CASE REPORT

Laparoscopically assisted full thickness skin graft for reconstruction in congenital agenesis of vagina and uterine cervix

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In patients with agenesis of the vagina and cervix but with a functional endometrium, the traditional treatment is hysterectomy with construction of a neovagina. We report successful treatment by laparoscopically assisted full thickness skin graft for reconstruction in a patient with congenital agenesis of the vagina and uterine cervix concomitant with haematometra and ovarian endometrioma in a 12 year old girl. Postoperatively, the vaginal skin graft healed well, and menstruation first appeared 4 weeks later. In our opinion, a combined laparoscopic and vaginal procedure with full thickness skin graft is an efficacious alternative in managing such genital defects.

Key words: agenesis/cervix/laparoscopy/skin graft/vagina

Introduction

Congenital agenesis of the uterine cervix and vagina in the presence of a functioning endometrium is a rare Mullerian duct anomaly (Rotter, 1958; Williams, 1963). This entity may also be associated with an obstructive phenomenon after menarche that leads to cyclic abdominal pain and abdominopelvic masses. Surgical reconstruction of the internal genitalia, restoration of menses, and maintenance of a patent genital tract for this congenital malformation are challenging problems for gynaecologists (Buttram, 1983). Traditionally, a hysterectomy with creation of an artificial vagina has been the treatment of choice because of the high likelihood of failure in preserving a functioning uterus and the risk of ascending infection. Here the use of a combined vaginal and laparoscopic technique is reported to create an epithelial-lined genital tract with a full thickness skin graft in a patient with congenital agenesis of the vagina and uterine cervix.

Case report

A 12 year old girl presented with a 3 month history of cyclic lower abdominal cramping pain. She had had normal pubarche, adrenarche, and thelarche starting at age 11 years, but denied ever having had vaginal bleeding. Physical examination revealed a healthy looking girl, 164 cm tall and weighing 43.7 kg. The pubic and axillary hair was normal. Pelvic examination revealed normal-appearing external genitalia but only a small shallow dimple at the vaginal introital site which invaginated 1.5 cm on pressure. There was no evidence of a bulge or haematoccolpos. Rectal examination revealed a tender globular mass in the right side of the pelvis, with a slightly enlarged uterus. An intravenous pyelogram (IVP) revealed a normal urinary tract with a soft-tissue pelvic mass impinging on the right side of the uterus. Pelvic sonography showed a large fluid-filled uterus which measured 8.1 cm in longitudinal diameter, consistent with a haematometra. The vagina was not visible. The right side of the pelvis contained a 12 cm mass that appeared to be contiguous with the uterus, and was thought to represent a blood-filled endometrioma. Magnetic resonance imaging (MRI) revealed bilateral fluid-filled tubular structures which were thought to represent haematosalpinx. No obvious uterine cervix or vagina was seen.

The serum level of CA-125 was 52 ng/dl. Serum concentrations of luteinizing hormone (LH), follicle stimulating hormone (FSH), oestradiol, prolactin and thyroid stimulating hormone (TSH) were all within normal limits. The karyotype was 46, XX. Therefore, we made a provisional diagnosis of vagina and uterine cervix agenesis associated with haematometra, right ovarian endometrioma, and bilateral haematosalpinx. After lengthy discussion with the patient and her parents, it was decided that everything possible should be done in an attempt to preserve future childbearing potential. Hence, we decided to perform vaginal and cervical reconstruction procedures, using a combined laparoscopic and vaginal procedure.

At laparoscopy, the large right pelvic mass proved to be an endometrioma with bowel adherent to its surface. The patient had an enlarged ovoid uterus. Bilateral haematosalpinx and a normal left ovary were present. Several haemosiderin deposits were noted on the pelvic peritoneal surface. Enucleation of the mass, bilateral salpingoplasty, and enterolysis were performed. Subsequently, a routine McIndoe procedure (McIndoe, 1950) was performed to create a vagina. A transverse incision was made at the hymenal ring, and the vaginal canal between the urethra and rectum was opened by blunt and sharp dissection, with the aid of laparoscopy to ensure correct orientation. The dissection was carried to the level of the lowest pole of the uterine cavity. There was no cervical...
structure on gross inspection. A tissue biopsy sample was taken and the histopathology report confirmed the absence of cervical tissue. A sounding probe was inserted into the uterine cavity through this area, and a cervical canal was made. More than 200 ml of thick, tenacious brownish material was extracted from the uterus. After these procedures, a 12×12 cm full thickness skin graft was harvested from the left groin region. The harvested skin was applied onto the cervix with 3–0 vicryl sutures. A no. 10 Foley catheter was passed into the uterine cavity through the opening from the newly created vagina to keep these canals patent, and an acrylic vaginal stent 2.5 cm in diameter and 7.5 cm in length was inserted.

