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Case Report

Torsion of a rudimentary uterine horn with multiple leiomyomas in a case of Mayer-Rokitansky-Küster-Hauser syndrome

M.S. Kim[1], E.D. Na[1], H.C. Kim[1], M. Kim[1], S.Y. Shin[1], M.H. Lee[1,*]

[1]*Department of Obstetrics and Gynecology, CHA Bundang Medical Center, CHA University, Seongnam (Republic of Korea)*

Summary

The Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is characterized by congenital hypoplasia or aplasia of the uterus and vaginal agenesis. A 53-year-old woman presented to the emergency department with severe abdominal pain. She was diagnosed with torsion of the right rudimentary uterine horn with multiple leiomyomas with MRKH syndrome. The twisted rudimentary uterine horn and multiple leiomyomas were successfully removed via laparoscopy. MRKH syndrome with uterine horn torsion is uncommon and ours is the first case of torsion in MRKH syndrome treated via a laparoscopic approach. Our report indicates that laparoscopy is ideal for the simultaneous diagnosis and treatment of pelvic masses in MRKH syndrome.

Key words: Laparoscopy; Leiomyoma; Mayer-Rokitansky-Küster-Hauser syndrome; Torsion.

Introduction

MRKH syndrome is characterized by congenital hypoplasia or aplasia of the uterus and agenesis of the vagina. The incidence of MRKH syndrome has been estimated to be 1 per 4,500-5,000 females [1]. MRKH syndrome is usually identified at the time of evaluation for primary amenorrhea, infertility, and abnormalities of the internal genitalia. These patients are characterized by a normal female karyotype, secondary sexual characteristics, and ovarian function [2]. As ovarian function is normal in most patients with MRKH syndrome, the development of estrogen-dependent pathologies, such as leiomyomas and adenomyosis, is possible in the rudimentary uterine horn [3, 4] or in other pelvic areas [5]. Here, we report a case with severe abdominal pain that was diagnosed as torsion of a right rudimentary uterine horn with multiple leiomyomas in a patient with MRKH syndrome.

Case Presentation

A 53-year-old nulligravida woman presented to the emergency department of our university-affiliated hospital with severe abdominal pain. She was referred to the department of obstetrics and gynecology and diagnosed with multiple leiomyomas via abdominopelvic computed tomography (CT), which could not be differentiated from an ovarian fibrothecoma. The uterine structure was not defined on CT scan. She had undergone diagnostic laparoscopy for primary amenorrhea at the age of 20 years in another university hospital and was diagnosed with infertility, without any further investigation or subsequent treatment. The patient reported having a normal sexual life after marriage. On pelvic examination, she had normal external genitalia and a blind vaginal pouch measuring 6 cm in length, with the cervix absent. Ultrasonography and magnetic resonance imaging (MRI) revealed multiple solid masses in the pelvis, suspected as torsion of intraligamentary leiomyomas with an indistinct uterus (Figure 1). Being diagnosed with MRKH syndrome with uncertain pelvic masses causing significant abdominal pain, she was scheduled for laparoscopy. On laparoscopy, the abdominal cavity showed multiple leiomyomas ($7 \times 6 \times 6$ cm, $5 \times 5 \times 4$ cm and $2 \times 2 \times 2$ cm) protruding from the right rudimentary uterine horn (Figure 2A, 2B). The base of the rudimentary horn was rotated around 720 degrees clockwise (Figure 2B). The largest leiomyoma was entirely calcified with a stony-hard consistency. In the position of the uterus, only a thin transverse cord-like structure, 5-6 mm in diameter, was noted instead of a normal uterus, suggesting MRKH syndrome. A left small rudimentary horn, around 15 mm in size, was noted in the left pelvic position (Figure 2C). The fallopian tubes were distally attached to the bilateral rudimentary horns. The right ovary was slightly enlarged, with a 3×2 cm cystic mass and the left ovary looked normal. All the leiomyomas in the right rudimentary uterine horn and right adnexa were removed laparoscopically; the procedure was uneventful (Figure 2D). Pathological examination revealed a leiomyoma and endosalpingiosis in the right ovary. At the 1-month postoperative follow-up, the patient had recovered well.

