International Journal of Reproduction, Contraception, Obstetrics and Gynecology
Choudhary A et al. Int J Reprod Contracept Obstet Gynecol. 2023 Jun;12(6):1892-1895
www.ijrcog.org
pISSN 2320-1770 | eISSN 2320-1789

DOI: https://dx.doi.org/10.18203/2320-1770.ijrcog20231576

Case Report

A rare case of broad ligament fibroid in Mayer-Rokitansky-Küster-Hauser syndrome

Arpit Choudhary, Navneet Takkar, Dilpreet Kaur Pandher*, Mohit Satodiya

Department of Obstetrics and Gynaecology, Government Medical College and Hospital, Chandigarh, India

Received: 27 March 2023
Accepted: 01 May 2023

***Correspondence:**
Dr. Dilpreet Kaur Pandher,
E-mail: dr_dilpreet@yahoo.com

ABSTRACT

Mayer-Rokitansky Kuster Hauser (MRKH) syndrome with lieomyoma is a rare disorder. Women with this syndrome have normal 46 XX karyotype, normal secondary sex characteristics and primary amenorrhea. There is a scarcity of cases in the literature where fibroid develops in women suffering from MRKH syndrome. Here, we present a case of an ectopic broad ligament fibroid found in a 41-year-old woman with MRKH syndrome type II. A 41-year-old married nulliparous female diagnosed with MRKH syndrome 20 years back, presented with an abdominal mass. Trans-abdominal ultrasound suggested a large mass on right side arising from paracolic area with heterogeneous echo texture. However, the ovarian vs leiomyoma origin of mass was of diagnostic dilemma. CEMRI confirmed the same findings and ruled out ovarian origin of the mass along with the presence of rudimentary uterus. Following this, the patient was taken for laparotomy followed by total abdominal hysterectomy with bilateral salpingoopherectomy, the fibroid was found to be arising from broad ligament and was removed along with the hypoplastic uterus and bilateral Fallopian tubes and ovaries. The case we presented exhibits the development of leiomyomas in patients with MRKH syndrome, although rare, is always a possibility and should be kept in mind as a differential diagnosis while evaluating an abdominopelvic mass. MRI is an accurate modality both for delineating the mass, confirming its origin as well as diagnosing MRKH syndrome.

Keywords: Mayer-Rokitansky-Kuster-Hauser syndrome, MRKH, Leiomyoma, Mullerian duct anomalies

INTRODUCTION

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is named after the four physicians German anatomist August Franz Josef Karl Mayer (1829), Austrian anatomist Carl von Rokitansky (1838), German gynaecologist Hermann Küster (1910) and Swiss gynaecologist Georges Andre Hauser (1961) who over the period of 130 years thoroughly described the syndrome.[1] It is a congenital disorder of females affecting primarily the reproductive organs but may also involve other organ systems. It is associated with an absence or incomplete development of mullerian ducts. It is is characterized by congenital aplasia or hypoplasia of the uterus and the upper part (2/3) of the vagina in women showing normal development of secondary sexual characteristics and a normal 46, XX karyotype. It affects 1 in 4500 women globally.

Myomas are rather common benign lesions that usually arise from the myometrium of normal uterus. The occurrence of myoma in a patient of MRKH syndrome is a very rare finding and only few cases have been reported.

Here, we report a patient of MRKH syndrome with a large leiomyoma originating from the broad ligament with rudimentary uterus.

CASE REPORT

A 41-year-old, nulligravida female with MRKH syndrome diagnosed 20 years back presented to our institute with

complaint of heaviness in lower abdomen and increase in frequency of micturition for 5 months, awareness of mass per abdomen for 5 months, not associated with pain abdomen, loss of weight and appetite or alteration in bowel habits. She first went to her primary healthcare provider, where she was advised to undergo an ultrasound which was suggestive of a solid abdominopelvic mass approximately 17.3×16.5×11.5 cm in size of likely ovarian pathology. Thereafter she was referred to our institute for further management.

