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Case Report

Uterine Didelphys, an Uncommon Mullerian Duct Anomaly in a Multiparous Woman with Endometrial Carcinoma

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Abstract: Uterine didelphys is a rare deformity with most cases associated with favourable pregnancy outcomes. Uterine carcinoma in such uterine malformation is equally rare with many cases diagnosed late due to failed pre-surgery evaluation associated with routine endometrial biopsy. We report a case of incidental uterine didelphys found during surgery in a 62-year-old para 2 +3, woman with endometriod variant of endometrial carcinoma of the right uterus (grade II, and at least stage III). She, successful, had surgery and currently undergoing adjuvant chemoradiation with no complaint. This case demonstrates the possibility of uterine didelphys in a patient with endometrial cancer even when there has been successful pregnancies and deliveries. Clinicians should be aware of this rare possibility especially in a background of preceding repeated miscarriages. Diligently performed endometrial biopsy can still yield adequate specimen for pre-surgery histological diagnosis despite the documented high failure rate associated with the procedure. Where available, contrast-enhanced magnetic resonance imaging is a useful pre-surgery evaluation test with option of exploratory laparotomy performed to avoid delay in diagnosis and treatment of cancers where this is not available but with high index of suspicion.

Keywords: Uterine Didelphys, Endometrial Cancer, Case Report, Incidental Finding

1. Introduction

Uterine didelphys is a component of Mullerian duct anomalies (MDA) spectrum arising from the complete non-fusion of both Müllerian ducts, resulting in two distinct uteri [1, 2] The human uterus is of paramesonephric origin and failure of fusion of the Mullerian ducts or subsequent resorption of the tissue results in a spectrum of MDAs. Didelphys uterus occurs in 0.5 -5% of the general population [3] and is associated with more cases of infertility than normal uterine anatomy. In pregnancies, such a uterus causes various complications across the trimesters. In the early trimester, there can be recurrent miscarriages while in the later part there can be preterm labour and abnormal presentation with resultant increased incidence of caesarean delivery [4, 5]. other pregnancy related problems will include cervical incompetence due to congenital anatomical weakness of internal os resulting in pregnancy accommodation problems [6, 7]. Successful pregnancies have been severally associated with didelphys uteri and this might account for the said rarity. We present a case of didelphys uterus found incidentally during abdominal hysterectomy in a multiparous woman with endometrial carcinoma.

2. Case

A 62year old with 2 alive babies and 3 previous miscarriages, 8years post-menopausal presented to our Gynaecology Oncology service following a referral from a secondary health facility where she had presented after no appreciable improvement in her complaints of post-menopausal bleeding per vagina and increasing

associated abdominal swelling of seven- and four-months duration, respectively. She has been to several private facilities with the same complaints prior to presentation to this source of referral. There were associated weight loss, easy fatiguability, loss of appetite and dull aching, constant low back pain. Following evaluation, she had endometrial biopsy done at the source of referral and histology report of the specimen showed endometrial cancer grade 2 on account of which she was referred to our service.

The patient had previously 2 caesarean deliveries. The first caesarean delivery was on account of breech presentation while the second was on account of her previous caesarean section and her desire to have repeat caesarean delivery. These live deliveries were preceded by 3 cases of miscarriages at gestational ages of six, nine and ten weeks, respectively. She was on 3-monthly progestin injectable contraception for about 4 years prior to her attaining menopause. Her cervical cancer screening history showed that she had 5 cervical cytology tests between age 34 and 41 years with the last cytology test done 3 months prior to onset of her present complaints. All the cytology tests were reported normal. She was diagnosed hypertensive 6 years earlier and had been well controlled on medications. Clinical examination showed an obese (BMI = 34.2 kg/m^2) with no other abnormality from physical examinations. The vaginal examination showed single vagina and single ectocervix flushed with the fornices. The uterus was about 14 weeks sized with free adnexae and Pouch of Douglas. Abdominopelvic ultrasound indicated an enlarged uterus with multiple fibroid nodules. There was no other radio-imaging report available for review. The diagnosis of endometrial cancer was made on account of which she was counselled for surgical intervention.