Discussion and Conclusion

We reported a case with severe abdominal pain that was diagnosed as torsion of a right rudimentary uterine horn with multiple leiomyomas in a patient with MRKH syndrome. The etiology of MRKH syndrome is poorly understood, and it may be associated with significant urologic, skeletal, cardiac, and auditory abnormalities [2, 6]. MRKH

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Table 1. — *Case reports on Mayer-Rokitansky-Küster-Hauser syndrome with torsion of the rudimentary horn.*

Study	Year of publication	Study location	Age	Diagnostic methods	Treatment
Yan *et al*. [10]	2002	China	52	Ultrasound CT† Laparotomy	Hysterectomy and bilateral salpingo-oophorectomy
Fletcher *et al*. [8]	2012	India	28	MRI‡ Laparotomy	Stalk of the leiomyoma was suture-ligated
Kundo *et al*. [9]	2014	US	40	Ultrasound CT Laparotomy	Right salpingo-oophorectomy, excision of the right and left hemi-uteri, and left salpingectomy
Present	2020	Korea	53	Ultrasound CT MRI Laparoscopy	Right salpingo-oophorectomy, excision of the right rudimentary uterine horn

† CT, computed tomography; ‡ MRI, magnetic resonance imaging.

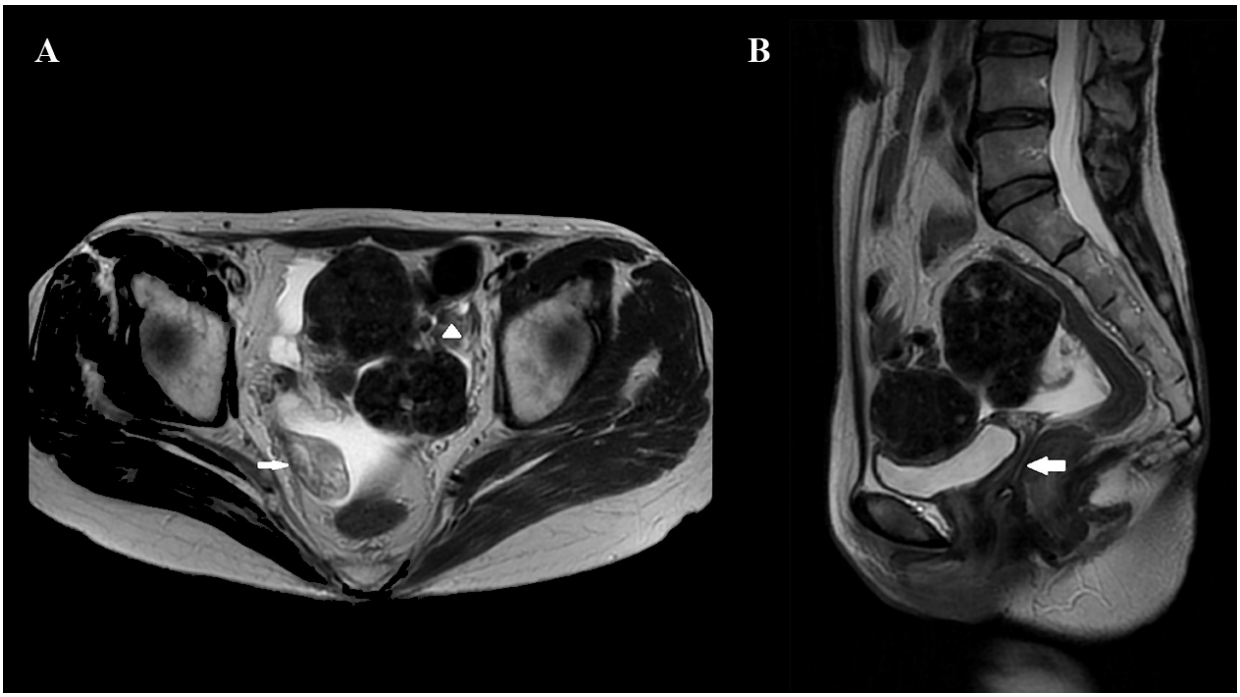

Figure 1. — Magnetic resonance imaging (MRI). (A) Axial T2-weighted images showing a normal right ovary (white arrow). The right rudimentary horn was twisted (white arrowhead), suggesting uterine torsion. (B) Sagittal T2-weighted MRI showing two heterogeneously enhanced circumscribed masses in the pelvis. The absence of the uterus and the upper two-third segment of the vagina was noted (white arrow). MRI, magnetic resonance imaging.

syndrome is subdivided into 2 types [6]. Type A occurs in an estimated 44% of MRKH patients, with isolated Müllerian duct malformations presenting as shallow vaginal dimples with the cervix, uterus, and upper vagina being absent, and it is not associated with other anomalies. Type B occurs in an estimated 56% of MRKH patients, who have a similar Müllerian agenesis as in type A, along with varying degrees of associated congenital renal malformations (renal agenesis, renal ectopia, and horseshoe kidney), ovarian anomalies (decreases ovarian volume, higher position of ovaries, hypoplasia), and skeletal abnormalities (scoliosis, spina bifida, and sacral lumenization), and unilateral auditory defects [6, 7]. In our case, bilateral kidneys with a normal shape and position were identified on the abdominopelvic CT. In addition, no lumbosacral bony abnormalities were defined on CT or MRI.

As the ovarian function is normal, the development of estrogen-dependent pathologies is possible in various areas of the pelvis in patients with MRKH syndrome [3-5]. A rudimentary uterine horn is a typical site where estrogen-dependent smooth muscle cells are found and is a most likely site for the occurrence of leiomyomas or ade-

