CASE REPORT

Laparoscopic extirpation of an aplastic ectopic uterus in a patient with Mayer–Rokitansky–Küster–Hauser syndrome

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The Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome comprises the combined hypoplasia of the vagina and the uterus. In recent years, variable anomalies of the development of the Müllerian duct [as classified by the American Fertility Society (AFS)] have been operated on by the laparoscopic approach. We describe here a laparoscopic extirpation of an aplastic ectopic uterus in a patient found to have a hypoplastic vagina and ectopic uterus attached to the right pelvic wall. The uterus was linked to a normotopic right Fallopian tube and right ovary which were covered with endometriosis and adhesions. While the uterine vessels and the round ligament could be demonstrated on the right side, these structures were missing on the contralateral left side of the uterus. However, a normal tube and ovary were present on the left pelvic wall. Although not explicitly described in the AFS classification, this constellation most likely corresponds to the AFS classification stage Ie.

Key words: ectopic uterus/Mayer–Rokitansky–Küster–Hauser syndrome/operative laparoscopy

Introduction

In 1829, Mayer described partial and complete duplications of the vagina associated with cardiac, oropharyngeal and renal defects. Rokitansky (1838) published 19 autopsies on adult females with utero-vaginal agenesis. Following these preliminary descriptions, Küster (1910) reported several cases with genital abnormalities found to be associated with skeletal and renal anomalies. In addition, Hauser and Schreiner (1961) emphasized the need to distinguish the features of this malformation from those of testicular feminization. All these observations were grouped as a clinical entity called Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome. Tarry et al. (1986) defined the MRKH syndrome as a spectrum of anomalies of the Müllerian duct including vaginal aplasia which occur in genotypically and phenotypically normal females. In 1988, the American Fertility Society (AFS) published a standardized classification of these variable anomalies of the Müllerian duct.

So far, laparoscopic removal of the remnants of the Müllerian duct in patients with MRKH syndrome suffering from intractable lower abdominal pain has only been described three times. Canis et al. (1990) were the first to report the laparoscopic extirpation of a unicornuate uterus in a patient with MRKH syndrome stage IIb according to AFS classification. Following this description, Yeko et al. (1992) and Chapron et al. (1995) described laparoscopic surgery in patients with stage Ie syndrome also suffering from lower abdominal pain. Here we report laparoscopic surgery in a patient with an unpublished variant of MRKH syndrome stage Ie.

Case report

At the age of 19 years, the patient first presented in our outpatient clinic with lower abdominal pain and primary amenorrhoea. Her past medical and surgical history was uneventful. She complained about cyclically recurrent right lower abdominal pain which always lasted for 4 days. On examination, normally developed external genitalia were found, and an apparently normal vulva, labia and clitoris. However, the total length of a hypoplastic vagina was found to be ~1 cm. On rectal examination, no uterus could be palpated. The serum endocrine profile was uneventful. On ultrasound examination, a hypoplastic uterus with normal ovaries was suspected to be in an ectopic position. Physical and radiological examinations did not reveal any skeletal anomalies. To further evaluate the abdominal symptoms of this patient, a diagnostic laparoscopy was performed, during which the ectopic position of a hypoplastic uterus was confirmed. The uterus was found to be attached to the right pelvic wall. On this occasion, the patient did not wish to undergo further diagnosis and treatment.

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At the age of 26 years, the patient presented again in our outpatient clinic with persistent lower abdominal pain. After several years of vaginal sexual intercourse, the vaginal length was now 6 cm. The patient’s endocrine findings were uneventful and confirmed a normogonadotrophic eugonadal state. On pelvic ultrasound, a uterus of 4 cm with an endometrial lining of 7 mm was described which was found adherent to the right pelvic wall. The ovaries were determined to be normal. On computed tomography scan, a paracolic position of the uterus was confirmed; in addition, small cysts in the right ovary and multiple bilateral renal cysts were seen. On i.v. pyelography, spongiform renal degeneration with normotopic ureters was suspected.