The patient gave the history that she never attained menarche for which she was evaluated at the age of 18 years and was diagnosed with hypoplastic uterus and blind ending vagina. Patient had also undergone vaginoplasty in 2004. Patient adopted a child 18 years back. She has been non coital since past 10 years. There was no similar history in family.

During examination, her vitals were stable. Patient had well developed secondary sexual characteristics. On per abdomen examination, an abdominopelvic mass could be palpated which was up to 26 weeks gravid uterus in size, firm in consistency, had regular margins with limited side to side mobility, it appeared to be bilobed, non-tender with no local rise of temperature. On local examination, pubic hair were present, vagina was atretic and vaginoplasty area appeared to be obliterated. On per rectal examination the same mass was felt which was free from rectal mucosa and occupying the whole of pelvis.

She had undergone a CECT before reporting to us (Figure 1) which was suggestive of large lobulated heterogeneously enhancing solid mass in pelvis 18.6×17.9×12.6 cm. A small pear-shaped mass was seen adjacent to the large mass likely rudimentary uterus and bilateral moderate hydronephrosis. However, the ovaries were unable to be visualized.

As the ovarian origin of the mass was of contest and the ovaries were unable to be visualized on CECT. Serum tumor marker levels were performed to rule out ovarian malignancy. It showed CA 125 of 24.8 U/ml, CEA-1.96 mg/ml, inhibin B-1 pg/ml, LDH-167 IU/L and AFP-1.3 ng/ml which were all within normal limits

A transabdominal ultrasound was done which was suggestive of a large mass more on right side likely in the right paracolic gutter/ broad ligament area with heterogeneous echotexture. Picture was suggestive of leiomyoma or leiomyosarcoma. A CEMRI (Figure 2) was ordered which as suggestive of a similar large abdominopelvic mass having well-defined fat planes with adjacent organs and posteriorly compressing the great vessels. Uterus was not separately visualized due to its likely hypoplastic nature. Bilateral ovaries were visualized separately and were normal. However, the nature of the mass was doubtful of malignancy. Patient underwent a 2D echocardiography as a part of her pre anesthesia evaluation which was suggestive of dextrocardia.

Since the ovarian origin of the tumor was not clear preoperatively from rudimentary/ broad ligament fibroid, preoperative bilateral DJ stenting was done for optimal identification of ureter during the primary procedure to prevent undue injury to the ureters. The patient was subsequently taken up for surgery. The abdomen was opened through midline incision. Intraoperatively, a large lieomyoma (Figure 3) approximately 12×15 cm was seen arising from the broad ligament separate from the uterus (true broad ligament leiomyoma), the uterus was hypoplastic. The broad ligament fibroid was removed followed by total abdominal hysterectomy and bilateral salpingo-opherectomy. Patient stood the procedure well. A planned DJ stent removal was done on post-operative day 16 and the patient was discharged on post-operative day 20 after an uneventful post-operative period. Patient was followed up in OPD after 2 weeks and was healthy with no post-operative complaints. Histopathology report was consistent with the intraoperative findings and suggestive of benign spindle cell tumor leiomyoma and hypoplastic uterus with normal adnexa.

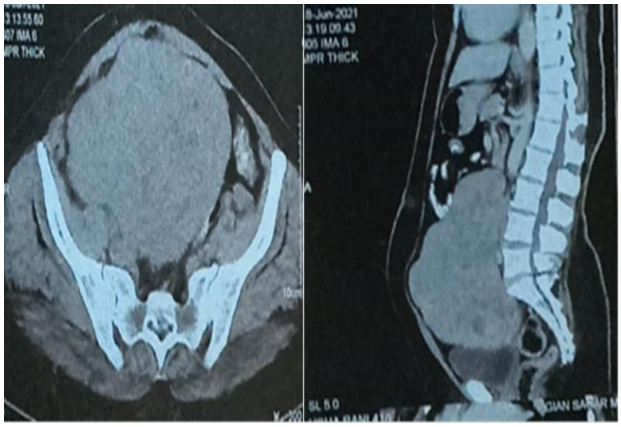

Figure 1: CECT pelvis of the large lobulated heterogeneously enhancing solid mass likely fibroid and a small hypoplastic uterus adjacent to the mass.