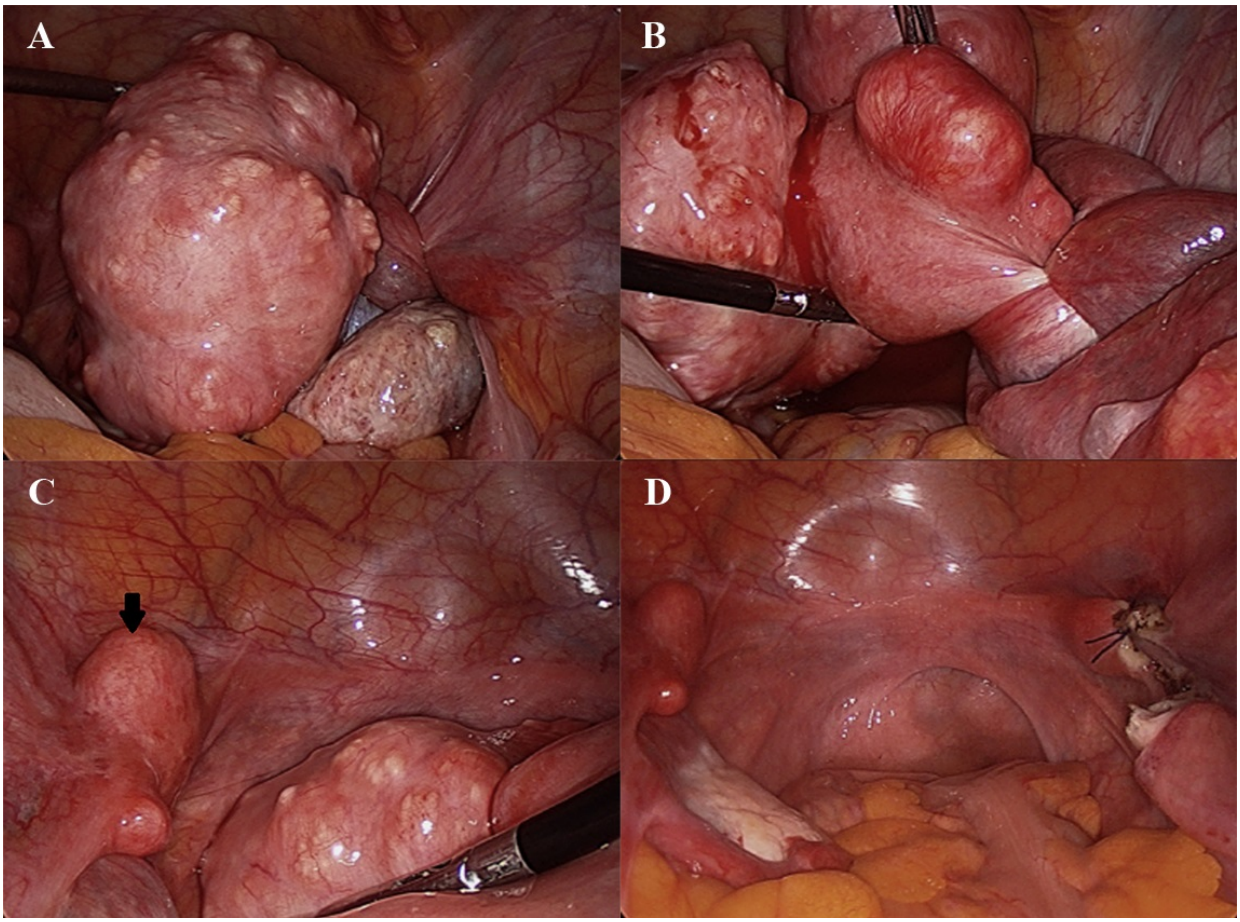

Figure 2. — Laparoscopic findings. (A) Multiple leiomyomas protruding from the right rudimentary uterine horn. (B) The base of the right rudimentary horn was rotated around 720 degrees clockwise. (C) A left small rudimentary horn may be seen (arrow). (D) All the leiomyomas and the right rudimentary uterine horn and right adnexa were removed.

nomyosis in patients with MRKH syndrome. Few cases of leiomyomas arising from the rudimentary uterine horn have been reported in the literature [3, 4, 8-10]. Secondary to its mass effect, the risk of torsion in large pedunculated leiomyomas arising from a small uterine remnant is increased [9]. To date, 3 case reports have described the torsion of the rudimentary uterine horn with protruding leiomyomas [8-10]. Of note, in all these 3 cases, the pelvic mass was removed by laparotomy (Table 1).

Our case suggests that leiomyoma-associated torsion should be suspected when a pelvic mass accompanied by acute lower abdominal pain is observed in patients with MRKH syndrome. The differential diagnoses of pelvic masses in MRKH syndrome include masses arising from the ovaries, gastrointestinal stromal tumor of the intestine, extravesical leiomyoma of the bladder, and leiomyomas arising from the rudimentary uterine horn [3]. The initial medical diagnosis should be based on patient history and gynecological examination. The three most common methods of diagnosing MRKH syndrome are MRI, ultrasound, and laparoscopy [1, 6]. However, in MRKH syndrome with a pelvic mass, imaging tools have limitations with respect to diagnosing the origin of the mass because of the indistinct uterus and abnormal anatomy [11]. In a review of 6 cases of MRKH syndrome with ovarian tumors, laparoscopy was shown to offer the possibility of concurrent diagnosis and treatment [11].

To our knowledge, this is the fourth case of torsion of a rudimentary uterine horn reported in a patient with MRKH syndrome and the first case of torsion to be treated with laparoscopy. Our report also reveals the importance of imaging tools and laparoscopy as a diagnostic and therapeutic method in MRKH syndrome with a pelvic mass.

