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Primary Amenorrhea In A Patient With Haematometra And Non-Communicating Rudimentary Horn With Cervical Agenesis: A Case Report

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Abstract : *Mullerian agenesis is most common cause of primary amenorrhea.Prevalence is 1 in 4000-10,000 females.Regardless of treatment option selected , patients should be thoroughly counseled and prepared psychologically before initiating treatment. A 25 year old married girl reported to OPD with history of primary amenorrhea and cyclical pain in lower abdomen every month since 3 years.She had history of vaginoplasty 2 years back.Clinically she was found to have well developed secondary sexual characteristics with 2 solid masses one in left iliac fossa and one suprapubically deviated to right side and good vaginal length with blind upper end of vagina.CECT revealed multi-cystic left ovarian mass compressing the vagina with haematometra.Reconstructive surgeries of genital tract such as cystectomy , rudimentary horn resection ,implantation of right tube over right thin part cornu of like part of unicornuate uterus and communication of uterus and vagina was done.On follow up, patient came after 2 months with onset of menstruation. Evaluation for associated congenital renal anomalies should be done.Routine gynaecological care and screening is recommended in patients with reconstructive surgeries.*

Keywords: *Amenorrhea , Mullerian agenesis.*

I. Introduction

Mullerian agenesis, also called Mayer-Rokitansky- Kuster-Hauser syndrome or MRKH, named after August Franz Joseph Carl Mayer, Carl Freiherr von Rokitansky, Hermann Kuster, and A. Hauser, is a congenital malformation characterised by a failure of the mullerian duct to develop, resulting in a missing uterus and variable degrees of vaginal hypoplasia of its upper portion.[1] It is the most common cause of primary amenorrhoea. An individual with this condition is hormonally normal that is, they will enter puberty with development of secondary sexual characteristics including thelarche and adrenarche .Their chromosome constellation will be 46XX.Ovaries are intact and ovulation usually occurs.[2] Typically, the vagina is shortened and intercourse may, in some cases, be difficult and painful. Medical examination supported by gynecologic ultrasonography demonstrates a complete or partial absence of the cervix, uterus, and vagina.

II. Case Report

Our case is Mrs. Meenu w/o Mr. Suraj aged 25 years , resident of Tonk Phatak Jaipur of middle class family.She presented to gynae OPD on 17/02/2014 with chief complaints of Primary amenorrhoea with cyclical pain in lower abdomen every month since 3 years. Patient did not achieve menarche.She complained of lower abdominal pain for 3 to 4 days every month since 3 years.Pain was not associated with nausea or vomiting or any radiating pain of any kind and not associated with any vaginal discharge. Her married life is of 3 years and is nulliparous.Patient had vaginoplasty 2 years back in Ajmer.Her general physical and systemic examination was not significant. On PER ABDOMINAL EXAMINATION - Two lumps felt, one was solid mobile mass felt in left iliac fossa about 8 x 6 cm in size , second mass arising from pelvis.On PER VAGINAL EXAMINATION – there was good vaginal length with blind upper end of vagina and a mass of about 8 x 6 cm size mobile and solid felt in left iliac fossa. Other mass was suprapubically located , deviated to right side.Per speculum examination revealed good vaginal length.CBC , Liver function tests , Renal function tests , Urine examination were normal .CA-125 was raised.Ultrasonography findings showed bilateral ovaries congested , free fluid in POD , 4 x 3 cm cystic lesion seen adjacent to posterior wall of uterus and mild echogenic fluid in endocavity s/o pyometra.CECT revealed multi-cystic left ovarian mass compressing the vagina with haematometra / pyometra.

Diagnostic laparoscopy was done.It revealed 2 uterus – one enlarged and one rudimentery horn communicating with a pedicle with main uterus on right side. Left Hematosalpinx and right fallopian tube were normal . Both ovary were bulky and enlarged.

After all pre operative preparations and consent, patient was taken for surgery.On inspection of peritoneal cavity- 1. Uterus unicornuate 10-12 week size due to haematometra. 2.Left ovary enlarged 10 x 12 x 10 cm forming chocolate cyst. 3.2 x 1 x 3 cm thick band connecting unicornuate uterus to rudimentery horn. 4.Right ovary looking like endometrion connecting to rudimentery horn. 5.Cervix absent and uterus not connected to vagina. After inspection, reconstructive surgery was decided -Bilateral ovarian cystectomy with resection of rudimentery horn and drainage of haematometra was done.Then implantation of right tube over the right thin part cornua of like part of unicornuate uterus and right round ligament implantation over unicornuate uterus done.Passage was created through the vagina to the uterus and catheter was passed through the passage and inflated with 10 ml normal saline.Upper part of vagina was stitched with lower part of uterus.Vaginal catheter was kept for 1 month.Patient was followed monthly till one year to watch for normal cyclic menstruation and formation of vagina.

