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A rare case of atretic left ovary with partial congenital absence of ipsilateral fallopian tube with unicornuate uterus

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Abstract

Mullerian anomalies, estimated to be present in 5% -8% of women, represent the majority of female genital tract malformations. Occasionally, Mullerian-tract abnormalities coexist with gonadal developmental disorders. We report a rare case of unicornuate uterus with unilateral ovarian atresia and partial absence of ipsilateral fallopian tube. She underwent laparoscopy for primary infertility evaluation, where this incidental finding was seen.

Keywords: Mullerian anomalies, unicornuate uterus, Laparoscopy

1. Introduction

Embryological malformations of the Mullerian ducts result in an aberration in the normal anatomy of the uterus, resulting in congenital uterine anomalies. During the course of development of the two Mullerian ducts, these aberrations can occur at any of the three stages—formation, fusion and canalisation of the ducts. Depending upon the stage of deviation, Mullerian anomalies can be differentiated from each other^[1]. The prevalence of Mullerian anomalies has been calculated to be 5.5% in general population and 8% in women with infertility, making it an important factor to be considered by obstetrician and gynaecologists treating women for reproductive health and fertility^[2].

As early as the first 6 weeks of intrauterine life, due to absence of the anti-mullerian hormone (AMH) in the female fetus, the Mullerian ducts develop into the female reproductive system. Partial or complete arrest in the development of one of the Mullerian ducts results in the formation of an unicornuate uterus. Furthermore, governed by the type of arrest, unicornuate uterus can be of four types - (i) absent rudimentary horn, (ii) non-cavitary (non-functional) rudimentary horn, (iii) cavitary communicating rudimentary horn and (iv) cavitary non-communicating rudimentary horn^[3-4].

We report a case of a rare association of unicornuate uterus with partial absence of left tube and atretic left ovary, incidentally found on diagnostic hysteron-laparoscopy during infertility evaluation of a patient. Asymptomatic unilateral absence of a portion of a fallopian tube with or without adjacent ovarian agenesis is a very rare condition. The true incidence is unknown. Two etiologic causes are possible. Asymptomatic segmental torsion of the uterine tube and/or ovarian pedicle may occur for uncertain reasons during adulthood, in childhood, or even during the fetal stages. Consequently, torsion may give rise to necrosis and autoamputation. Alternatively, the absence of these organs may be congenital, associated with developmental alterations of the mesonephric and paramesonephric ducts.

2. Case

A 27 year old nulligravida, presented with primary infertility of 9 years. Her husband is a farmer and stays at home with her. Their frequency of intercourse is 3-4 times/week. There is no history of dyspareunia or vaginismus. Her periods were regular and uneventful. She had no significant medical or surgical history. There was no history of a chronic illness or drug intake. She and her husband were both non-smoker and non-alcoholic. Blood Investigations and endocrinal parameters were within normal limits. She underwent hysterosalpingography, which revealed unilateral tubal block and abnormal contour of the uterine cavity (banana sign). (figure 1)

