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MULLERIAN ANOMALY AN UNUSUAL PRESENTATION OF MRKH

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A B S T R A C T

Anomalies of the uterus may be congenital or acquired and typically present with abnormalities of the menstrual cycle, pelvic pain, infertility, or pregnancy complications such as primary amenorrohea, preterm labour, recurrent pregnancy loss or abnormal foetal presentation. The true incidence of congenital Mullerian anomalies, of which uterin malformations constitute the majority, is unknown. Prevalence of uterine anomalies ranging from 0.5% to 6% of women. In our case we analysed retrospectively that the endometrium present in the rudimentary mullerian band was responsive because of which the minimal, thickening on the band grew into the size of a normal uterus on giving estrogen therapy & this uterus was not having any connected fallopian tubes or ovaries. We tried to find out the ovaries as the ovaries has to be there though at the various ectopic sites but couldn't get it on laparoscopy.

INTRODUCTION

Anomalies of the uterus may be congenital or acquired and typically present with abnormalities of the menstrual cycle, pelvic pain, infertility, or pregnancy complications such as primary amenorrohea, preterm labour, recurrent pregnancy loss or abnormal foetal presentation. The true incidence of congenital Mullerian anomalies, of which uterine malformations constitute the majority, is unknown. Prevalence of uterine anomalies ranging from 0.5% to 6% of women.[1]

American Society of Reproductive Medicine classifies uterine malformations as follows[2]

TYPE I	Segmental Mullerian Hypoplasia or Agenesis
	A.Vaginal
	B.Cervical
	C.Uterine Fundus
	D.Tubal
	E.CombinedAnomalies
TYPE II	Unilateral Hypoplasia or agenesis (Unicornuate uterus)
	A.Rudimentary horn with cavity,communicating to unicornuate uterus
	B.Rudimentary horn with cavity,not communicating to unicornuate uterus
	C.Rudimentary horn with no cavity
	D.Unicornuate uterus without a rudimentary horn
TYPE III	Failure of Fusion of Mullerian ducts(Uterus didelphys)
TYPE IV	Partial Failure of Fusion of Mullerian Ducts (bicornuate uterus)
	A.Complete (division to internal os)
	B.Partial
TYPE V	Nonresorption of midline septum (septated uterus)
	Complete Septation
	Partial Septation
TYPE VI	Arcuate uterus
TYPE VII	Diethylstilbestrol related anomalies

Case report

History

An unmarried female aged 16 years came to our side in Nov 2015 with chief complaints of primary amenorrohea and nondevelopment of secondary sexual characters i.e. absent breast & axillary hair with scanty pubic hair. Ultrasound pelvis reported uterus and left ovary not visualized but right ovary normally seen.

There was no history of any chronic medical illness or any surgical history in the past.No similar history in her family, belonged to lower middle socio-economic group & was taking Indian vegetarian diet. Her height was 4feet 5 inches. Weight was 45kg. BMI was 22.

Clinical Examination: Breast development: Tanner stage I, pubic hair tanner stage II

Per Abdominal Examination:
Soft, non-tender

Per Speculum:
Cervix could not be well visualized

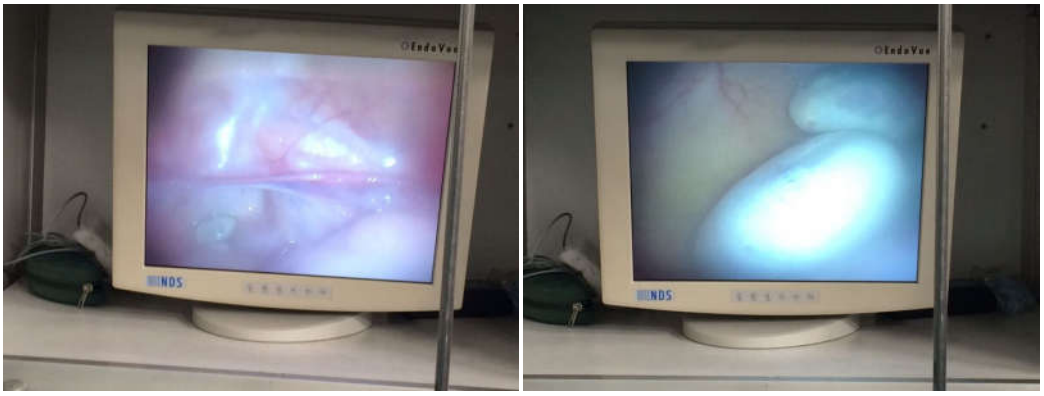
Per Vaginum:
Vagina fully & properly developed
Cervix could not be felt
Uterus not felt

Per Rectal Examination:
Uterus not felt

We investigated her for serum LH, FSH, Prolactin (PRL) & Thyroid profile. It was found that her S.FSH-108.72Miu/ml,S.LH-30.57Miu/ml,S.PRL-15.93 ng/ml & S.TSH-1.09µIU/ml.

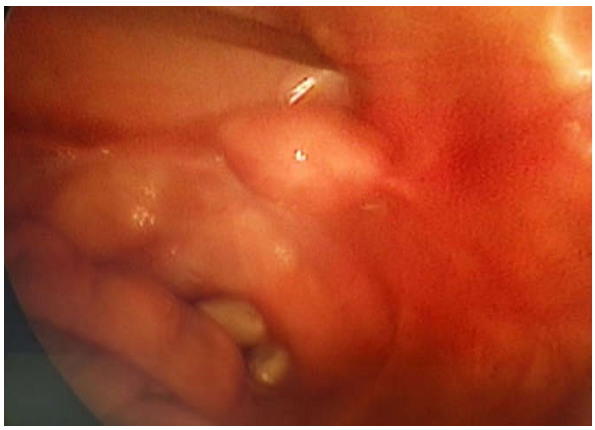
MRI pelvis could not be done due to unaffordability but patient's family were insistent to confirm the presence of uterus, so we planned Diagnostic Laparoscopy for her at our hospital.

