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Case Report

A Case of Spontaneous Twin Pregnancy in Unicornuate Uterus with Ipsilateral Left Ovarian and Renal Agenesis

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Abstract

Objective: To present a case of a successful twin pregnancy in unicornuate uterus with ipsilateral ovarian and renal agenesis.

Design: Case report.

Setting: Department of Obstetrics and Gynecology, Maternity hospital, Dr Sulaiman Al Habib Medical Group, Olaya, Riyadh, KSA

Patient(s): A 27-year-old woman P0+3 with history of 3 consecutive recurrent miscarriages came in infertility clinic for detailed fertility work up. She was found to have unicornuate uterus with ipsilateral ovarian and renal agenesis on MRI.

Intervention(s): The patient had spontaneous twin pregnancy, managed well with (Transvaginal shirodkar) cervical cerclage at 16 weeks and progesterone supplementation, ended up in preterm labour and had cesarean section at 32 weeks gestation.

Result(s): Successful twin pregnancy with live birth of twin babies with birth weight of 1.7 and 1.1 kg.

Conclusion(s): We presented a very rare clinical condition that demonstrates a successful twin pregnancy in a unicornuate uterus with no horn and ipsilateral ovarian and renal agenesis concomitantly. The absence of one ovary and one kidney in our case may be explained by the abnormal development of organs derived from a unilateral urogenital ridge. Although the reproductive outcome of women with unicornuate uterus is associated with many obstetric complications, a successful pregnancy is possible.

Keywords: *Congenital Uterine Anomalies; Unicornuate Uterus; Twin Pregnancy*

Introduction

Congenital uterine anomalies result from an abnormal formation, fusion or reabsorption of Müllerian ducts during fetal life. These anomalies are present in 1 to 10% of the unselected population, 2 to 8% of infertile women and 5 to 30% of women with a history of miscarriages [1]. Müllerian duct anomalies (MDA) are uncommon but can be a treatable form of infertility [2]. Patients with MDA are known to have higher incidences of infertility, repeated first trimester spontaneous abortions, fetal intra-uterine growth retardation, fetal malposition, pre-term labor and retained placenta [2].

Regarding embryology the female reproductive tract develops from a pair of Müllerian ducts that form the following structures: fallopian tube, uterus, cervix and the upper two-thirds of the vagina. The ovaries and lower third of the vagina have different embryological origins derived from germ cells that migrate from the primitive yolk sac and the sino vaginal bulb, respectively. Normal development of the Mullerian ducts depends on the completion of three phases: organogenesis, fusion and septal resorption. Failure in any of these phases' results in mullerian duct anomalies. Organogenesis is characterized by the formation of both Müllerian ducts. Failure of this results in

uterine agenesis/hypoplasia or a unicornuate uterus. This type II classification anomaly results from complete or near-complete arrested development of one of the Müllerian ducts [3]. Four possible subtypes can develop: (i) absent rudimentary horn, (ii) non-cavitary (non-functional) rudimentary horn, (iii) cavitary communicating rudimentary horn and (iv) cavitary non-communicating rudimentary horn. The last one may obstruct and present with abdominal pain, subsequently requiring surgical intervention [2,3].

A unicornuate uterus is present in 0.1% of the unselected population. The reproductive performance of women with unicornuate uterus is poor, with a live birth rate of only 29.2%, prematurity rate of 44%, and an ectopic pregnancy rate of 4%. Moreover, women with this anomaly, present rates of 24.3% first trimester abortion, 9.7% second trimester abortion and 10.5% intrauterine fetal demise.

The occurrence of Müllerian anomalies with concomitant gonadal developmental abnormalities is very rare [8,9]. Urinary tract abnormalities occur more frequently in class I and class II uterine anomalies than with those in classes III, IV, or V [10]. Approximately 40% of patients with unicornuate uterus have urinary tract anomalies, usually of kidneys [6,7].

Regarding diagnosis out of all imaging modalities including Hysterosalpingography (HSG), Ultrasound and 3-D ultrasounds, MRI is currently considered the best imaging modality for detection of MDA. It lacks radiation and provides clear delineation of both the internal and the external uterine anatomy. MRI has been shown to have excellent agreement with the clinical diagnosis of the subtypes of MDA [4,5].