She had exploratory laparotomy performed on the 17th of November 2021. At surgery, two uteri with separate endocervical canal but single ectocervix were found and contained multiple fibroid nodules with a large subserous component on the cornua region of the right uterus (Figure 1). The bladder, intestine, colon, peritoneal surfaces, omentum, liver and undersurface of the diaphragm were grossly free of tumour seedlings. A total abdominal hysterectomy, bilateral salpingo-oophorectomy and infra-colic omentectomy were performed. Pelvic and para-aortic lymph nodes were palpated, and no enlarged lymph node felt. The surgical specimens were sent for histological assessment. Section of the right endometrium showed infiltrative malignant glands. The left, smaller uterus showed no malignant cells on the cut section. The right ovary, fallopian tube, cervix and the omental tissue showed tumour cells. Immunohistochemical staining showed negative receptors for oestrogen and progestogen and HER2 positive of 85%. Final histological diagnosis was well differentiated endometriod variant of endometrial carcinoma involving the right uteri. The tumor was classified as at least stage IIIC according to FIGO (International Federation of Gynecology and Obstetrics) 2009. The patient recovered satisfactorily following surgery and was referred to Radiation and Clinical Oncology unit for adjuvant treatment.

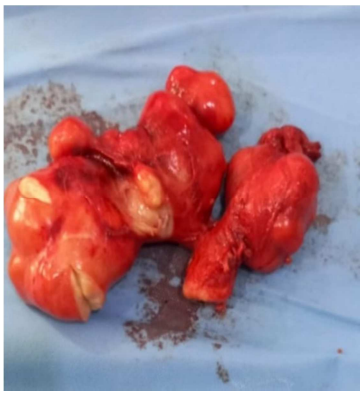

Figure 1. *Hysterectomy specimen of the case showing didelphys uterus with single distal cervix. Both uteri containing fibroid nodules with a huge subserous fibroid nodule attached to the right side of the right uterus.*

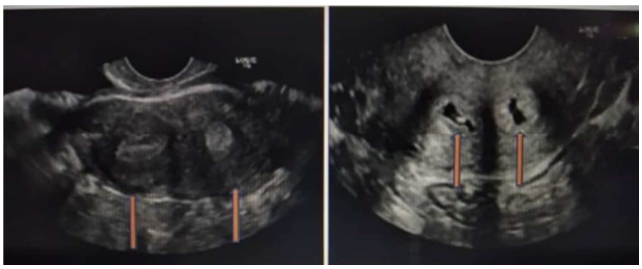

Figure 2. *Ultrasound image showing didelphys uterus (a) with two endocervices (b) [7].*

3. Discussion

Müllerian duct anomalies (MDA) are various forms of congenital defects that occurs from non-fusion of the Müllerian ducts during embryologic development. The incidence ranges between 0.5-5% of the general population. [1, 3]. Didelphys uterus is MDA class III from the modified version of the initial classification system by American Fertility Society [8]. It accounts for approximately 5% of all cases of MDAs [2]. The condition is, though considered rare, can still be associated with successful pregnancies resulting in live babies like in this case reported. Most women with a didelphys uterus are asymptomatic with better fertility potentials than those with other Mullerian duct abnormalities like septate or bicornuate uterus which are more common amongst the MDAs [3, 9]. Common symptoms in uterine didelphys include increased risk of spontaneous abortion. Our patient had 3 miscarriages before she was able to carry to term two pregnancies. While didelphys uterus is not an indication for caesarean delivery [3] especially when not associated with vaginal septum like in our patient. The indications for our patient caesarean deliveries can be considered social due to an emotional sentiment usually associated with reproductive experiences like repeated miscarriages in this case [10, 11].

