



A CASE REPORT OF A MULTIPARA WITH MULTIPLE OVARIES AND TUBES ASSOCIATED WITH A NORMAL UTERUS FOUND AT SECOND REPEAT CAESAREAN SECTION.

¹Osaze O. Oboigba, ²Nosakhare O. Enaruna, ²Jedidiah D. Sodje

¹*ENOS Hospital, Plateau, Nigeria.*

²*Department of Obstetrics and Gynaecology, University of Benin Teaching Hospital, PMB 1111, Benin City, Nigeria.*

ABSTRACT

Duplication of the tubes and ovaries are rarely reported perhaps due to their association with other major anomalies or being missed or overlooked because of the lack of clinical relevance. While female genital tract anomaly is likely to suggest a sense of challenge with fertility, the presence of multiple tubes and ovaries might indeed increase the frequency of ovulation and pregnancy, including ectopic pregnancy. We present a case of a woman with multiple ovaries and tubes associated with a normal uterus found at a second repeat caesarean section who was unaware of the condition.

INTRODUCTION

Congenital anomalies of the female reproductive system may involve the uterus, cervix, fallopian tubes, ovaries or vagina. Depending on the specific defect, a woman's obstetric and gynaecological health may be adversely affected. Uterine anomalies are the most common of the Mullerian anomalies, but the true incidence is not known since many women are asymptomatic.¹ Uterine anomalies are associated with both normal and adverse reproductive outcomes; they occur in approximately 3–4% of fertile and infertile women, 5–10% of women with recurrent early pregnancy loss, and up to 25% of women with late first or second trimester pregnancy loss or preterm delivery.²⁻⁵ Overall, uterine anomalies are associated with difficulty maintaining a pregnancy, but do not usually impair the ability to achieve a pregnancy.^{3,6} The ovaries and tubes are rarely seen during routine obstetric scans for foetal growth and wellbeing done usually by the abdominal route. However, during caesarean section, the presence of duplicated

adnexal structure can be easily observed. Such discoveries are startling but have been rarely reported. The origin of this anatomical variation established during embryogenesis in the female reproductive tract might reflect genetic aberrations. For example, it is possible that the additional ovaries and Fallopian tubes contain a mixture of genetically different tissues, formed through a process of early fusion of embryos (chimera).⁷

The embryological development of the female reproductive tract can provide some explanation for the anomaly of reproductive organ duplication. Gametes are derived from primordial germ cells (PGC) that are formed in the epiblast during the second week and are thereafter moved to the wall of the yolk sac. During the fourth week, these cells begin to migrate from the yolk sac toward the developing gonads through the dorsal mesentery, where they arrive by the end of the fifth week. Mitotic divisions increase their number during migration and when they arrive in the gonad. The importance of this is that if they fail to reach the gonadal ridges, the gonads do not develop. Hence, the PGC have an inductive influence on the development of the gonad into an ovary.⁸ It can be inferred that the gene which codes for this expression has a tendency of being doubly expressed either from genetic mutation or by an

Corresponding Author: DR. NOSAKHARE O. ENARUNA

*Department of O&G,
University of Benin Teaching Hospital,
P.M.B. 1111, Benin City, Nigeria.
E-mail: nosaenrus@gmail.com, Tel: +2348033835761*

unknown predisposing factor. There is, however, the possibility of the reunion or disunion of these gametes during the process of migration, and after complete division of twins entrapped in a single zygote, this can result in a double expression of the gonads at the gonadal ridge.

The paramesonephric ducts arise as a longitudinal invagination of the epithelium on the anterolateral surface of the urogenital ridge. Cranially, the duct opens into the abdominal cavity with a funnel-like structure. Caudally, it first runs laterally to the mesonephric duct, crossing it ventrally to grow caudo-medially.⁸ It is also possible that double expression in the gene regulating the signalling for the epithelial invagination results in formation of multiple paramesonephric ducts, thus leading to multiple fallopian tubes. It is, however, not clear how the uterus remains a fused organ in the setting of multiple adnexal appendages. Perhaps the usual finding of uterine anomalies without involvement of the tubes or ovaries implies the variable and isolated contributions of the different parts of the reproductive tract to the developmental anomalies. Ovarian duplication may also occur in conjunction with the duplication of genital ridge, and probably from a duplicated Mullerian duct, but this has rarely been reported in our environment.