Abbreviations

MRKH, Mayer-Rokitansky-Küster-Hauser; CT, computed tomography; MRI, magnetic resonance imaging.

Authors' Contributions

All authors have read and approved the manuscript. Conceptualization: MH Lee. Data curation: M Kim, SY Shin. Investigation: HC Kim. Writing - original draft preparation: MS Kim. Writing - review and editing: ED Na.

Ethics Approval and Consent to Participate

This study was approved by the Research Ethics Committee of the CHA Bundang Medical Center (Ethical approval number: 2019-03-045-001). Written informed consent was waived by the Institutional review board approval from the CHA Bundang Medical Center.

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Conflict of Interest

The authors declare no conflict of interest.

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References

[1] "ACOG Committee Opinion No. 728: Mullerian agenesis: diagnosis, management, and treatment". *Obstet. Gynecol.*, 2018, *131*, e35-e42.
[2] Oppelt P., Renner S.P., Kellermann A., Brucker S., Hauser G.A., Ludwig K.S., *et al.*: "Clinical aspects of Mayer-Rokitansky-Kuester-Hauser syndrome: recommendations for clinical diagnosis and staging". *Hum. Reprod.*, 2006, *21*, 792-797.
[3] Rawat K.S., Buxi T., Yadav A., Ghuman S.S., Dhawan S.: "Large leiomyoma in a woman with Mayer-Rokitansky-Kuster-Hauser syndrome". *J. Radiol. Case Rep.*, 2013, *7*, 39-46.
[4] Girma W., Woldeyes W.: "Leiomyoma arising from Mullerian remnant, mimicking ovarian tumor in a woman with MRKH syndrome and unilateral renal agenesis". *Ethiop. J. Health Sci.*, 2015, *25*, 381-384.
[5] Hoo P.S., Norhaslinda A.R., Reza J.N.: "Rare case of leiomyoma and adenomyosis in Mayer-Rokitansky-Kuster-Hauser syndrome". *Case Rep. Obstet. Gynecol.*, 2016, *2016*, 3725043.
[6] Bombard D.S., 2nd, Mousa S.A.: "Mayer-Rokitansky-Kuster-Hauser syndrome: complications, diagnosis and possible treatment options: a review". *Gynecol. Endocrinol.*, 2014, *30*, 618-623.
[7] Strubbe E.H., Willemsen W.N., Lemmens J.A., Thijn C.J., Rolland R.: "Mayer-Rokitansky-Kuster-Hauser syndrome: distinction between two forms based on excretory urographic, sonographic, and laparoscopic findings". *AJR Am. J. Roentgenol.*, 1993, *160*, 331-334.
[8] Fletcher H.M., Campbell-Simpson K., Walcott D., Harriott J.: "Mullerian remnant leiomyomas in women with Mayer-Rokitansky-Kuster-Hauser syndrome". *Obstet. Gynecol.*, 2012, *119*, 483-485.
[9] Kundu K., Cohen A.W., Goldberg J.: "Acute torsion of uterine remnant leiomyoma with Mayer-Rokitansky-Kuster-Hauser syndrome". *Fertil. Steril.*, 2014, *102*, 607-609.
[10] Yan C.M., Mok K.M.: "Uterine fibroids and adenomyosis in a woman with Rokitansky-Kuster-Hauser syndrome". *J. Obstet. Gynaecol.*, 2002, *22*, 561-562.
[11] Miao Y., Wen J., Huang L., Wu J., Zhao Z.: "Diagnosis and management of ovarian tumor in Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome". *Biomed. Res. Int.*, 2018, *2018*, 2369430.

Corresponding Author:
MEE-HWA LEE, M.D., Ph.D.
Department of Obstetrics and Gynecology, CHA Bundang Medical Center, CHA University,
59, Yatap-ro, Bundang-gu, Seongnam-si, Gyeonggi-do, 13496 (Republic of Korea)
e-mail: yeegen@cha.ac.kr