occur as isolated developmental defect or as part of a complex of anomalies. Vaginal agenesis is estimated to occur in 1 in 4000 - 5000 live female births.1 It is most commonly associated with Mayor- Rokitansky-Kuster-Hauser (MRKH) syndrome and androgen insensitivity syndrome. 2, 3 Isolated defects in the development of vagina though very rare but have been reported in literature.4 The management of vaginal atresia depends on degree of atresia, associated functioning or non-functioning uterus and expectations of patient. Depending on the case, the management varies from non-surgical technique such as self-dilation with vaginal dilators (Frank's technique) to surgical techniques which involve native vaginoplasty and

foreign tissue vaginoplasty. Native vaginoplasty include Vecchietti operation (conventional or laparoscopy and Balloon vaginoplasty (Assiut innovation). Foreign tissue vaginoplasty include various graft like free skin graft, sigmoid vaginoplasty, amnion graft, pedunculated skin graft, pelvic peritoneum graft, free graft from urinary bladder, buccal mucosa, absorbable adhesion barrier. Nonsurgical techniques such as Frank's technique have been shown to have a good success rate combined with minimal risk and preferred as first-line treatment.5However this technique may not be suitable for some women. This may be due to previous multiple vaginal operations leading to significant scarring and inability of the vaginal tissue to adequately stretch by dilation alone. In addition, some women fail to dilate due to psychological difficulties despite support. For these women, surgical reconstruction of a neovagina is needed. The two most commonly performed procedures were lining of the neovaginal space with a split thickness skin graft (the McIndoe-Reed procedure) and lining of the neovaginal space with a section of intestine. Both the procedures are major and associated with significant risks and complications. The McIndoe-Reed skin graft commonly has vaginal stenosis and is troubled with unsightly scars at the harvest site. The problems associated with intestinal vaginoplasty are excessive foul-smelling vaginal discharge and diversion colitis. An increased risk of malignant change in these grafts has also been reported. Laparoscopic Vecchietti & Davydov technique is associate with stress incontinence. The expectations of patients varies with the condition involved .In cases with complete mullerian agenesis outcome expected is successful sexual intercourse .But women having segmental vaginal atresia and functioning uterus primary outcome expected is to have regular outward menstrual flow and subsequently successful pregnancy. Usually when these patient present late, they present with severe endometriosis and land up undergoing hysterectomy with BSO. Badway et al and Hafeez et al managed similar type of case with TAH with BSO.6, 7 If the cases can be detected earlier, it is possible to preserve uterus with vaginoplasty. K Dhiya et al and Fotopoulou C et al did

vaginoplasty using amnion to establish utero-vaginal continuity for girls between 15 to 18 years of age.8,9In our case we have tried the vaginoplasty using full thickness pludendal skin graft without much surgical disfigurement. We could able to attain the primary objective of patent vagina, although patient need follow up for her obstetric outcome.

CONCLUSION

Although vaginal atresia with functioning uterus is a rare condition, all adolescent girls who complain of cyclical pain abdomen with primary amenorrhoea needs prompt evaluation and early intervention. Now techniques and technologies have improved to provide not only better postoperative structural anatomy and reduced morbidity but also better quality of life.

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