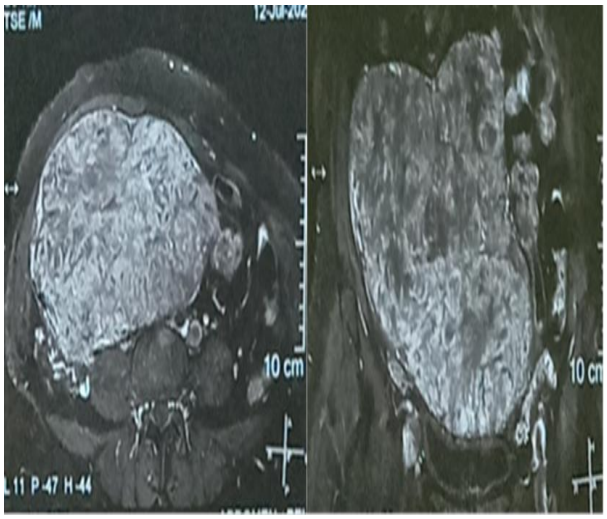

Figure 2: CEMRI pelvis of the same mass with separate normal looking ovaries.

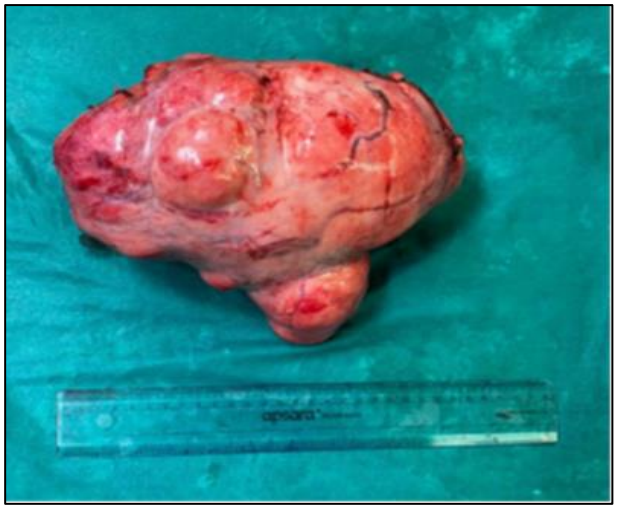

Figure 3: Gross morphology of the fibroid removed during the procedure.

DISCUSSION

MRKH syndrome is a rare congenital disorder which is usually diagnosed in adolescence when the patient is unable to achieve menarche. It occurs due to absence or incomplete development of mullerian ducts. It is characterized by aplasia or hypoplasia of uterus with a blind ending vagina. These patients have normal secondary sex characters like well-developed breast, pubic and axillary hair, normal height with normal labia majora and minora but absent uterus. They have female karyotype (46, XX) with normal functional ovaries and normal hormonal profile. The syndrome is classified into two types according to involvement of structures other than reproductive organs. Type I MRKH syndrome / isolated MRKH syndrome is more common and is represented by abnormalities limited to the reproductive organs. Type 2 MRKH syndrome also known as MURCS (mullerian duct aplasia, renal dysplasia and Cervical Somite anomalies) is characterized by multi organ involvement in the form of uterovaginal hypoplasia or aplasia, renal, bone, and cardiac malformations.[2] Our patient belonged to type II MRKH syndrome due to presence of cardiac involvement in the form of dextrocardia.