III. Discussion

Conventional transabdominal, translabial, or transrectal ultrasonography; three-dimensional ultrasonography; and magnetic resonance imaging can be used to evaluate the mullerian structures and characterize anatomy.

In 2–7% of patients with mullerian agenesis, active endometrium is found in these uterine structures. These patients will present with cyclic or chronic abdominal pain. Laparoscopy may be useful in the treatment of patients with functional rudimentary uterine horns.When obstructed hemi-uteri are identified (uterine horns with presence of active endometrium without an associated cervix and upper vagina), then laparoscopic removal of the unilateral or bilateral obstructed uterine structures should be performed. Endometriosis can develop from retrograde menstruation from the obstructed uterine horn, which presents as dysmenorrhea and pelvic pain. Surgical excision results in resolution.Evaluation for associated congenital, renal, or other anomalies is essential as prevalence is 53%.Management includes psychosocial counseling to address the functional and emotional effects of genital anomalies as well as correction of the anatomical defect. Discussion of assisted reproductive techniques and use of a gestational carrier (surrogate) is appropriate.

Non surgical creation of Neo VaginaTiming is best planned when the patient is emotionally mature and expresses the desire for correction. Successful self-dilation requires patients to manually place successive dilators on the vaginal dimple for 30 minutes to 2 hours per day. [3]90%–95% of the patients are usually able to achieve anatomic and functional success by vaginal dilation. Sexually experienced patients may present with natural dilation of the vaginal dimple and occasionally require no additional dilation therapy. Surgical creation of a vagina requires ongoing postoperative dilation or vaginal intercourse to maintain adequate vaginal length and diameter. Aim of surgery is to create a vaginal canal in the correct axis of adequate size and secretory capacity to allow intercourse. Most common is the modified Abbe–McIndoe operation. Procedure involves dissection of a space between the rectum and bladder, placement of a mold covered with a split-thickness skin graft into the space, and the diligent use of vaginal dilation postoperatively which is essential to prevent significant skin graft contracture.[4]The laparoscopic Vecchietti procedure is a modification of the open technique where a neovagina is created using continuous dilation with an external traction device that is temporarily affixed to the abdominal wall .

Figures

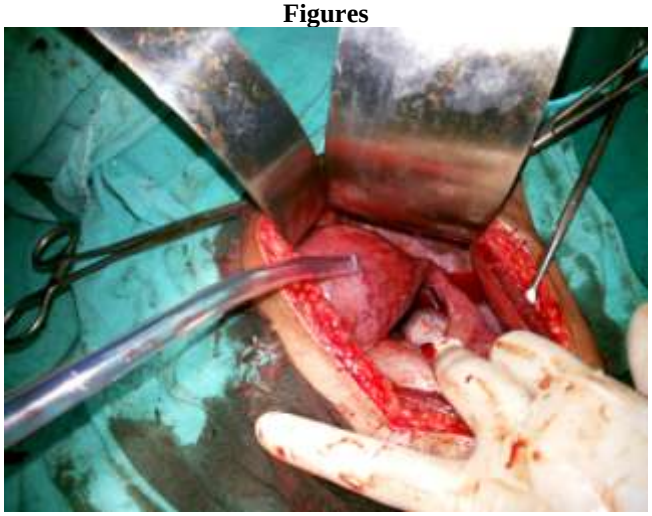

Fig 1: Thick band connecting unicornuate uterus to rudimentory horn.