Endometrial carcinoma is a common gynecological cancer. However, co-existence of uterus didelphys and endometrial carcinoma in one patient is rare. There are only few reports of endometrial cancer arising in patients with uterine malformations. Using multiple keywords in different combinations on the available databases between January

1990 and May 2020, Rockson, O, et al found only 27 cases of endometrial cancers arising within MDA reported. Our patient is one of the very few cases of endometrial cancer occurring in a didelphys uterus in our environment. Most patients with uterus didelphys had endometrial cancer on one side of the uterus, with high possibility of missing the diagnosis during evaluation. This patient presented had the cancer in the right uterus while the left uterus was free of cancer with ultrasound done as part of the pre-surgery evaluation missing the uterine didelphys. Ultrasound is the preferred investigation for detecting uterine defects, this ability is dependent on the type and techniques of the ultrasonography. Studies have shown that three-dimensional (3-D) ultrasound and when performed through the transvaginal route (TVS) give the best results [12, 13]. Our patient had transabdominal ultrasound using a 2-D ultrasound which the commonly available type of ultrasound in our setting. Other possible contributing factors for the ultrasound missing the didelphys uterus in this patient will include the obesity and the associated multiple fibroid nodules in the uteri especially with the subserosal components (Figure 1). The commonly find pictures from ultrasound are the two uteri with associated separate endocervices (Figure 2). These were found during the histological diagnosis of the surgical specimen from the case we are reporting.

In the past, the commonest method of investigation for endometrial cancer was diagnostic dilatation and curettage (D&C). Recent trends favour minimally invasive procedures like endometrial biopsy using pipelle and hysteroscopy-guided biopsy as the gold standard with estimated sensitivity of 82.6% and the specificity of 99.7% [14]. Our patient had blind endometrial biopsy, a procedure associated with delay diagnosis as high as 32% of cases. This might be the case in this patient as she could have had failed endometrial biopsies in the several private facilities she presented to before the source of referral to our facility and the eventual pT3 stage.

The standard treatment for endometrial cancer is hysterectomy and bilateral salpingo-oophorectomy, pelvic lymphadenectomy and other biopsy as appropriate [15]. Our patient did not have lymphadenectomy because because we considered it not necessary based on pre-surgery endometrial biopsy results of grade 2 lesion and gross absence of tumour seedlings on the peritoneal surfaces. While the role of lymphadenectomy remains controversial in the surgical extent in the management of endometrial cancer, specifically those adjudged confined to the uterus, many patients with such low-grade lesions based on preoperative and intraoperative assessments have higher grade disease on final pathology [16] like the case of this patient we are reporting. Lymphadenectomy, also, assist in tailoring adjuvant therapy for those with adverse risk factors. Our patient had stage IIIC endometrioid endometrial carcinoma and she is currently on adjuvant chemoradiation therapy in our Radiation and Clinical Oncology unit.

4. Conclusion

In conclusion, uterine didelphys is a rare uterine anomaly and may go undetected as it is usually asymptomatic with good reproductive outcomes. The finding of uterine didelphys may be incidental during surgery for other gynaecological conditions like uterine cancer in this case report. While the risk factors and epidemiological characteristics of such endometrial cancer might not differ from that of normal uteri, histological diagnosis can be delayed because of high rate of failed endometrial biopsy of the appropriate uterus especially when such malignant lesion involve only one of the uteri as in this case. When done by experienced hand, endometrial biopsy which is still one of the first line procedure for histological specimen collection, can yield the desire result as seen in this case. Alternatively, the use of advanced radio-imaging techniques like contrast-enhanced magnetic resonance imaging of the pelvis serves useful roles in overcoming such pre-surgery diagnostic dilemma. But this imaging technique remains scarce and financially prohibitive to many patients in our environment. Active exploratory laparotomy in cases of highly suspected endometrial carcinoma in the presence of uterine malformation even if the histological report is negative from examination of endometrial biopsy to avoid a delay in treatment might be the only option.

Contributors

Olutosin Alaba Awolude contributed to case summary, literature review, drafted the manuscript and approved the final version.

Olayinka Raphael Kuboye and Sunday Oladimeji Oyerinde contributed to literature review, reviewed all drafts of the manuscript, and approved the final version.

Patient Consent

Obtained from the patient.

Conflict of Interest Statement

The authors declare that they have no conflict of interest regarding the publication of this case report.

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