Torsion Adnexal Mass in a Patient with Mullerian Agenesis

Tharun Ganapathy Chitrambalam[1], Ramyasree Paladugu[2], Jeyakumar Sundaraj[3], Pradeep Joshua Christopher[4], Sundeep Selvamuthukumaran[5]

[1, 2, 3, 4, 5] Department of General Surgery, SRM Medical College and Research Institute, SRM University, Kattankulathur, Tamilnadu, India.

INTRODUCTION

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is characterised by underdevelopment of vagina and uterus. This may be due to early arrest in the development of the Mullerian duct causing aplasia of the vagina and uterus. The paramesonephric duct or mullerian duct develop to form the fallopian tubes, corpus and cervix of the uterus along with upper portion of the vagina. The mullerian ducts have smooth muscle cells existing at the proximal ends, from which the leiomyoma originates. Since the uterus is underdeveloped, these leiomyomas of larger size may have a mass effect increasing the risk of torsion due to large volume disparity. Here we report one such case of a torsion of a huge leiomyoma arising from the left adnexa in a patient with MRKH syndrome who presented as acute abdomen.

Mullerian agenesis also termed to be Mayer-Rokitansky-Küster-Hauser syndrome is a syndrome first studied by Mayer; later by Rokitansky, Küster and Hauser. This is described as a spectrum of anomalies such as congenital hypoplastic or absence of uterus, hypoplastic fallopian tubes and blind vagina in a female (46XX genotype) with normal secondary sexual characters and normal endocrine status. The incidence of MRKH syndrome is 1 in 4000 female births.[1] The ovaries develop normally as they are derived from the genital ridge and the hence the sex hormones are found to be normal. Associated anomalies include renal, skeletal, cervicothoracic and auditory defects in the most severe form.[2] The pathogenesis of formation of leiomyomas in MRKH syndrome is not yet known but it is found that these leiomyomas have oestrogen and progesterone receptors as seen in the normal uterine histopathological picture responding to the sex steroids mainly oestrogen released from the ovaries. This acts as a promotor for the increase in the size of leiomyomas in the rudimentary uterine tissues. The risk of torsion in a large pedunculated leiomyoma arising from a small uterine remnant is increased secondary to its mass effect.[3]

PRESENTATION OF CASE

A 43-year-old unmarried female, presented to the emergency room with acute onset of severe abdominal pain, not relieved by medications for 2 days. Patient did not attain menarche. She is a known case of Mullerian agenesis. No history of urinary complaints. No past history of use of any contraceptives or vaginal dilators. On physical examination; pallor was noted and vitals were found to be within normal limits. Abdomen was tense with diffuse tenderness and guarding over the lower abdomen. A 10 x 8 cm vague tender mass was palpable in the hypogastrium, partly mobile and bowel sounds were present. Breasts and pubic hair are well developed to Tanner's stage V. Labia majora and minora was underdeveloped for age. Vaginal introitus was not seen. Rectum was found be loaded with stools.

Corresponding Author:
Jeyakumar Sundaraj.
Department of General Surgery,
SRM Medical College and Research Institute, SRM University, Kattankulathur, Tamilnadu, India.
E-mail: drtarunchitrambalam@gmail.com

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Investigations

On imaging; Ultrasonography and Computerized Tomography (CT) of abdomen suggested a well-defined soft tissue density lesion measuring 13 x 9.6 x 11 cm noted in pelvis anterior to the bladder, occupying the pelvis and left lower quadrant of abdomen with minimal ascites. No evidence of dilated bowel loops or pneumoperitoneum.

Treatment

Diagnostic laparoscopy revealed a 14 x 10 cm left adnexal mass involving left ovary which was double twisted around its pedicle causing complete gangrene (Figure 1). Uterus was found to be underdeveloped. Right ovary with fimbriae, fallopian tubes were normal. Laparoscopic left adnexal en-mass excision with left salpingo-oophorectomy was done. Specimen was retrieved in to as shown in figure 2, via a mini-Pfannenstiel incision.

Outcome and Follow-Up

Histopathology (Figure 3) showed features of secondary degeneration and gangrenous changes in the leiomyomata, left fallopian tube and left ovary due to torsion. Patient was followed up for a period of 1 year who remained asymptomatic throughout with 1st year review abdomen scan showing no abnormalities.