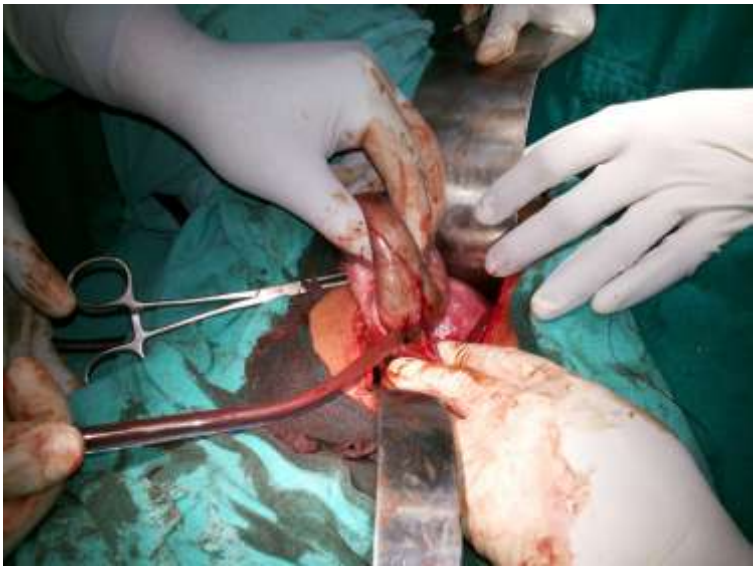

Fig 2: Ovarian cystectomy

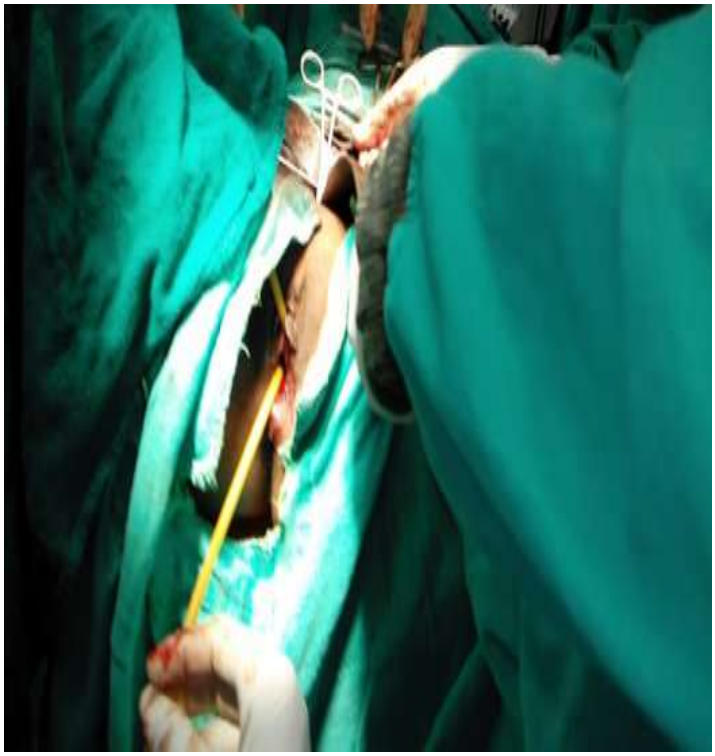

Fig 3: Vaginal catheter introduced.

IV. Conclusion

Evaluation of primary amenorrhea begins with careful history and physical examination including the assessment of the internal and external genitalia as well as determination of FSH, thyroid stimulating hormone (TSH), and prolactin concentrations.[5]Evaluation for associated congenital renal anomalies should be done. Routine gynaecological care and screening is recommended in patients with reconstructive surgeries.Clinicians should be aware of presence of more than one etiology which causes atypical presentations and accomplishes a systematic strategy for the evaluation of amenorrhea potential to avoid long-term side effects.[6]One of the most important steps in the effective management of mullerian agenesis is psychosocial counseling before any treatment or intervention.

References

[1]. Londra L, Chuong FS, Mayer-Rokitansky-Kuster-Hauser syndrome: a review , Int J Womens Health , 2015 Nov 2;7:865-70.
[2]. Akhter N1, Begum BN, Newaz M ,The MURCS Association: Mullerian Duct Aplasia, Renal Hypoplasia and Cervicothoracic Somite Dysplasia , Mymensingh Med J ,2015 Jul;24(3):600-5.
[3]. Callens N, De Cuypere G, De Sutter P, An update on surgical and non-surgical treatments for vaginal hypoplasia , Hum Reprod Update , 20(5): 2014 Sep-Oct; 775-801.
[4]. Kamada Y , Sakamoto A , Tabuchi K , Matsuda M , Opening the Anterior Vaginal Vault: A Novel Approach to Vaginoplasty with a Modified McIndoe Procedure Using an Artificial Dermis, J Pediatr Adolesc Gynecol,28(5), Oct 2015:e119-22.5.
[5]. Bombard DS, Mousa SA, Mayer-Rokitansky-Kuster-Hauser syndrome: complications, diagnosis and possible treatment options: a review.Gynecol Endocrinol, 30(9): 2014 Sep; 618-23.
[6]. Kebaili S1, Chaabane K, Mnif MF: Gonadal dysgenesis and the Mayer-Rokitansky-Kuster-Hauser Syndrome in a girl with a 46, XX karyotype: A case report and review of literature,Indian J Endocrinol Metab,May 17(3),2013,505-8.