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SUCCESSFUL LAPAROSCOPIC MANAGEMENT OF OVARIAN GANGRENE IN MRKH WITH ERAS PROTOCOL : A CASE REPORT.

Gynaecology

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ABSTRACT

The Mayer-Rokitansky-Küster-Hauser ***(MRKH) syndrome*** *is characterized by congenital aplasia 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. Women with this syndrome usually present at young age with* ***primary amenorrhea as first sign*** *hence we are reporting this rare case of ovarian gangrene with MRKH syndrome with* ***late and uncommon presentation****.* With increasing availability of laparoscopic facilities, **successful laparoscopic management** can be undertaken. Enhanced recovery after surgery **(ERAS), ERP or "fast-track" protocol**, aiming to minimize physiologic stress of surgery and optimize rehabilitation of patients can be applied in gynaecological conditions, routinely.

KEYWORDS

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome, Ovarian gangrene, Enhanced recovery after surgery (ERAS)/ ERP /"fast-track" protocol, ERP with laparoscopic surgery

INTRODUCTION:-

Mayer Rokitansky Kuster Hauser syndrome is second most common cause of primary amenorrhea after gonadal dysgenesis. Incidence of MRKH syndrome has been estimated as **1 in 4500 female live births**. In **1829 Mayer** described congenital absence of vagina as one of the abnormalities found in still born infants.**Rokitansky in 1838** and **Kustner in 1910** described an identity in which vagina was absent and small biparitite uterus was present with normal ovaries and anomalies of renal and skeletal system was frequently observed. **Hauser** and associates further described the spectrum of anomalies associated with the syndrome hence over the years this disorder came to be known as Mayer Rokitansky Kuster Hauser syndrome.

MRKH syndrome –

1. **Type I** MRKH - Isolated MRKH or Rokitansky sequence
2. **Type II** MRKH - MURCS association (Mullerian duct aplasia, renal dysplasia and cervical somite anomalies).

MRKH type I syndrome is characterized by complete uterus aplasia in the presence of two rudimentary horns linked by a peritoneal fold and normal fallopian tubes.

Type II MRKH is characterized by uterine hypoplasia accompanied by aplasia of one of the two horns or by a size difference between the two horn rudiments, with tubal malformation of one or both tubes.

Association of MRKH syndrome with **various malignant ovarian masses** (sertoli and epithelial cell tumour) and with **dermoid cyst** of ovary has been reported till date [1]. However very few cases of MRKH syndrome with large twisted ovarian cysts have been reported.

The **etiology** [4] of MRKH syndrome has remained quite unclear until now although the spectrum of malformations encountered suggests developmental field defect involving organ systems which are closely related during embryogenesis. I*ncreasing number of* ***familial cases*** *support hypothesis of genetic cause transmitted as autosomal dominant trait with incomplete penetrance and variable expressivity* More precisely, MRKH syndrome may be attributed to an initial affection of the intermediate mesoderm, consequently leading (by end of fourth week of fetal life) to an alteration of blastema of cervicothoracic somites and pronephric ducts.These latter subsequently induce differentiation of mesonephric and then Wolffian/ Müllerian ducts.

CASE SUMMARY:-

34 year old lady came to our outpatient department with **complaints of** dull aching pain in right lower abdomen since 4 months which was gradual in onset, progressive in nature, nonradiating, not associated with bladder and bowel complains. There was no history of cyclical abdominal pain, bleeding or discharge per vaginum. Her appetite was normal and she had normal sleep pattern and bowel habits. Patient married at the age of 27 years and she got divorced after 4 years of marriage because of inability to conceive. Patient again got remarried 1 year back. There was no history of coital difficulty, dyspyrunia or bleeding while coitus.

On general **physical examination** her height was 155 cm, weight was 56 kg. Breasts showed Tanner stage 5 development, axillary and pubic hair were normally developed. Respiratory and cardiovascular systems were normal.

On per **abdominal examination** there was tenderness present in right iliac fossa on deep palpation, however, there was no guarding or rigidity.

Per speculum examination showed blind vagina, with vaginal length of 6cm. On **per vaginal** examination, the length of vagina was confirmed to be 6 cm, fullness was felt in right adenexa, there was no palpable uterus, while of per-rectal examination, firm to cystic mass felt in right adenexa measuring about 8*6 cm, uterus could not palpated, left fornix was free and rectal mucosa free.

She was a previously diagnosed and worked up case of Type I MRKH, with normal **hormonal profile** (FSH, LH, Prolactin, Thyroid) and XX **karyotype**. Differential diagnosis of MRKH syndrome with right adenexal mass(OVARIAN TORSON/ HYDROSALPHIX) was made.

