Laparoscopic creation of a neovagina and recovery of menstrual function in a patient with Rokitansky syndrome: A Case Report

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A 16-year-old woman experiencing primary amenorrhoea and cyclic pelvic pain was diagnosed with Rokitansky syndrome, which was characterized by the absence of the uterus and the upper two-thirds of the vagina, normal fallopian tubes and ovaries and a 4 × 3 cm Müllerian remnant containing functioning endometrium located near the left adnexa. With a combined laparoscopic-vaginal operation, the remnant was anastomosed with the apex of the retrohymenal fovea. The operation allowed not only the creation of a neovagina but also the recovery of a regular menstrual activity and the theoretical restoration of the reproductive capacity of this patient. An accurate pre- and intra-operative evaluation of patients with Rokitansky syndrome is necessary to identify those who might benefit from this procedure.

Key words: laparoscopic surgery/Müllerian anomalies/primary amenorrhoea/Rokitansky syndrome

Introduction

Mayer–Rokitansky–Küster–Hauser syndrome is a rare congenital anomaly characterized by the lack of development of the vagina and uterus, with normal Fallopian tubes and ovaries. The treatment of this condition consists in the creation of a neovagina, allowing satisfactory sexual intercourse. Owing to the agenesis of the uterus, the only chance for these women of recovering menstrual and possibly reproductive functions relies on the future availability of uterine transplantation (Altchek, 2003).

Rokitansky syndrome, however, may present with different anatomical and clinical pictures: in particular, Müllerian remnants can frequently be observed. Usually, such remnants are represented by small non-cavitary streaks of fibrous tissue located near the adnexa or in other areas of the pelvis. In rare cases, they can resemble a small uterine corpus and contain functioning endometrium (Fedele et al., 2000; Deligeoroglou et al., 2005).

We report a case in which a Müllerian remnant containing functioning endometrium was successfully attached to the apex of the retrohymenal fovea, leading to the creation of a neovagina and the recovery of menstrual function.

Case

A 16-year-old woman was referred to our Department for primary amenorrhoea and cyclic pelvic pain. She had already undergone in another hospital a karyotype evaluation, which resulted in 46 XX, and an abdomino-pelvic magnetic resonance imaging and ultrasound, with the diagnosis of suspected Rokitansky syndrome. At ultrasound, there were no associated renal anomalies. At physical examination, she presented normal axillary and pubic hairs, normal puberal development and agenesis of the upper two-thirds of the vagina, with a 2-cm-deep retrohymenal fovea. At rectal examination, only a uterine fibrous remnant located in place of the uterus could be palpated, with no other pelvic masses. A transabdominal pelvic ultrasound confirmed absence of the uterus and demonstrated the presence of a bilateral Müllerian remnant, adjacent to the ovaries. The ovaries appeared normal. The right Müllerian remnant measured 2 × 1.5 cm and appeared non-cavitated, whereas the left-sided remnant measured 4 × 3 cm and contained endometrial echoes 25 mm long and 7 mm wide. After signing the informed consent, the patient was scheduled for diagnostic laparoscopy and possible treatment of her condition, including the anastomosis of the Müllerian remnant with the apex of the retrohymenal fovea to restore menstrual activity.

Laparoscopy was performed according to the standard technique. After induction of the pneumoperitoneum and the intraumbilical insertion of the laparoscope through a 10-mm trocar, three suprapubic ancillary ports were inserted under direct vision. The preoperative diagnosis was confirmed. The size of the left Müllerian remnant was similar to that of a small uterine corpus; no endometriosis was present (Figure 1). An intraoperative ultrasound was performed with a laparoscopic probe confirming the cavitation of the remnant containing endometrial tissue (Figure 2). The decision was taken to
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attempt anastomosis of the uterine remnant with the apex of the retrohymenal fovea.

Through a 0.5 cm hole made with a monopolar needle, a hysterometer was introduced in the cavity of the uterine remnant, pushing it down towards the vesico-rectal space (Figure 3A). Further steps described were carried out vaginally. An H-shaped incision of the retrohymenal fovea was performed, and a tunnel between the bladder and the rectum created by blunt and sharp dissection until the communication with the pelvic cavity was obtained. With the aid of an assistant pushing the remnant from above, the operator grasped it from below and entered the cavity by means of serial transverse sections of the inferior aspect of the remnant in a caudo-cranial direction and guided by the intrauterine hysterometer. The discharge of a small amount of blood from the cavity was observed. The edges of the cavity of the Müllerian remnant were anastomized with the edges of the retrohymenal incision with multiple interrupted 2–0 monocryl sutures, displacing the remnant downward (Figure 3B). A cystoscopy was performed confirming the integrity of the bladder; saline solution was placed in the Douglas pouch and the rectum was filled with air to confirm absence of rectal lesions. Operating time was 120 min and blood loss was 50 ml. Post-operative course was uneventful and the patient was discharged on the 8th post-operative day. The application of an estrogenic vaginal cream was prescribed for 15 days after surgery.