MRKH syndrome is one of the most common cause of primary amenhorea. It may either be sporadic, familial, or genetic in etiology. Transmission is autosomal dominant, and multiple patients with this genetic condition may have variable presentation. Loss of function mutation in the WNT4 gene has been linked to MRKH syndrome in the medical literature.[1] The association of MRKH syndrome with myoma was 1st reported in 1977 by Beecham and Skiendzielewski and only limited cases of such association have been reported since then.[3]

Leiomyomas are common benign, monoclonal tumors of smooth muscle cells of the myometrium containing varying amounts of fibrous tissue and grow in size because of estrogen stimulation. Classically fibroids originate from the uterus however, fibroid mass may rarely arise from adjacent structures such as cervix, broad ligaments and other mullerian remnants. From embryology studies it is known that Mullerian ducts though primarily endodermal in origin have smooth muscles cells at the proximal ends. Thus, fibroids can originate even in hypoplastic uterus and from mullerian remenents.[4]

Blontzos et al reviewed medical literature of 35 patients with MRKH and leiomyoma. The origin of leiomyoma in MRKH patients was the uterine remnant in 65% of patients. Other origins include fibrous myometrial bands, broad ligament, round ligament, and parametrial tissue.[5] Considering that patients with MRKH have ovaries with normal hormone levels, fibroids in these patients undergo the same patterns like in normal patients. In addition, it has been suggested that estrogen receptor sensitivity or concentration in the remnant tissue may be potentially responsible.[2]

In our patient the leiomyoma grew large enough to cause pressure symptoms of significant severity that it led to bilateral hydronephrosis. Initially the ovarian vs uterine/ broad ligament origin of the mass was a diagnostic dilemma, the distinction of which was important and CEMRI was helpful in ruling out the ovarian origin of the mass.

Due to the rapid nature of growth of the mass the risk of malignancy could not be ruled out. Imaging findings weighed towards benign nature of the mass however CEMRI findings indicated doubtful malignant nature of the mass. To differentiate between leiomyoma and lieomyosarcoma a true cut biopsy was performed.

Because of the large size of the mass which distorted the pelvic anatomy and the close relation between the mass and ureters in imaging, additional precautionary steps were taken preoperatively (i.e., Bilateral DJ stenting) for ease in intraoperative identification of the ureter which helped in preventing undue injury to the ureter.

Differential diagnosis of such presentation include ovarian fibroma, GIST (gastrointestinal stromal tumor), extra vesical leiomyoma of the urinary bladder. Ovarian malignancies in such patients should always be ruled out as patients with MRKH syndrome have normal functioning ovaries and are thus at-risk malignant transformation of ovaries like any other female. Furthermore, their inability to conceive increases the risk of ovarian malignancies due to incessant ovulation. Our patient was adequately explained this risk of ovarian malignancy after which the patient opted for prophylactic bilateral oophorectomy.

Similarly, leiomyomas should not be forgotten as a potential differential diagnosis while evaluating an abdominopelvic mass in a patient with MRKH syndrome just because of absence of uterus. Leiomyomas can arise from adjacent structures as well as uterine remnants in

such patients. MRI in such patients is an accurate diagnostic modality. Such fibroids if symptomatic should be thoroughly investigated and surgically removed.

CONCLUSION

The case we presented exhibits the development of leiomyomas in patients with MRKH syndrome, although rare, is always a possibility and should be kept in mind as a differential diagnosis while evaluating an abdominopelvic mass. MRI is an accurate modality both for delineating the mass, confirming its origin as well as diagnosing MRKH syndrome. Such masses should be thoroughly investigated and treated in a timely fashion. When dealing with large abdominopelvic masses pre-operative DJ stenting can be done to prevent undue injury to the ureters.

ACKNOWLEDGEMENTS

Author would like to thanks to whole nursing and medical team who contributed in one way or the other for the efficient management of the client.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Choudhary A, Takkar N, Pandher DK, Satodiya M. A rare case of broad ligament fibroid in Mayer-Rokitansky-Küster-Hauser syndrome. Int J Reprod Contracept Obstet Gynecol 2023;12:1892-5.