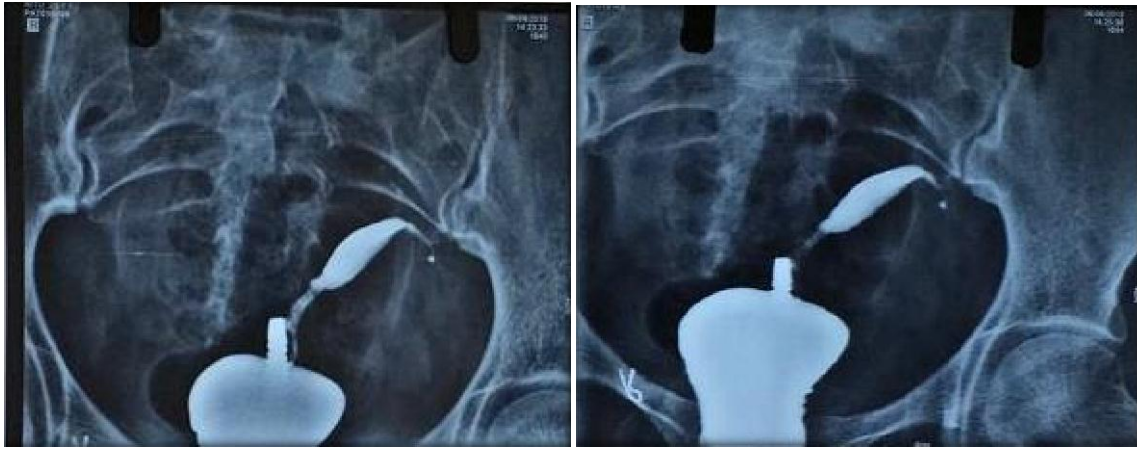


Fig 1: Hysterosalpingogram showing Unilateral tubal block and abnormal uterine cavity contour. (Banana sign)

On ultrasound examination, uterus and right ovary were normally visualised. Left ovary was not visualised. Further on Magnetic Resonance Imaging, an unicornuate uterus with rudimentary right horn (figure 2) and atretic left ovary (figure 3) was reported with no other system anomaly.

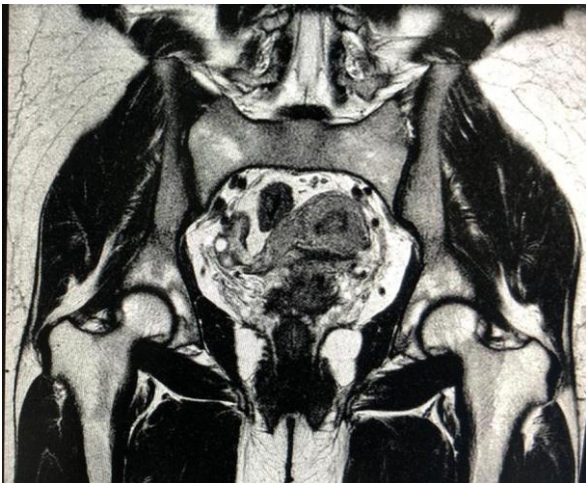


Fig 2: MRI film showing a left unicornuate uterus with rudimentary right horn



Fig 3: MRI Film showing atretic left ovary.

The patient underwent diagnostic laparoscopy with hysteroscopy. During laparoscopy, findings were of an unicornuate uterus with an underdeveloped right horn. Uterine

Fundus was towards left side. Right adnexa was normal. Bladder peritoneum and gut were adherent to the left tube and left round ligament. Left side adhesiolysis was done, which revealed partial absence of tube on left side. Left ovary was not visualised. On chromopertubation, bilateral spill was absent. Hysteroscopy showed a left unicornuate uterus and revealed a patent left cornu. Uterine cavity was normal with healthy endometrium. On the right-side opening was seen, suggestive of a communicating horn. No other system anomaly was detected. Patient was counselled for IVF and discharged the next day.

3. Discussion

Our findings of a left unicornuate uterus with partial absence of left tube and atretic left ovary is a rare condition which was incidentally found during infertility work-up of an otherwise asymptomatic patient, implicates the silent association between unification defects and reproductive outcomes. Therefore, its important to increase a clinicians' awareness regarding Mullerian anomalies, due to their absent to varied symptomatology. A unicornuate uterus is often associated with abortions, preterm labour and malpresentations.⁵⁻⁶ Thus, in order to provide optimal reproductive health advice, counselling and management – possibility of such a rare anomaly should be kept in mind.

4. Comment

Risks associated with unicornuate uterus have been widely acknowledged, including increased risk of miscarriage, ectopic pregnancy in rudimentary horn and adverse obstetrics outcomes. Eventhough, the relationship between unicornuate uterus and infertility remains unclear. Studies to date suggest that 3D USG is also very accurate and can be used as a diagnostic tool; limitations include a possible under-diagnosis of unicornuate uteri and lack of availability in some centres. MRI has a significant role in screening diagnosing congenital uterine anomalies which may become more important in the future.⁷ Combined hysteroscopy and laparoscopy allows for a direct visualization of the internal and external contour of the uterus, and is therefore considered by many to be the gold standard. The main advantage is that it allows concurrent diagnosis and treatment, whereas the disadvantage is the invasiveness of the procedure. Thus, a general awareness of these Mullerian anomaly variants and judicious use of diagnostic modalities among clinicians; can evidently aid in early diagnosis and providing precise management options in reproductive healthcare.

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