Thirty days after the operation, the patient had her first menstruation and has been having regular menses, although limited in quantity, during a 24-month-follow-up. She has not yet attempted sexual intercourse. Follow-up visits were performed every 6 months. Pelvic examination and vaginoscopy showed a 6 cm-long vagina without evidence of uterine cervix and vaginal fornices, but with a well-healed utero-vaginal anastomosis resembling a normal external uterine orifice. We were able to perform a hysteroscopy, which showed a small cavity coated with functioning endometrium. At transvaginal ultrasound, a small uterus with a normally appearing endometrial cavity was observed.

Comment

We report a case in which a Müllerian remnant was used to replace the uterus in a patient affected by Rokitansky syndrome, allowing the recovery of a regular menstrual activity. We believe that this is an important achievement in a 16-year-old woman, helping her to acquire psychological fulfillment as a woman.

We attempted to perform this operation with the aim of creating a neovagina by means of traction exerted on the vaginal fovea by the uterine remnant, whose round ligament and ovarian vessels had been stretched downwards by the anastomosis. This was based on our experience, which is consistent with previously published series (Deffarges et al., 2001), showing that a utero-vaginal anastomosis is effective in creating a neovagina in patients with cervical atresia associated with vaginal aplasia. Anatomically, the procedure was successful: a 6 cm long neovagina allowing easy introduction of two fingers was obtained within 6 months after corrective surgery. We were not able to assess functional success because the patient has not yet attempted sexual intercourse.

An extremely important implication of this operation is the potential restoration of the reproductive capacity of this patient. On the basis of the previous experiences with patients treated for cervical atresia (Deffarges et al., 2001), we believe that this patient may have the possibility to obtain a spontaneous and successful pregnancy despite the reduced size of her uterus. The procedure therefore holds the possibility of fully treating Rokitansky syndrome, restoring the anatomy and the function of both the vagina and uterus.
An operation of simultaneous hysteroplasty and vagino-plasty allowing the recovery of menstruations has been previously reported in 41 (Chakravarty, 1977; Chakravarty and Gun, 1977) and three patients (Singh and Devi, 1980), respectively. The hysteroplasty consisted of the unification of bilateral uterine remnants. All the procedures were carried out by laparotomy; the presence of functioning endometrium was not assessed preoperatively and no pregnancies were reported. To our knowledge, there has been no further report of such an operation in the last 25 years. We propose a new operation which, taking advantage of both a minimally invasive surgical approach such as laparoscopy and a very reliable diagnostic technique such as transabdominal and intraoperative ultrasoundography, could allow to improve the outcome of uterine reconstruction in selected patients with Rokitansky syndrome.

Successfully performing this operation has been possible as a consequence of the favourable anatomical characteristics of this patient, namely the presence of both a cavitated uterine remnant of adequate dimensions to replace the uterus and a 2-cm-deep retrohymenal fovea. The greatest limitation of this operation is the need for such a uterine remnant, which is unfortunately absent in most patients with Rokitansky syndrome. A retrohymenal fovea, on the contrary, is quite frequently observed in these women, and it is already known that its presence increases the possibility of creating a neovagina with the non-surgical method of Frank.

Considering that the remnant successfully used in the present case measured 4x3 cm only, we believe that an accurate preoperative and intraoperative evaluation of patients with suspected Rokitansky syndrome, especially when cyclic pelvic pain is experienced, is of paramount importance to identify those who might benefit from this procedure. This might constitute an argument for avoiding reconstructive surgery in children; before puberty, in fact, small uterine remnants may more easily be overlooked.

In conclusion, a cavitated uterine remnant was used for replacing the uterus in a patient with atypical Rokitansky syndrome, achieving the restoration of menstrual function. This operation holds the theoretical possibility of a successful pregnancy in this patient. An accurate preoperative and intraoperative evaluation of patients with Rokitansky syndrome is necessary to identify those who might benefit from this procedure.

References


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