The most common classification system is that developed by the American Society of Reproductive Medicine [2,3] (Figure 1).

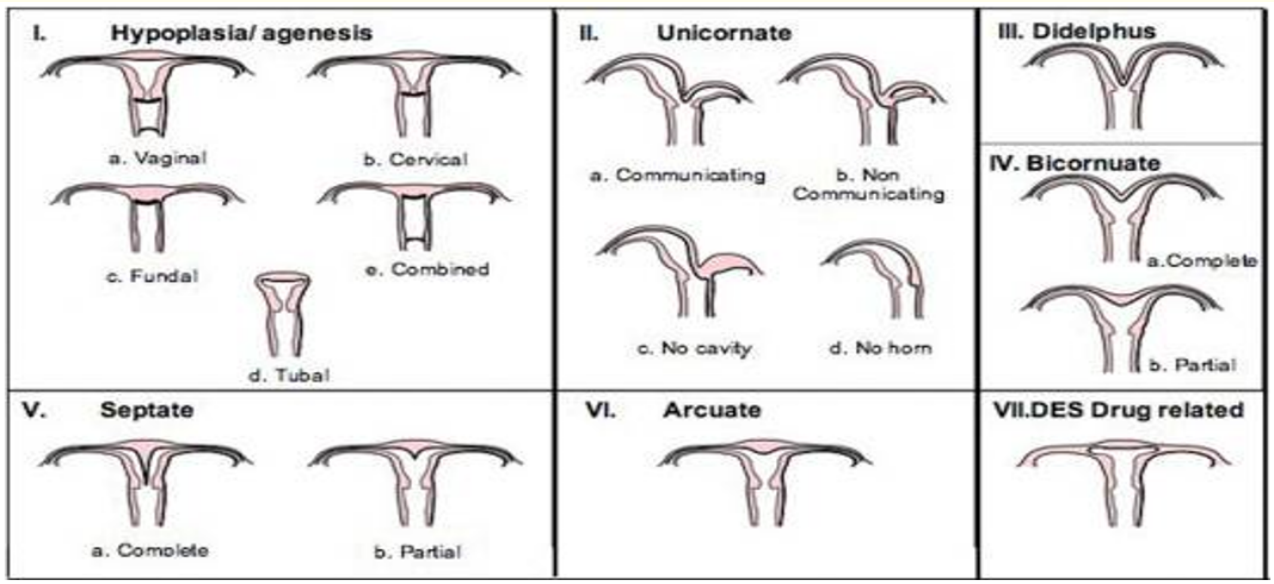

Figure 1: *The classification system of Müllerian duct anomalies used by the American Fertility Society.*

It is important to classify MDA properly because the associated risks of poor pregnancy outcome and treatment can vary widely between anomalies [2,3] (Table 1).

Class	Treatment
I - Hypoplasia/agenesis	No reproductive potential; medical intervention in the form of *in vitro* fertilisation of harvested ova and implantation in a host uterus needed
II - Unicornuate	
Non-communicating, cavitary horn	Always surgically resected, as it is associated with dysmenorrhoea, haematometra, endometriosis and ectopic pregnancy
Non-communicating, non-cavitary horn	Surgery not currently recommended. No complications of endometriosis etc. as there is no endometrium
Communicating, cavitary horn	Also surgically removed because pregnancy that implants in the rudimentary horn rarely is viable
No horn	No treatment. Reproductive potential is possible
III - Didelphys	May consider metroplasty; however, full-term pregnancies have occurred
IV - Bicornuate	Surgical intervention rarely needed; may consider metroplasty
V - Septate	Often treated with transvaginal hysteroscopic resection of the septum. Conception is possible 2 months after surgery

Table 1: *Summary of the therapeutic interventions performed for the different classes of Müllerian Duct anomalies.*

