Original Research Paper



Gynecology

MULLERRIAN DUCT ANOMALIES & PRESENTATION CASE SERIES

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(ABSTRACT) Mullerrian duct anomalies are congenital structural anomalies of uterus, cervix fallopian tubes presenting different clinical symptoms & signs. At Cama hospital mumbai we reported 4 cases of some of this rarely reported cases of mullerian duct anomalies one of which operated for Haematocolpos drainage & vaginal septum resection.

KEYWORDS:

INTRODUCTION

Female reproductive system develops from pair of mullerian ducts that forms uterus ,fallopian tubes ,cervix & upper 2/3rd of vagina .ovaries & lower 1/3rd of vagina has origin from germ cells of primitive yolk sac & sinovaginal bulbs respectively. Urogenital system develops from intermediate mesoderm so its not uncommon to find mullerian duct anomalies with renal anomalies. normal development of mullerian duct depends upon completion of three phases organogenesis ,fusion ,reabsorption. failure of organogenesis can lead to absent uterus , hypoplastic uterus, failure of fusion can lead to bicornuate ,diadelphis uterus, failure of reabsorption of septum can lead to septate ,arcuate uterus. seconadary sexual characters are normal in mullerian duct anomalies .

Mullerian duct anomalies are uncommon but treatable form of infertility as well as uncommon cause of amenorrhoea. pateient with mda have high incident of repeated first trimester abortion, iugr, malposition, preterm labour.and retained placenta

Details of cases -

Case 1

OPQ 26 yrs MS 6 months came in opd on 20/6/17 with c/o foul smelling blood stained pv discharge since 1 month. Pt has h/o dysparenua. menstrual history s/o regular menses with moderate bleeding painless, Pt nulligravida. No h/o chronic illness in past.

On P/S If forniceal bulge extending along If wall of vagina up to introitus seen. shown in fig 1



On P/V two uterus felt Rt felt normal Lf felt bulky

Pt admitted on 20/6/17 adviced blood investigaions & USG .Blood

investigations WNL. USG pelvis s/o Uterus diadelphys with large collection in vagina s/o hematocolpos, lf ovarian complex cyst most likely hemorrhagic cyst. USG (A +P) s/o uterus didelphys with hematocolpos Rt ovary normal Lf Not visualized, Lf kidney agenesis.

22/6/17 Pt posted for diagnostic laproscopy & Haematocolpos drainage per vaginally On Laproscopy In situ Uterus diadelphys seen, both ovaries normal Both fallopian tubes normal no Intrapelvic bulge seen. shown in fig 2



Operating from vaginally faint vaginal septum ridge seen. 2 cm vertical incision taken on Lf lateral side of ridge to drain Haematocolpos All collection drained out collection Brownish fluid with rotten egg foul smell ,collection sent for culture & sensitivity Procedure stopped as nature of collection not known , extent of cavity , any intrabdominal extent/fistula cant b ruled out .

Pt started on iv antibiotics analgesics post op pt stable .Daily Betadine dushing of cavity done .

On culture minimal growth of Streptococcus spp.susceptible to B lactum, Macrolid. Fluoroquinolones antibiotics .

4/7/17 Pts MRI(A+P) Plain+ contrast done post op,

MRI (A+P) s/o uterus didelphys ,no fluid collection in vagina , complete longitudinal vaginal septum extending from Lf posterolateral vaginal wall to Rt anterolateral vaginal wall with small lf anterolateral compartment & large rt posterolateral compartment septum measures $8\,\mathrm{mm}$ in thickness .

After course of antibiotics & 2 wks vaginal dushing pt posted for vaginal septum resection 10/7/17 vaginal septum resection done under spinal anaesthesia.

After vaginal septum resection single vaginal cavity formed, Two cx visuallised.l.



Pt given iv antibiotics, anagesics ,local antibiotic & anesthetic ointment to reduce post op pain & infection Post op pt stable

24 yr female ABC with c/o cyclical pain in abdomen & breast since 8-9 yrs with h/o primary amenorrhoa.