After surgery, the patient remained in bed and was maintained on intravenous antibiotics for 6 days. On postoperative day 7, the vaginal stent and intrauterine Foley catheter were removed so that the condition of the neovagina and ‘cervix’ skin graft could be assessed. The graft showed no evidence of rejection. After vaginal irrigation with 10% povidone–iodine, a condom-covered Lucite mould was placed in the vagina. The following day, the patient was instructed in removal and insertion of the vaginal mould, and she was taught to wear the mould continuously for 4 months. She went home on the 9th postoperative day, and returned to the clinic regularly to ensure proper usage of the vaginal mould. Two months postoperatively, good healing of the vaginal skin graft was noted. The uppermost part of the lower artificial genital tract was somewhat scarred and fibrotic, but a no. 3 Hegar dilator could still be passed through the cervical canal without effort. The patient experienced vaginal bleeding 4 weeks postoperatively. Four months later, the vaginal mould was inserted only during sleep. The patient had regular menstruation during 1 year follow-up period. No haematometra was found on serial sonography and patency of the cervix was confirmed by insertion of a Hegar dilator at clinic visits.

Discussion

Congenital vaginal agenesis is estimated to occur in one in 5000 phenotypic females (Capraro and Gallego, 1976; Griffin et al., 1976). Approximately 8% of women with vaginal agenesis have a uterus, but half or more of these have uterine or cervical abnormalities. The incidence of vaginal and cervical agenesis with a functioning endometrium, has not been determined, because of its rarity. Our patient had agenesis of the vagina and cervix and a functional uterus concomitant with pelvic pathology. This abnormality in Mullerian duct development may represent a variant of the Mayer–Rokitansky–Kuster–Hauser syndrome.

The goal of surgical treatment should be to preserve reproductive function whenever possible. However, unlike pure vaginal agenesis, in which many successful pregnancies have been reported following vaginoplasty (Solomons, 1956; Murray and Gambrell, 1979; Fujimoto et al., 1997), absence of the vagina and cervix makes preservation of fertility difficult. Zarou et al. (1973) reported a pregnancy following the surgical correction of congenital atresia of the cervix in 1973, although there might have been some residual cervical tissue in their patient. Niver et al. (1980) studied three cases of congenital atresia of the uterine cervix and vagina in 1980. All three cases ended with total hysterectomy because of uncontrollable ascending infection from the neovagina. Thus, a majority of clinicians conclude that total hysterectomy with the McIndoe procedure (McIndoe, 1950) is the appropriate treatment for this kind of patient. Our decision to try to preserve the patient’s childbearing potential was based both on the wishes of the patient and her parents, and our belief that the risk of ascending infection could be reduced by using a full-thickness skin graft. The addition of laparoscopy to the treatment protocol offered several advantages (Lee et al., 1995), in terms of diagnosis and management.

Early and accurate preoperative diagnosis of these varieties of Mullerian duct anomalies is important and may help avoid unnecessary surgical intervention. In our case, while the sonography and MRI findings suggested the absence of the cervix and vagina with haematometra, these imaging studies could not confirm the diagnosis. The use of laparoscopy allowed this anomalous entity and the coexisting pelvic pathology to be diagnosed and treated simultaneously. Meanwhile, laparoscopy facilitated the creation of the artificial space between the rectum and bladder via the McIndoe procedure (McIndoe, 1950). Laparoscopic guidance also helped to avoid damaging the visceral organs, which might occur during blind dissection.

We used a full thickness skin graft to create the anatomical cervix, in order to avoid ascending infection and to maintain the patency of the genital tract. In this patient, the persistent patency of the cervical canal 12 months postoperatively was evident from the regular egress of menstrual effluent. This has been very encouraging; however, continued patency is as yet uncertain. Another question remains as to whether the newly created cervix will provide an effective barrier to ascending infection in the face of frequent sexual intercourse. Furthermore, if the patient were to conceive, an array of obstetric challenges will need to be addressed, particularly cervical competence. Further evaluation and follow-up are necessary.

In summary, abnormalities of Mullerian development may result in various urogenital anomalies. Accurate preoperative diagnosis of these various types is important and may avoid unnecessary surgical intervention. Hysterectomy can be avoided and fertility can be preserved in some patients with agenesis of the vagina and cervix with a functional uterus. A combined laparoscopic and vaginal procedure with a full thickness skin graft may be a reliable alternative in managing such genital defects.

References

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