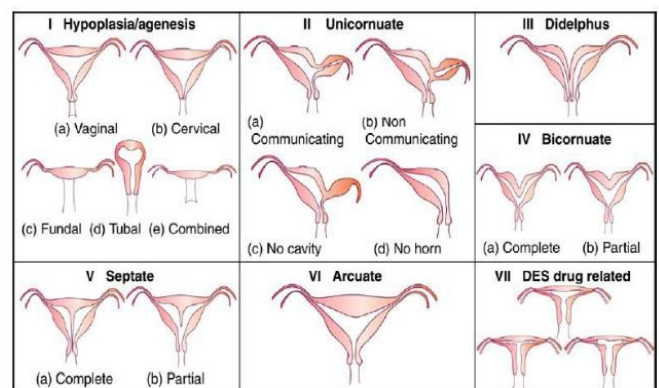
Mullerian anomalies affect 4% of females and congenital anomalies of the female reproductive tract are typically classified into three main categories: agenesis and hypoplasia, lateral fusion defects, and vertical fusion abnormalities. A fourth group is composed of women exposed to diethylstilbestrol (DES) in utero.^{9,10} Agenesis and hypoplasia can occur with any or multiple Mullerian structures as seen in approximately 1 in 5,000 to 1 in 40,000 girls in a condition such as Mayer-Rokitansky-Küster-Hauser Syndrome.¹¹ Lateral fusion defects occur due to failure of migration of one Mullerian duct, abnormal fusion of the Mullerian ducts, or absorption of the intervening septum between the ducts. This is the most common category of Mullerian defects and can result in symmetric or asymmetric and non-obstructed or obstructed structures.¹² Depending on the population studied and the imaging modalities used, the most common uterine malformations are arcuate, septate, or bicornuate uteri (fig.1).^{2-4,6} Vertical fusion defects result from abnormal fusion

of the Mullerian ducts with the urogenital sinus, or problems with vaginal canalization. These conditions can cause menstrual flow obstruction. (Fig. 1).¹³

The aetiology of congenital anomalies of the female reproductive tract is poorly understood. Karyotypes are normal (46XX) in 92% of women with Mullerian anomalies, and abnormal in 7.7% of these women.¹⁴ Most of these developmental abnormalities are infrequent and sporadic and are thus attributed to polygenic and multifactorial causes.¹⁵ The diagnosis of these conditions requires a high index of clinical suspicion as many of the anomalies are accidental findings. These occasional discoveries will certainly enrich the advancing medical literature, and probably add to the knowledge of anatomical ambiguity which may be linked to less obvious dysmorphic attributes. The diagnosis of such a condition is frequently missed during antenatal care, despite the routine use of abdominal ultrasonography. However, it is important to detect such an anomaly considering the possibility of fertility challenges, as well as increased rates of multiple pregnancy and ectopic gestation.

This is a report of a multipara who had four ovaries and four Fallopian tubes in a mirror-image fashion and had had two normal vaginal deliveries followed by two caesarean sections. She had a repeat caesarean section in the 37th week, indication being bleeding placenta praevia, and with a favourable maternal and neonatal outcome.

Fig 1: The American Fertility Society classifications of mullerian anomalies



Case presentation

A 37-year-old multipara with two living children was verbally referred from a primary health centre with a diagnosis of antepartum haemorrhage at 36 weeks gestational age.

She booked index pregnancy at 20 weeks gestation at the referring centre and had regular uneventful antenatal care visits until about 36 weeks when she started bleeding. She was unaware of results of her booking investigations. She had tetanus toxoid immunization twice during index pregnancy. Ultrasound scan done at about 20 weeks was normal, and no further scans were done. She had had 4 deliveries and 2 miscarriages between 2002 and 2018 with the last two deliveries requiring emergency caesarean section (CS) for prolonged obstructed labour, outcome being fresh stillbirths. Following the last pregnancy and delivery she commenced Depo-Provera for contraception but discontinued it in order to get pregnant. There was no other significant history.

On examination, she was anxious, afebrile with a temperature of 36.9°C. She was not dehydrated but pale clinically. Pulse rate was 96bpm, respiratory rate was 26cpm and breathe sounds were normal. The abdomen was enlarged with left flank bulge. It moved with respiration, and there was no tenderness. The symphysio-fundal height was 36cm and was compatible with the gestation. There was a single foetus in oblique lie, cephalic presenting in the right occipito-posterior position with the head five-fifths palpable per abdomen. There was no contraction in 10 minutes. The foetal heart tone was heard and regular at 135bpm with the foetal stethoscope. The estimated foetal weight was 3.5kg clinically. The vulvo-vagina was smeared with blood but bleeding had reduced. And no further vaginal examination was done.