On p/s examination no cervix seen vagina wall wnl

On p/v blind vagina up to 4-5 cm.

Previous investigations 25.12.17 usg A+P s/o grossly contracted Rt kidney? Absent Rt kidney

Very small infantile uterus? Absent Uterus .one small ovary is visuallised in pelvis other not seen 14.9.16 Karyotyping by G banding s/o 46 XX apperantly normal karyotype 16.1.17 EC renal scan s/o Lf Kidney Normal cortical function & excretion .Absent Rt Kidney 30.8.16 Pt admitted for Diagnostic Laproscopy ,CBC,LFT, RFT, URM, WNL, LH,FSH WNL. 6.7.17 Diagnostic laproscopy s/o Two small horizontally oval bulge in pelvis with fusion medially s/o bicornuate uterus, No cx seen .B/L Ovaries& Fallopian tubes Normal . shown in fig 3



PT adviced excision of Uterus for relief of symptoms & IVF with surrogacy for infertility.

Case 3-

EFG 24 yrs MS 7 yrs nulligravida came with h/o Primary amenorrhoa On examination -P/S Blind vagina with no cx

P/V-Blind Vagina 3-4 cm with no cervix. Uterus not felt.

Previous reports 23.2.17 s/o Hypoplastic uterus with normal ovaries. Agenesis of Lfkidney.

6.1.17 USG s/o? Mullerian agenesis,? Atropic Uterus

19.6.17 karyotyping 46XX Normal Female karypotype

27.6.17 Pt admitted for diagnostic laproscopy

CBC,LFT,RFT,URM, WNL

On Laproscopy Uterus absent, B/L ovaries visuallised normal, B/L fallopian tubes visuallised communicating with each other. shown in



Pt adviced IVF with surrogacy for infertility or vaginoplasty if willing.

Case 4-

KLM 25yrs married since 4yrs, G3A2 with ftg, k/c/o PIH, came with c/o PV discharge, previous scan in ANc period(8/5/17) s/o left kidney not visualized in left renal fossa, ? agenesis, ? ectopic. With SLIUG of 26wks 6days

Pt is k/c/o PIH on tablet Lobet 200mg TDS, and T Nicardia 20mg BD 26/7/17: Pt admitted for safe confinement On Clinical Examination: Uterus 34wks? IUGR with severe PIH Patient posted for elective LSCS i/v/o G3A2 with FTG with IUGR with severe PIH

Intra Op: Left corneal structure, broad ligaments absent., unicornuate uterus with corneal structures and broad ligament only on right side, shown in fig 5



lower segment not formed. Placenta partially adherent.

Outcome: Baby 1.8kg with mother. Post LSCS: Patient stable antihypertensives tapered

DISCUSSION:

Apart from anomalies of ueterus cervix & vagina ,Mullerian duct anomalies are most commonly associated with Renal anomalies anomalies of urogenital system .Mayer Rokitansky Kuster Hauser syndrome type 1 chracterised by isolated absence of proximal 2/3rd of vagina. type 2 marked by other malformations vertebral, cardiac, urologic.

In above few cases associated renal anomalies are also present.

Secondary sexual characteristics well developed & genotype 44XX present.

Commonly presents at young age with primary amenorrhoa. In young married females may presents with primary infertility. As well as in pregnant women can present with IUGR, Malpresentation, Repeated abortions, Retained placenta.

For diagnostic purpose MRI is best option ,HSG ,USG,cab be used as baseline investigations.

Diagnostic Laproscopy is best option to view pelvic organs & decide further line of management.

Management in this cases includes surgical to resection of septum, Heamatocolpos drainage, excision of uterine horns if symptomac . vaginoplasty in case of blind vagina, aplastic vagina, absent vagina done. In Infertility cases IVF with surrogacy is better option, So in cases presenting with amenorrhoa, infertility, BOH posiisblity of Mullerian duct anomalies must be kept in mind.

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