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Case Report

A case of large leiomyoma arising from rudimentary horn in Mayer-Rokitansky-Küster-Hauser syndrome, managed by minimally invasive surgery

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ABSTRACT

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is a congenital disorder marked by aplasia or hypoplasia of the uterus and vagina as a result of arrest in the development of the müllerian ducts. Prevalence being 1 in 4000–5000 live births of females. Leiomyoma is the most common uterine tumor, their occurrence from rudimentary uterus in females with MRKH syndrome is very rare and only a few cases have been documented in the literature. Here we report a 38-year-old female, known case of MRKH syndrome presenting with USG suggestive of 7.9x7.4x6.0 leiomyoma in proximity to the hypoplastic uterus, undergoing a laparoscopic removal of fibroid with right ovarian cystectomy (incidental finding). Therefore, it is important to consider such unusual diagnosis of fibroids originating from primitive horns while treating these patients for gynecological symptoms as well as infertility, and consider them managing endoscopically.

Keywords: Mayer-Rokitansky-Kuster-Hauser syndrome, Leiomyoma, Fibroid, Mullerian duct, Laparoscopy

INTRODUCTION

Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is a congenital condition that affects the reproductive organs in females.[1] The upper vagina, cervix, and uterus are underdeveloped or absent in this syndrome due to arrest in the development of the müllerian ducts, the external genitalia are normal, with normal 46 XX karyotype, normal female secondary sex characteristics, presenting with primary amenorrhea.[1,2]

According to the involvement of structures other than those connected to the reproductive organs, there are three different kinds of this syndrome. Type I is the most prevalent, and it is characterised by defects that only affect the reproductive system. The second type, type II, is uncommon and is characterised by the existence of symmetric uterine remnants and abnormal uterine tubes, which are typically linked to ovarian disease, congenital renal disorders, bone abnormalities, and hearing impairments. The third type is referred to as MURCS type, and it includes renal, skeletal, and cardiac abnormalities with uterovaginal hypoplasia or aplasia.[1] Because tissue patterning and organ morphogenesis in the human embryo are complex processes that depend on a combination of timely signalling from genetic factors, soluble morphogens, chemical factors, and mechanical forces, several potential aetiologies, including monogenic, oligogenic, polygenic, multifactorial, and environmental factors, should be taken into consideration at this point.[3]

Fibroids of the uterus are the most common benign pelvic tumors in women worldwide. Their diagnosis is usually not missed because of the widespread and well-established use of ultrasound in gynecological clinics. Hence, the development of an unusually large myoma in a patient with MRKH syndrome is a rare event.

CASE REPORT

38-year-old, previously diagnosed (laparoscopically) with MRKH syndrome presented with c/o chronic lower abdomen pain with abdominal with ultrasound report s/o left adnexal mass 7.9×7.4×6 cm with minimal vascularity in close proximity to the uterus s/o broad ligament fibroid/ pedunculated fibroid.

On examination

(1) P/A: Ill-defined firm mass felt in midline with restricted mobility (6*5*cm); (2) P/S: Blind vagina noted with a vaginal length ~3 cm; (3) P/R: 6 cm mass felt above the vault, anterior to the rectum.

Investigations

Patient investigated with routine blood tests and tumor markers found to be within normal limit. Radiological investigations done and noted as follows.

Ultrasound s/o: An anteverted small uterus with left adnexal mass of size 7.9×7.4×6 cm showing proximity to the uterus, with ovaries seen normally. Impression as broad ligament fibroid or pedunculated uterine fibroid.

MRI s/o: An enlarged, bulky left uterine cornua of 6.8×8.9×9.8 cm. Impression as diffuse adenomyosis of the left uterine myometrium.

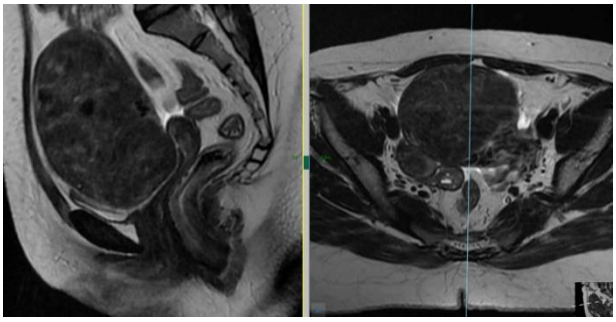

Figure 1: MRI images s/o diffuse adenomyosis.