We entertained a clinical impression of antepartum haemorrhage secondary to a low-lying placenta. She was counselled on the diagnosis and options of management were discussed. She gave consent for an elective CS.

Her laboratory results showed a haematocrit of 25%, white blood cells 5700 cells/mm³, platelets 307 cells/mm³, serum urea 35mg/dl and serum creatinine 0.5mg/dl. Her serum electrolytes, bedside clotting time and urinalysis were all within normal limits. Her retroviral screening was negative, the random blood sugar was 120mg/dL,

blood group was B Rhesus D positive and genotype was AA.

Fig. 2: From right to left; the right round ligament, the right anterior Fallopian tube and ovary with infundibulum, and the right posterior Fallopian tube and ovary lying on the infundibulum

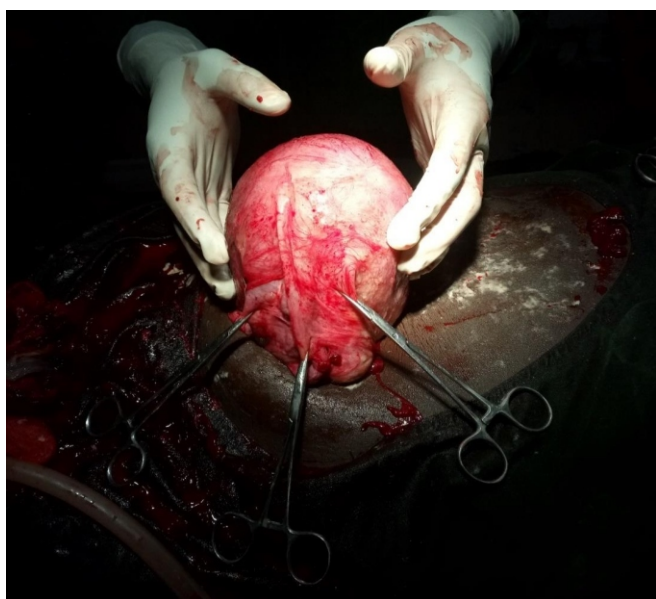


She was transfused with two (2) units of blood and was given 12mg of injection Dexamethasone 12 hourly for the next 48 hours. She had an emergency CS at 36 weeks and 5 days following another episode of unprovoked bleeding per vaginam. A midline incision was utilized, and findings included adhesions over the lower uterine segment, intact amniotic membrane, clear amniotic fluid, a live male 3.4kg neonate in good condition, and placenta previa type 2 anterior. Uterus was about 20 weeks' size with double right and left ovaries attached, along with double Fallopian tubes, the ipsilateral inter-tubal distance was approximately 6cm for each pair of tubes. The anterior tube was about 3cm above the posterior tube on each side, with the ipsilateral round ligament emerging 6cm below the anterior tube. The estimated blood loss was 1.2 L.

She had an uneventful post-operative recovery and was discharged to the lying-in ward the following day, beyond which she continued to make steady recovery. She was counseled on the outcome of delivery and other operative findings on the second postoperative day and was discharged home 8 days after surgery in good condition. She was seen at the

6th week postnatal clinic and in good condition. A pelvic scan showed the uterus was normal size and non-gravid. She was further counseled and discharged from follow up with a referral to the family planning clinic for detailed contraceptive advice.

Fig. 3: From left to right; the left round ligament, the left anterior Fallopian tube and ovary (ovary not very visible due to adhesions on the uterus), the left posterior Fallopian tube and ovary (ovary partially visible)



DISCUSSION

The presence of multiple adnexal structures discovered at caesarean section will create a disturbing sense of missed diagnosis, although the clinical relevance of such discovery will not be immediately apparent. There is however no unique approach to the management of duplicated tubes or ovaries accidentally found during laparotomy (Fig 2 and 3), and in index case, consent was not given to remove the extra tubes or ovaries. Without a high index of suspicion, antenatal diagnosis is unlikely, but a combination of investigative modalities such as salpingo-hysterography, ultrasound scan and laparoscopy will invariably play a pivotal role in the diagnosis and further management of this condition. Salpingo-hysterography can reveal the number of patent tubes and partially occluded tubes but will not be able to show the number of non-patent tubes, especially those whose non-patency ensued at the endometrial cavity. Thus, the presence of normal

looking patent tubes found at radiological studies does not rule out Mullerian abnormalities involving extra tubes which invariably will be non-patent or non-communicating with the endometrial cavity.