Case Report

This is case report of 27 years old lady, married for 6 years, P0+3 with no alive full term pregnancy and three consecutive recurrent miscarriages visited in infertility clinic for investigations and detailed work up. Regarding her obstetric history, she had three 1st trimester consecutive spontaneous miscarriages (at gestation of 6 weeks, 7 weeks and 8 weeks) followed by evacuation and curettage in last 2 miscarriages. Her menstrual history was unremarkable showing regular cycles with average menstrual flow. There was nothing significant found in medical or surgical history. Both husband and wife are not close relative and are nonsmoker. Family history was also unremarkable. On general examination, patient was stable, nothing abnormal was noticed on abdominal-pelvic examination. Regarding investigations ultrasound report revealed small uterus, absent left fallopian tube, right ovary was normal in texture and size while left ovary was not visualized, right kidney was compensatory hypertrophied while left kidney was absent. Husband semen analysis was normal. Her hormonal profile including (FSH, LH, Prolactin, and Testosterone) was within normal range. Antiphospholipid (APS-LA, ACA, B2 glycoprotein) antibodies were not found positive. Infection TORCH screening was negative. Hysterosalpingography (HSG) report conducted in 2010 was showing unicornuate uterus and right sided patent fallopian tube with free spill while left fallopian tube was not visualized. MRI report was showing unicornuate uterus with absent left fallopian tube and ovary and unilateral left renal agenesis.

In the next appointment she came back to review the result of investigations, in the same visit she complained that she missed her last period and has become overdue almost 20 days (secondary amenorrhea) as last menstrual period was 2-3-14, so serum BHCG was advised to rule out pregnancy, showing result of 849. Subsequently as she found pregnant so was booked for antenatal care, advised ultrasound and all routine booking investigations as well as advised to take low dose aspirin 81 mg orally and Innohep (LMWH) 3500 iu subcutaneously once daily in view of three consecutive 1st trimester recurrent miscarriages to improve pregnancy outcome. Viability scan was revealing twin viable intrauterine diamniotic pregnancy of 6 weeks with CRL OF 0.33 cm and 0.37 cm. She was diagnosed as spontaneous twin pregnancy and her expected date of delivery was calculated as EDD = 9-12-14 by dates and by 1st scan EDD = 11-12-14 at 6 wks. Proluton depot 500 mg injection was also initiated on weekly basis. Ultrasound done at 15 weeks was showing viable monochorionic diamniotic MCDA twins with short cervix of 1.5 cm. Transvaginal shirodkar cervical cerclage was applied at 16 weeks gestation to avoid second trimester miscarriages and preterm labour. After that she had an uneventful pregnancy with close antenatal surveillance. Obstetric ultrasound examinations at the first, second and third trimester of her pregnancy showed a normal insertion of the placenta, normal amniotic fluid index and viable twin gestation. At 32+5 weeks gestation she was presented with preterm pre labor rupture of membranes since 2 hours with no abdominal pain, fever and any other symptom in emergency department, admitted in hospital for observation, pro-

phylactic intravenous antibiotics and fetal close monitoring. She had already received 2 doses of steroids almost 1 week ago in view of twin gestation to improve lung maturity and neonatal outcome. At time of presentation ultrasound was showing 1st Twin 1 breech, 2nd Twin 2 cephalic, parameters of T2 were below 10th percentiles corresponding to 29 weeks with oligohydramnios of 5 cm while parameters of T1 were between 10th - 50th percentiles equivalent to 31+ weeks, umbilical artery doppler: Twin 1 - S/D ratio: 2.7; RI: 0.62 and Twin 2 - S/D ratio: 3.19; RI: 0.68, estimated fetal weights were 1732 gm for T1 and 1298 gm for T2, other findings were normal.

In labour ward after few hours she started to contract, CTG was showing reactive trace for both twins with regular contractions. Initially conservative management was done with hydration and analgesia but preterm contractions continued so Emergency cesarean section was conducted at 32+5 weeks gestation by giving C shaped incision in lower uterine segment and both twins were delivered safely. Shirodkar cervical suture was removed at same time. Outcome was Twin 1 delivered as breech baby girl, weight 1750 gm, A/S 6/8 while Twin 2 delivered as cephalic baby girl weight 1150 gm, A/S 5/8 at 1, 5 minutes. Both babies were examined by paediatrician and shifted to neonatal intensive care unit NICU. They stayed in NICU for one month and then discharged safely home. Patient was discharged home on 4th postoperative day and postnatal period was uneventful.