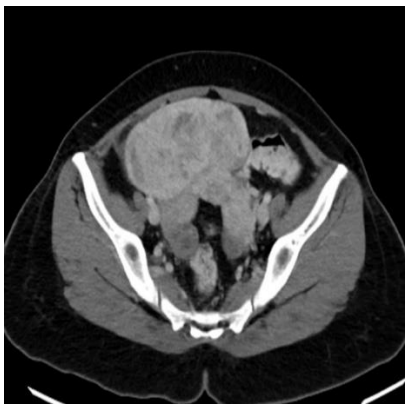

Figure 2: CT scan image s/o left uterine cornual fibroid.

CT scan s/o

CT scan s/o was 5.7×10.1 cm solid mass from the left uterine cornua likely fibroid. The impression was fibroid.

Treatment

Patient underwent laparoscopic myomectomy with right ovarian cystectomy.

Intraoperative findings are (1) left pelvic wall: left ovary, left fallopian tube, left rudimentary horn with attached large 9x8 cm left sub-serosal pedunculated fibroid, (2) midline fusion defect of mullerian system noted, (3) right pelvic wall: incidently right ovarian cyst 4×3 cm, right ovary, right fallopian tube attached to small hypoplastic uterus with blunt left end. Confirmed with histopathology report.

Following are the intraoperative images.

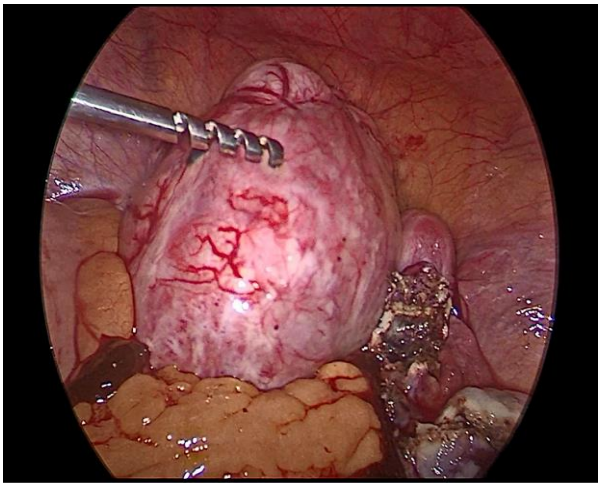

Figure 3: A large 8×8 cm left sub-serosal pedunculated fibroid note.

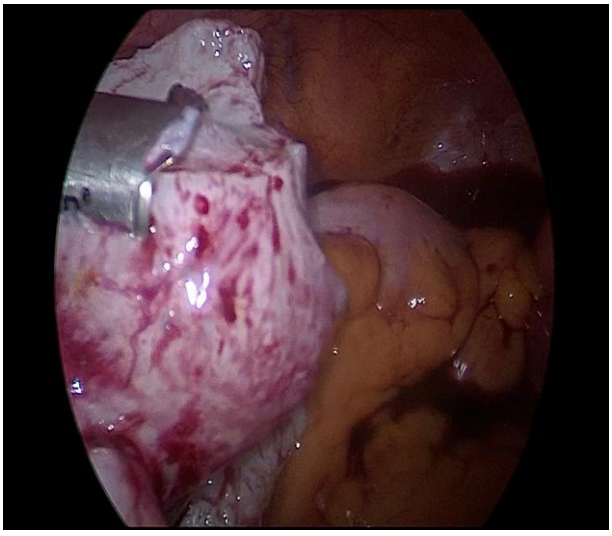

Figure 4: Laparoscopic myomectomy.

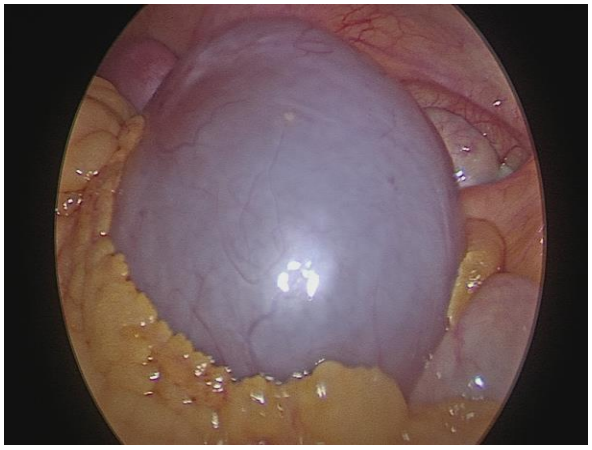

Figure 5: Right ovarian cyst.

