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Case Report

Rare case: enucleation of multiple fibroids after intraoperative vaginal sonographic detection of the solitary pelvic kidney by patient with Mayer-Rokitansky-Kuster-Hauser syndrome

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ABSTRACT

Mayer-Rokitansky-Kuster-Hauser syndrome (MRKH) is a congenital anomaly of the genital tract that occurs in about 1 in 4000 women. MRKH syndrome can be associated with renal, skeletal, heart and hearing abnormalities. The frequency of renal/urinary tract abnormalities is 33%. Only a few cases of fibroid development in MRKH syndrome have been described in the literature. The diagnosis and surgery of a fibroid in MRKH syndrome may be complicated in associated kidney abnormality by an atypical kidney position, as in this case: pelvic kidney on one side and renal agenesia on the contralateral side. Authors present the case of a 47-year-old female patient with a known MRKH syndrome and a pelvic kidney on the right side who had presented with an unclear tumour in the right lower abdomen. A completed CT scan revealed the tumour directly next to the pelvic kidney. A malignancy could not be excluded with certainty, so that a laparoscopy in laparotomy readiness was indicated and performed. During surgery, two rudimentary uterine horns were found; on the right side retroperitoneally, below the uterine horn, the tumour was located and directly below it there was a soft tissue alteration, probably the kidney. For safety, a vaginal sonography was performed in between, to clearly identify the only kidney and to avoid damaging it. The tumour could be extirpated laparoscopically without kidney injury. The two uterine horns were removed simultaneously. Histologically the fibroid could be confirmed. In addition, three other fibroids (one on the left side and two on the right side) were detected. Due to the high probability of a simultaneous kidney abnormality in the MRKH syndrome, authors suggest an accurate kidney diagnosis preoperatively. If necessary, in the case of a pelvic kidney and/or renal agenesia, as in this case, an additional intraoperative kidney check should be performed.

Keywords: Mayer-Rokitansky-Kuster-Hauser syndrome, Multiple myomas, Myoma, Solitary kidney, Uterus malformation

INTRODUCTION

Mayer-Rokitansky-Kuster-Hauser syndrome (MRKH) is a congenital anomaly of the genital tract that occurs in about 1 in 4000 women. It is a vaginal atresia often with rudimentary uterine horns or uterine aplasia with normal development of secondary sexual characteristics.[6,7] Since the external genitals look unremarkable, the MRKH is detected late.[6] At 20%, MRKH syndrome is the most common cause of primary amenorrhe.[6] MRKH syndrome can be associated with renal, skeletal, cardiac and hearing abnormalities.[5] Although it is very rare, a fibroid can develop from a rudimentary uterine horn.[5] Only a few cases of fibroid development in MRKH syndrome have been described in the literature.[3]

The diagnosis and surgery of a fibroid in MRKH syndrome may be complicated by an atypical renal position, such as in this case: pelvic kidney, in the presence of an associated renal abnormality.

CASE REPORT

In a 47-year-old female patient the MRKH syndrome was already known. This diagnosis was confirmed by laparoscopy 27 years ago. The second laparoscopy was performed 16 years ago with the creation of a neovagina using the meshgraft technique. A renal anomaly in the sense of a pelvic kidney on the right side was known. Now the patient was admitted to this case department with an unclear tumour in the right lower abdomen. After this finding could be confirmed sonographically and with the known pelvic kidney, a CT was indicated. The following findings were described in the CT: the left kidney was not attached, the right kidney was located in the small pelvis, cranially and ventrally of the right kidney there were unclear findings of about 4.5 cm in size. A malignancy could not be excluded with certainty, so that a laparoscopy in laparotomy readiness was indicated and carried out.

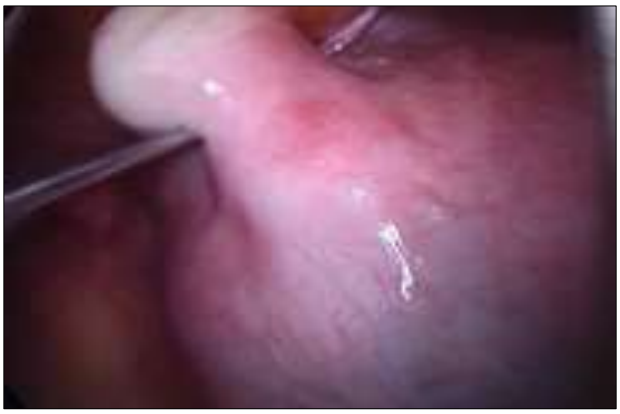

Figure 1: The right rudimentary horn with the fibroid below it.

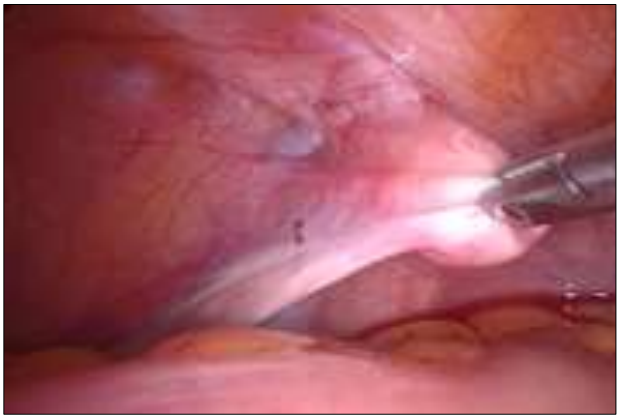

Figure 2: The left rudimentary horn.