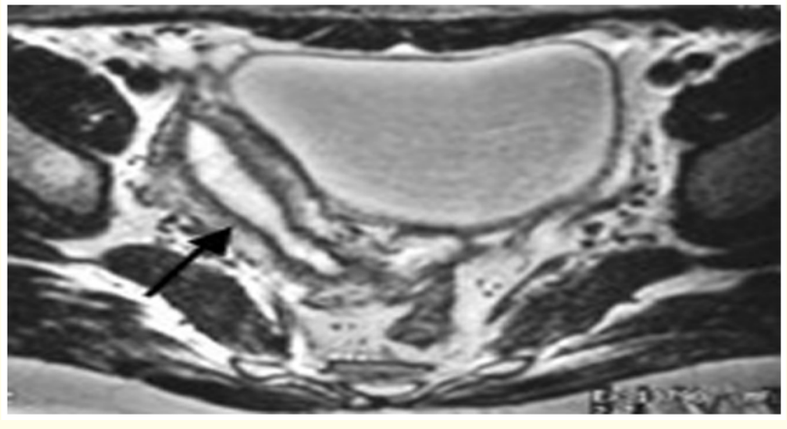

Figure 2: *MRI report.*

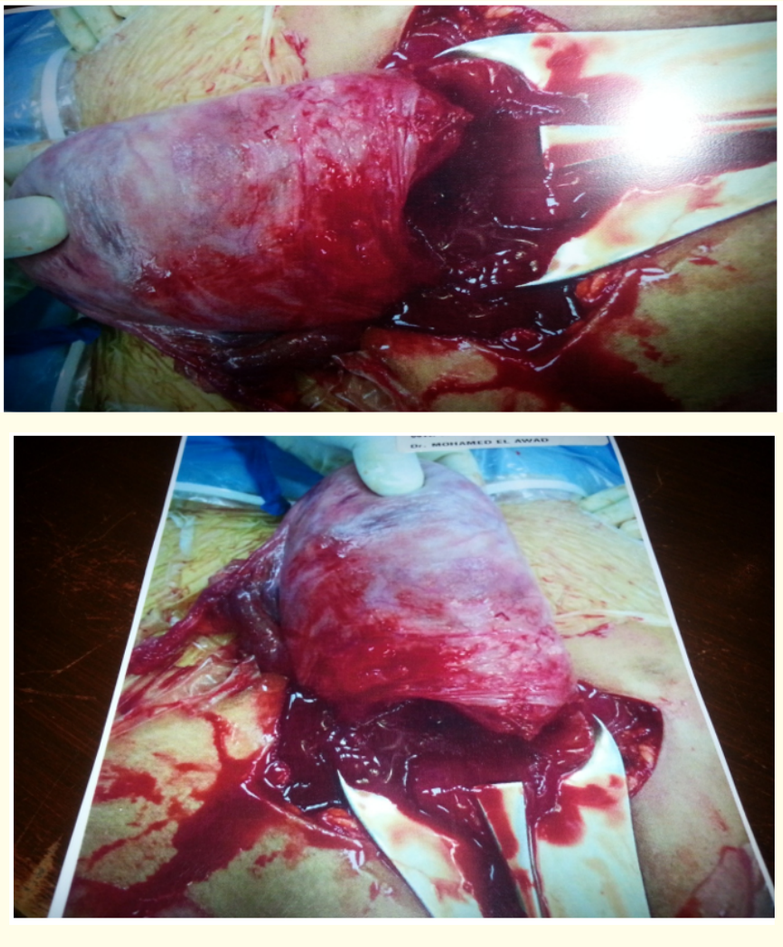

Figure 3: *Unicornuate uterus with ipsilateral left sided ovarian agenesis.*

Discussion and Conclusion

We present a case report of successful twin pregnancy in woman with unicornuate uterus together with ipsilateral ovarian and renal agenesis.