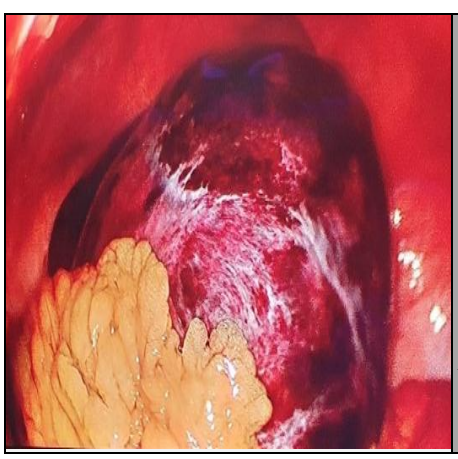

Figure 1. Gangrenous Left Adnexal Mass on Laparoscopic View.

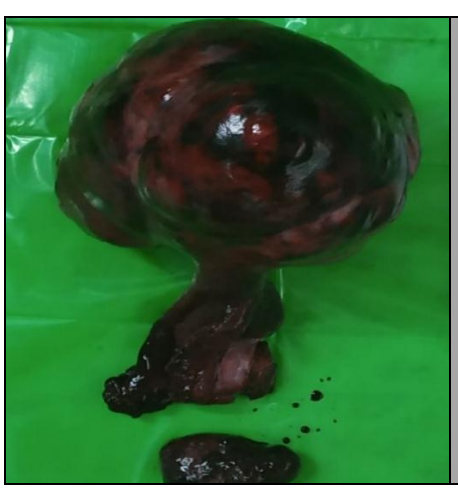

Figure 2. Retrieved Specimen of 14 x 10 cm Solid Gangrenous Adnexal Mass.

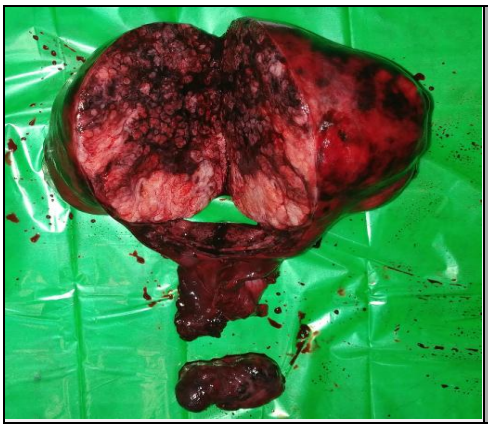

Figure 3. Cut Section of Specimen

DISCUSSION

The first case of leiomyoma of uterine remnant in a patient with Mayer-Rokitansky-Küster-Hauser syndrome was described in 1977 by Beecham and Skeindzeielwski.[2] Approximately 6 % to 10 % of these patients complain of chronic pelvic pain.[4] The cyclic pelvic pain in occurring in patients with MRKH can be due to hematometra, adenomyosis or a large leiomyoma. Torsion of these leiomyomas may present as acute surgical abdomen.[3] Although ultrasonogram of abdomen is the first investigation modality for evaluation of abdominal mass, MRI (Magnetic Resonance Imaging) is found to be diagnostic in evaluation of Mullerian anomalies and to exclude ovarian pathology accurately.[5,6] Diagnosis of such condition is difficult without surgical excision as the ovary cannot be separately identified from these tumors. Hence, the patients with acute abdomen; if found to have MRKH syndrome are to be counselled preoperatively regarding hysterectomy along with adnexectomy considering the torsion of the leiomyoma in rudimentary uterus.[7] The differential diagnosis includes ovarian fibroma, leiomyoma of the extravesical urinary bladder and intestinal GIST (Gastrointestinal Stromal Tumor). Psychological counselling is important to emphasize the fertility potential though most patients may never become pregnant but still they can become a mother through surrogacy.[8] If diagnosed early, excision of leiomyoma of the uterine remnant electively in patients with Mullerian agenesis prevents ovarian resection thereby preserving the fertility.

CONCLUSIONS

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is rare but the risk of torsion of a large pedunculated leiomyoma arising from a small uterine or adnexal remnant is increased secondary to its mass effect. Excision of leiomyoma of the uterine remnant electively in patients with Mullerian agenesis prevents ovarian resection thereby the patient can become a mother through surrogacy in a premenopausal women.

Data sharing statement provided by the authors is available with the full text of this article at jemds.com.
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