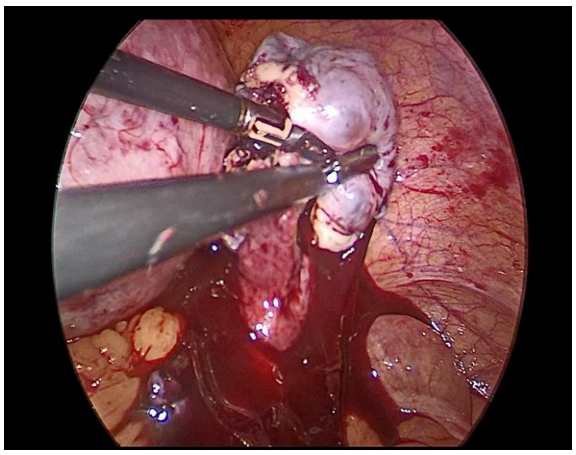

Figure 6: Right cystectomy.

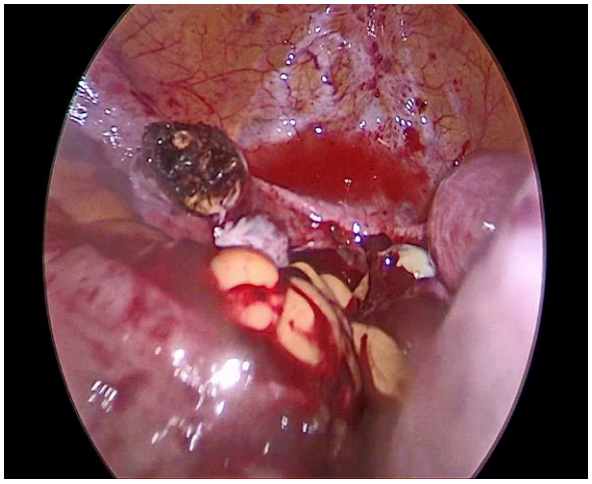

Figure 7: Post procedure.

DISCUSSION

Patients with MRKH syndrome have normally developed ovaries and fallopian tubes and many of them have two uterine remnants. Remnants contain fibromuscular tissue, a target tissue for ovarian steroids. And hence leiomyomas can originate.[4] Presence of solid pelvic mass is difficult to examine, especially when no vaginal reconstruction is done. Therefore, should be thoroughly evaluated. Hence it is important to have such rare diagnosis in mind of the fibroids arising from rudimentary horns while treating patients for gynecological complaints and surgeon should be skillful to remove such masses endoscopically.[4]

CONCLUSION

Although development of leiomyoma is rare in MRKH syndrome patients, it needs to be included in the differential diagnosis of pelvic mass in these patients.

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Ethical approval: Not required

REFERENCES

1. Harzif AK, Ambalagen S, Charilda FE, Mutia HD. A rare case of multiple leiomyomas on rudimentary uterus in a woman with Mayer Rokitansky Kuster Hauser (MRKH) syndrome: A challenging diagnosis and laparoscopic approach. Int J Surg Case Rep. 2021;81:105711..
2. Boruah DK, Sanyal S, Gogoi BB, Mahanta K, Prakash A, Augustine A, et al. Spectrum of MRI Appearance of Mayer-Rokitansky-Kuster-Hauser (MRKH) Syndrome in Primary Amenorrhea Patients. J Clin Diagn Res. 2017;11(7):TC30-TC35.
3. Herlin MK, Petersen MB, Brännström M. Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome: a comprehensive update. Orphanet J Rare Dis. 2020;15(1):214.
4. Gutsche MG, Chagas LA, Leal R, Cunha AL, Djahjah MC. Mayer Rokitansky Kuster Hauser syndrome: a case report and literature review. Radio Bras. 2011;44(3):192-4.

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