Ultrasound scan remains the gold standard technique for studying the ovaries and developing follicles, as well as excluding gross ovarian abnormalities. It is possible that extra ovarian tissue can be detected with ultrasound scan but without a high index of clinical suspicion, this is likely to be missed. This is especially so because in the face of ovarian hyper-stimulation, it is desirable to have multiple follicles, a situation that allows the ovaries to enlarge beyond the ovarian fossae thereby obscuring any chance of discovering adjacent extra ovarian tissue. In the patient we reported, there is no evidence of a pelvic scan done to monitor ovulation, and she did not have any record of early scan for pregnancy confirmation, situations which could have allowed detection of extra ovaries in her. Perhaps a deliberate attempt by the scan operator to search for adnexal anatomical aberrations beyond the clinical interest of folliculometry or confirmation of a pregnancy during abdominopelvic scanning will be instructive in the occasional discovery of multiple adnexal structures in some individuals.¹⁶

Laparoscopy, a modern tool of inestimable value in the diagnosis and treatment of several gynaecological conditions, is a useful modality in detecting duplicated organs or presence of extra tissues. It is however not readily available or accessible, especially in settings of poor resource which is prevalent in our environment. Where endoscopy is provided, it is usually at a very prohibitive cost, and so cannot be applied in many cases where it will be needed. Until laparoscopy and other endoscopic interventions become commonly available, a lot of what clinicians offer to their clients will continue to rely on astute clinical evaluation and sound clinical judgement, important qualities for the proper execution of patient care in low resource settings. Unfortunately, without associated complications, duplicated organs do not present as clinical features discernible through history and physical examination.

The diagnosis of duplicated adnexal organs is made more difficult in situations where pregnancies are achieved without prior management for infertility. Whereas gynaecological conditions often warrant

investigations to confirm diagnosis, decide on appropriate treatment, and evaluate progress of the condition, a pregnant woman presenting for antenatal care is often provided care without involving many investigations. And in many instances where radiological tests are requested, significant concern about safety for the foetus also exists. Moreover, the procedure of foetal examination with scan is often prescribed to be as safe as reasonably allowed (ALARA), an easy excuse for lack of details beyond observing foetal cardiac activities and foetal movements.¹⁷

Laparotomy for caesarean section will provide the best opportunity to find out about abnormal development of the uterus, its appendages and ovaries. Should diagnosis be made at caesarean section, appropriate intervention can be executed immediately; and follow up can be planned to allow prompt and timely intervention as appropriate. The recovery from the effect of pregnancy after the puerperium will probably make the pathology less obvious. However, after the puerperium, and with the resumption of ovarian function, knowledge of any anomaly will direct further follow up with standard investigative modalities.

It will appear like the main concern regarding duplicated organs will be the likelihood of other congenital anomalies,¹ or the risk of complications resulting from such anomalies such as malignant transformation. It is plausible to believe that with increased number of body parts such as ovaries and tubes, which are particularly prone to mitotic changes, there is also a chance that the risk of malignancy is similarly increased. Perhaps the unique roles of genetic studies and immuno-histochemistry can leverage the recent advances in cancer therapy and serve as a vista into further studies to determine the risks associated with extra adnexal structures.¹⁸

The importance of detecting these anatomical abnormalities to enable appropriate management cannot be overemphasized. Considering the possibility of increasing the individual risk for morbidity such as ovarian or tubal ectopic gestation, as well as the chance of increased risk of malignancy in some ectopic tissue like the ovary, it remains instructive to apply a high index of clinical and/or investigational radiological suspicion in all settings of evaluation of the patient in obstetric and gynaecological practice

There is the need for awareness by physicians to promptly examine for abnormal anatomical abnormality during caesarean section or other gynaecological surgeries and should not always be in a hurry to return the repaired uterus into the peritoneal cavity, as this can contribute to the problem of patient misdiagnosis despite prompt management. Such time spent on observation and appropriate intervention can save the patient from further gynaecological and obstetric challenge, as well as financial burden.

CONCLUSION

Female reproductive tract developmental anomaly such as duplications of the tubes and ovaries may not have any possible adverse effect on subsequent reproductive functions of a woman, regarding conception and the normal growth and development of a foetus. However, awareness of variations of the female anatomy might prove instructive in the occasional discovery of unusual findings, hence limiting the impact of misdiagnosis, and promoting continued vigilance and appropriate management.

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Authors' contributions

OO was involved in the initial drafting, writing and pre-editing of the manuscript. NE contributed to the final writing and edited the manuscript. JS contributed to the final writing and co-edited the manuscript. The three authors read and approved the final manuscript.

Consent for publication

The patient gave written informed consent for the publication of the case report as well as the images.

Competing interests

The authors declare that no competing interests exist.

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