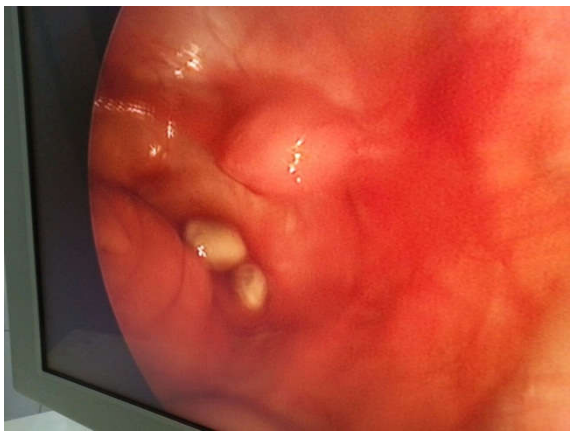
PER-OPERATIVE FINDING :A transverse band suggestive of uterine remnant with minimal central thickening was seen and bilaterally ovaries were not visualized .There were two white coloured oval 2x3x1 cm stone like structures lying in the pouch of Douglas.



There after we kept the patient on estrogen support i.e.ethinyl estradiol .01 mg one tab OD for two months followed by one tab BD for two months and then one tab TDS for further two months for the development of secondary sexual characters and referred her to the deptt of Medical Genetics &Endocrinology, SGPGI, Lucknow.

In the month of July 2016, she came to us with the complain of 2 episodes of cyclicalbleeding per vaginum, breast development tanner stage 3, axillary hair & pubic hairs tanner stage 3.





Her first episode of bleeding per vaginumstarted on 10-07-2016 remained for 8 days, flow-scanty & was associated with pain abdomen for one day. Similarly second episode started on 27-08-2016 remained for five days, flow scanty and was associated with pain abdomen.

Clinical Examination: Breast & Pubic and axillary Hair development Tanner stage III

Per abdominal examination:
Soft, non-tender

Per Speculum:
Two dimples seen at 11 O' Clock & 2 O' Clock position and a bleeding point observed at 11 O'clock position.

Per vaginum:
Vagina fully & properly developed
Two dimples felt at 11O' Clock & 2O' Clock
Uterus not felt

Per rectal examination

A Globular mass of around 5x4 cm felt deviated towards the right side

Management

As her cause of cyclical bleeding was not clear properly & to confirm the origin, we planned relook-laparoscopy for her after properconselling,written& informed consent & full pre-operative preparation.

Per-operative

We found well developed uterus deviated to right side of around 5x4x3 cm but bilateral tubes &ovaries were not visualized along with similar white coloured oval shaped structure of around 2x3x1cm present in pouch of douglas.

We continued the hormonal therapy for her and advised for IVF in future at higher centres and send her for repeat karyotyping for mosaic turner syndrome.

DISCUSSION

These anomalies are secondary to partial or complete absence of development of mullerian ducts. They are frequently associated with renal anomalies.The incidence is 1:4/6000[3]
There are two subclasses in this group:

Complete Bilateral Agenesis or Complete Type I

It is a rare anomaly and incompatible with life because of associated bilateral renal agenesis.

Incomplete Bilateral Agenesis or Incomplete Type I

It is also called Mayer-Rokitansky-kuster-Hauser syndrome, it is the most common presentation of vaginal atresia or

congenital absence of both uterus and vagina,which is also reffered to as Mullerian agenesis or aplasia.This syndrome is characterized by amenorrohea& absence of uterus,cervix and vagina.Sometimes patients have a shallow vaginal pouch,measuring up to 1.5 inches deep.

Most patients have small Mullerian bulbs (rudimentary uterus) without endometrium inside, but in 2-7% of cases, active endometrium is present, giving cyclical abdominal pain and in those cases, rudimentary uterus has to be removed.[4] Typically fallopian tubes or a distal portion of them are present, and normal ovaries are present because of their separate embryonic origin. Normal ovarian function is assosciated with normal development of secondary sexual characteristics.Approximately 15% of women with uterine agenesis also have defects of the urinary system, & 12% have scoliosis[5]

The perineum is usually normal, a recto-abdominal examination confirms the presence or absence of vagina, uteus & ovaries.

Ultrasound is very useful for excluding or confirming the clinical diagnosis and studying the existing portion of the vaginal canal.MRI is a more accurate diagnostic tool because we can evaluate the length of the atresia, the presence of cervix or rudimentary uterus, ovaries and assosciated urologic abnormalities.Laparoscopy is very useful, especially when ultrasound & MRI cannot give us sufficient information as in our case.

In these patients it is also advisable to study the morphology, volume and position of the ovaries as these can be entirely absent as in our case or it can be present but reduced to fibrous strands, or normal but in ectopic position.

CONCLUSION

In our case we analysed retrospectively that the endometrium present in the rudimentary mullerian band was responsive because of which the minimal thickening on the band grew into the size of a normal uterus on giving estrogen therapy& this uterus was not having any connected fallopian tubes or ovaries.

We tried to find out the ovaries as the ovaries has to be there though at the various ectopic sites but couldn't get it on laparoscopy.

Most probably the hypoplastic cervix got developed and therefore the cyclical bleeding was draining into the vagina.

Patient's family contended with the condition, whole scenario explained and kept for follow-up.

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