Case reports about the unicornuate uterus with gonadal and renal abnormalities are very limited in the literature. Firstly this case is representing a rare clinical condition that demonstrates a unicornuate uterus without rudimentary horn and ipsilateral ovarian and renal agenesis concomitantly. There is a strong association of mullerian anomalies with urinary tract anomalies but the occurrence of Müllerian anomalies with concomitant gonadal developmental abnormalities is very rare [8,9]. Urinary tract abnormalities occur more frequently in class I and class II (unicornuate) uterine anomalies than with those in classes III, IV, or V as seen in our case [10]. The absence of one ovary and one kidney in our case may be explained by the abnormal development of organs derived from unilateral urogenital ridge.

Another interesting finding in our case is spontaneous twin pregnancy in unicornuate uterus indicating that these women can have successful spontaneous pregnancy although there was a history of three consecutive abortions. Actually decreased muscle mass in unicornuate uterus results from failure of complete development of one of the Müllerian ducts. In unicornuate uterus with no horn, no surgical resection/treatment is required as spontaneous pregnancy potential is possible as seen in this case.

The unicornuate uterus is an infrequent type of Müllerian anomaly, which is due to a failure of development of one Müllerian tract. The unicornuate uterus represents 6.3% of congenital uterine anomalies, which have significant problems with reproductive outcome secondary to abnormal uterine vasculature and decreased muscle mass [8,9]. It has been suggested that first trimester abortion, intrauterine growth restriction, and stillbirths, may be explained by an abnormal uterine blood flow (absent or abnormal uterine or ovarian artery). Second trimester abortions and preterm deliveries are thought to be due to decreased muscle mass in the unicornuate uterus as well as cervical incompetence.

Cervical shortening and preterm labour are observed in this case as well as there was history of three consecutive miscarriages indicating that spontaneous abortion and premature delivery rates are very high in patients with unicornuate uterus (48% and 17%, respectively) with a total live birth rate of 40% [10].

The utility of ultrasound cervix length measurement for assessing the risk of preterm birth has been well documented, with an accepted cutoff value for cervix length of ≤ 25 mm before the 24^{th} week of gestational age. Cervical cerclage is the best treatment for women with a short cervix (< 25 mm), and particularly for women with a history of prior mid trimester pregnancy losses due to cervical insufficiency, Therefore, in our case report, cervical shortening is assessed by serial ultrasound cervical length measurement and then managed by classical (shirodkar) cervical cerclage at 16 weeks gestation.

Regarding preterm labour seen in our case, accumulating evidence suggests that the myometrial activity associated with preterm labor results primarily from a release of the inhibitory effects of pregnancy on the myometrium rather than an active process mediated through the release of uterine stimulants, and progesterone appears to play a central role [1].

Recent data suggest that progesterone may be important in maintaining uterine quiescence in the latter half of pregnancy by limiting the production of stimulatory prostaglandins and inhibiting the expression of contraction-associated protein genes (ion channels, oxytocin and prostaglandin receptors, and gap junctions) within the myometrium. However the role of progesterone in later pregnancy, however, is less clear. In fact, ACOG recommend progesterone supplementation only for prior spontaneous preterm birth and cervical shortening (≤ 15mm prior to 24 weeks) so we also administered progesterone treatment in form of weekly injections in our present experience [1].

According to the current guidelines of the American Congress of Obstetricians and Gynecologists (ACOG) for the management of IUGR [11], it is reasonable to consider serial growth ultrasound examinations in pregnancies at risk of IUGR and to rule out malpresentation as there is high risk of these complications in unicornuate uterus pregnancy.

Our case report shows that by adopting these strategies serial cervical length measurements, insertion of cervical cerclage, serial growth measurements by ultrasound and progesterone supplementation) the prognosis of pregnancy in a unicornuate uterus is not always impaired, although cervical incompetence, complications related to twin pregnancy, prematurity and cesarean delivery threatens to occur. But the reproductive outcome of women with unicornuate uterus is still associated with obstetric complications, a successful pregnancy is possible.

Nevertheless, the optimal management approach cannot be clearly stated. Further large observational and prospective studies are essential to investigate the treatment modalities needed during pregnancies in this uterine anomaly.

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