USG and CT scan (**Figure 1 & 2**) confirmed our clinical suspicion of right sided ovarian torsion, with normal contralateral ovary. Findings suggested an ill-defined mass of 8*6 cm in right adenexa solid to cystic in consistency,with no vascularity in the mass. Uterus was not visualized, in consistent with history of MRKH.

Her routine blood parameters were normal and her hemodynamic status permitted us to go ahead with **laparoscopy** for the same condition. She was enrolled for **ERAS protocol** and further management was started accordingly.



Examination under anaesthesia revealed blind vagina of 6 cm and absent uterus. **Per op** there was rudimentary stump at the site of uterus, suggestive of MRKH syndrome. Gangrenous ovarian mass 8*7 cm irregular surface with three twists (**Figure 3**) at its pedicle arising from right ovary was noted. On untwisting the pedicle, length of the pedicle was found to be `8 cm. The pedicle was coagulated and cut. The necrotic ovary was put in endo bag and retrived through camera port incision, at the end of procedure. Contralateral ovary was normal in size shape and texture. Thorough lavage and suction evacuation of blood clots and necrotic debris was done. Haemostasis was achieved. Her post operative recovery was uneventful, was discharged on day 2 of surgery **(ERAS Protocol** - enhanced recovry after surgery). Follow up call after 72 hrs and visit at day 7, 30 ensured complete recovery. At 3 month follow up visit, patient is symptom free with good QOL.

DISCUSSION:-

According to the intra-operative findings, our patient fitted into category of **MRKH type 1** syndrome. Since patient got married there was no coital difficulty due to adequate vaginal length, patient did not report to hospital for treatment of primary amenorrhoea/ infertility, it was torsion of ovarian cyst which gave rise to lower abdominal pain for which patient reported to hospital,at age of 34 years. Hence we are reporting this rare case of ovarian torsion with MRKH syndrome with **late and uncommon presentation**.

In **literature** association of malignant ovarian tumor have been described with pure gonadal dysgenesis, mixed gonadal dysgenesis & testicular feminizationsyndrome**. Tsaur et al** [2] described combination of MRKH syndrome and immature teratoma of ovary in 4-year girl while **Hyo et al** [3] reported case of serous papillary carcinoma of supernumerary ovary in MRKH syndrome. Few cases of MRKH syndrome with Dermoid Cyst of ovary has been reported till date. At present, ovarian pathologies are not considered to be part of MRKH or MURCS clinical spectrum, as no association between these pathologies and uterovaginal aplasia have been reported so far.

Implementation of an **ERP/ ERAS protocol along with laparoscopic route of surgery** is associated with decreased length of hospital stay, a decrease in rates of postoperative complication, decreased morbidity, and cost savings while preserving patient satisfaction, quality of life and more rapid return to baseline functional status. [5]

CONCLUSION:-

Women with MRKH syndrome suffer **extreme depression and anxiety**, so patient and the relatives should be **properly counselled with empathetic attitude**.We **conserved** contralateral healthy ovary of this patient, and further counseling was offered regarding **in vitro fertilization** with **own oocyte and surrogacy**, for which the patient is planning surrogacy in near future, after sorting out logistic issues. The cost to implementation of **ERAS Protocol (ERP)** is less whereas the benefits are tangible. As we, move towards quality metrics, ERP is the future, along with **minimally invasive surgeries** wherever feasible.

LEARNING POINTS:-

- Mayer-Rokitansky-Küster-Hauser **(MRKH) syndrome** is a rare congenital disorder that shows varying degrees of aplasia or hypoplasia of the uterus and vagina.
- Since the **ovaries are normal, oestrogen-dependent pathologies** such as leiomyomas, adenomyosis and neoplasms can develop in a rudimentary uterus or in Müllerian remnants.
- In cases of MRKH syndrome presenting with a **pelvic mass, differentials** that should be considered include masses arising from the **Müllerian remnants**, and masses arising from the **ovaries**, such as ovarian fibroma, gastrointestinal stromal tumour of the intestine and extravesical leiomyoma of the urinary bladder.
- **USG- Doppler & MRI** has an accuracy of 100% in diagnosing Müllerian anomalies, and can characterise pelvic masses thus guiding further management.
- With advancement in the treatment, **in vitro fertilization with own oocyte and surrogate** pregnancy can be offered to patient of MRKH desiorous of pregnancy, hence **individualse and treat conservatively**.
- In a health care world that is increasingly focused on evidence-based medicine, resource use, and measuring the quality of delivered care, **ERP & minimally invasive operations seems a natural fit** across the surgical specialties.

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