After extensive counselling, the patient decided to undergo laparoscopic extirpation of the ectopic uterus. During surgery, a uterine remnant of ~4 cm in diameter was detected on the
right pelvic wall, close to the iliac vessels and median of the uterus. The round ligament together with a normal Fallopian tube with fimbriae was found on the right side of the uterus. Extense peritubal and tubo-ovarian adhesions existed, and thus the right ovary was primarily not visible. After adhesiolysis, the ovary was found to be within the normal size range, but with surface bleeding and blue–darkish discoulouration consistent with endometriosis of the ovary. The right ovary was firmly attached to the pelvic wall; the left Fallopian tube was torn longitudinally with the fimbriae located over the terminal pelvic line. Near the tube, a left ovary of 4 cm in size and of normal appearance was detected. The left tube merged into a fibrous string which inserted on the uterine remnant. For laparoscopic extirpation of the uterus, three lower abdominal insertions of 5 mm were chosen. First, the lateral peritoneum was dissected down to the crossing of the uterine vessels over the ureter, thus to see completely the course of the ureter. This was followed by a meticulous adhesiolysis. As the right ovary presented with severe adhesions to the hyoplastic uterus and was morphologically altered by multiple endometriotic cysts, we decided to remove the right ovary together with the Fallopian tube. To do this, the infundibular ligament was dissected, and the peritoneum between the uterus and pelvic wall was opened. The uterine vessels were tied laterally to the ureter and both the uterus and the right adnexa were removed after dissection of the vesical peritoneum. All visible endometriotic lesions were treated by use of the argon beamer. The duration of this surgery was 2.5 h.

The patient’s post-operative course was uneventful, and she was discharged on the third post-operative day. The patient presented again without any abdominal complaints 3 months after surgery. Histological examination revealed a uterus of 3.5×2 cm in size, with a proliferative endometrium of 2 mm and a myometrium of 1.2 cm. Extensive microfocal endometriotic lesions were found on the serosa and subserosa of the uterus, the tube and the peritoneum adjacent to the right ovary.

Discussion
The MRKH syndrome comprises combined hypoplasia of the vagina and the uterus. In general, patients present clinically with primary amenorrhoa and with cyclically recurrent lower abdominal pain. For relief of the latter symptoms, removal of the remnants of the Müllerian ducts has been suggested, particularly when endometrial proliferation with subsequent formation of a haematometra is suspected. To accomplish this, three laparoscopic procedures have so far been described. Canis et al. (1990) described the laparoscopic removal of Müllerian remnants in a patient with AFS stage IIb. In this patient, a right-sided unicornuate uterus with normal ovary and tube was found, while the left-sided uterine rudiment presented with an enlarged Fallopian tube and an endometriotic ovarian cyst of 15 cm in diameter. Yeko et al. (1992) reported a laparoscopic treatment of a specific maldifferentiation composed of bilaterally normal tubes and ovaries, in which each tube ended in a Müllerian duct remnant of 3×3 cm. The relics of the Müllerian duct consisted exclusively of myometrium, and neither endometrial components nor endometriosis could be found in this specimen. Both uterine horns were symmetrically developed and were mediadly linked by a fibrous tissue. Uterine vessels could not be detected. This finding corresponded to the AFS stage Ie. Chapron et al. (1995) described laparoscopic surgery of Müllerian anomalies in a patient with AFS stage Ie. They reported a case of bilateral uterine aplasia with two uterine horns. The left horn was 2 cm, the right 5 cm in diameter; both were found attached to normal tubes and ovaries; there were minimal endometriotic lesions next to the ovaries. With the corresponding vaginal aplasia, this stage of the MRKH syndrome corresponded to the case reported by Yeko et al. (1992), except for Chapron et al.’s finding of endometrial tissue in the aplastic uterus.

In the patient whom we have described, laparoscopic surgery revealed a hypoplastic vagina and an ectopic uterus attached to the right pelvic wall. This uterus was linked to a right Fallopian tube and right ovary which were covered with endometriosis and adhesions. The right uterine vessels and the round ligament could be demonstrated. These structures were missing on the left side of the uterus. However, we found a normal tube and ovary on the left pelvic wall. This constellation most likely corresponds to the AFS classification stage Ie, although the ectopic localization of an aplastic uterus is not described in the AFS stages. A hypoplastic uterus was found on the top of the right uterine distal to the right uterine artery and vein in immediate proximity to the vesical peritoneum. In combination with a strongly adherent right adnexa, these findings forced us to remove all these structures. The four cases and their laparoscopic treatment which have been published so far emphasize the variety of anomalies of the Müllerian duct and underscore the need for laparoscopic techniques adapted for each individual.

References

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Laparoscopy and MRKH syndrome