During the operation, the following situation occurred

A uterine system was missing, the typical findings of a fold in the region of the vagina, in the area of the left pelvic wall outside the linea terminalis a streak gonad, above it a small rudimentary horn with connection to the inguinal canal, on the right side also a rudimentary horn, below it a tumour located retroperitoneally.

Figure 3: Opening of the peritoneum above the fibroid.

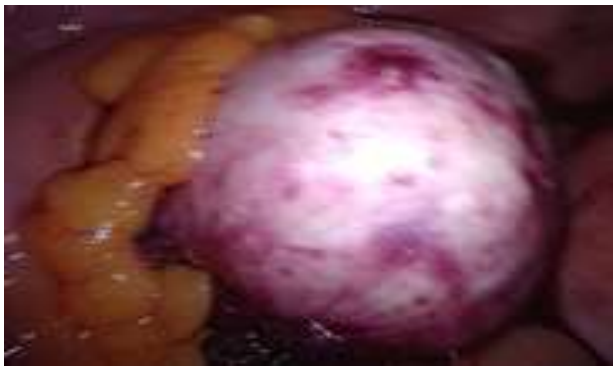

Figure 4: The dissected fibroid.

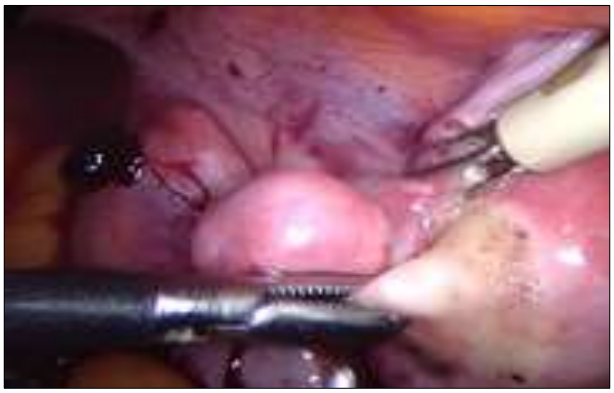

Figure 5: Removal of the right rudimentary uterine horn.

Underneath the tumour there was an obvious soft tissue alteration, which could correspond to the pelvic kidney. The peritoneum was opened above the tumour.

This finding was very solid. Since it was unclear whether it was the pelvic kidney or an additional tumour, the kidney was visualized by vaginal ultrasound and under vaginal sonographic view, which excluded that the prepared tumour was the only pelvic kidney, the tumour was removed.

It was a fibroid that was isolated from the pelvic wall and could be easily removed.

Meanwhile, the kidney was sonographically checked. The kidney is unchanged postoperatively after removal of the fibroid. An i.v. blue solution was also injected. The catheter urine showed a clear blue excretion, so that the excretory function of the right pelvic kidney was given.

Due to the fibroid that had developed from the rudimentary uterine horn, the two small uterine horns were removed simultaneously, and another small subserous fibroid of 1.2 cm was visible on the right horn.

The postoperative course was unremarkable, so that the patient could be discharged on the third postoperative day.

Histologically the fibroid (68 g) could be confirmed. On the right uterine horn there was another small ligamentary fibroid of 0.8 cm and on the left uterine horn a small regressive fibroid of 0.6 cm.

DISCUSSION

The MRKH syndrome is a rare disorder described as aplasia or hypoplasia of the uterus and the upper two-thirds of the vagina due to the developmental disorder of the Müllerian ducts. Women with this syndrome have a normal 46XX karyotype, secondary sexual characteristics, ovarian function and an underdeveloped vagina.[4] MRKH syndrome may be associated with renal, skeletal, cardiac and auditory abnormalities.[5] The incidence of renal / urinary tract abnormalities is 33%.[6]

The occurrence of a fibroid in MRKH syndrome is very rare.[5] Individual cases have already been described in the literature. Edmonds and Malik have mentioned 15 and 6 cases of MRKH syndrome and fibroids respectively in their studies after analyzing the literature.[1,3] The fibroids can develop simultaneously from both uterine horns.[2] Valecha has described a case of multiple fibroids in MRKH syndrome.[7]

In MRKH syndrome, the fibroids can reach different sizes. Rawat and Michiel have described cases with large fibroids in MRKH syndrome, corresponding to 30 cm and 14 cm.[5,8]

However, a case of multiple fibroids in MRKH syndrome and associated renal anomaly in the sense of a pelvic kidney on one side and renal agenesia on the contralateral side, as in this case, authors could not find in the literature. The specificity of the case is due to the location of the fibroids, immediately next to the single kidney, which makes surgery difficult. In order not to injure the only kidney, the operation was performed under simultaneous intraoperative renal control.

CONCLUSION

Due to the high probability of a simultaneous renal anomaly in the MRKH syndrome, authors recommend an accurate kidney diagnosis preoperatively. If necessary, in the case of a pelvic kidney and/or renal agenesia, as in this case, an additional intraoperative renal check should be performed.

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