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AN ABERRANT UTERUS: CASE REPORT

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SUMMARY

A case of an aberrant uterus is presented and literature reviewed. The patient presented with abnormal uterine bleeding, left iliac fossa pain and was managed by excising the aberrant uterus. This case was an enigma as it didn't present in the classical way one with anomalies of the uterus would present. Despite knowledge on the classical types of uterine anomalies, awareness of other possibilities is important.

INTRODUCTION

In women, the tubes, the uterus and the upper portion of the vagina are formed from fusion of the paired müllerian ducts. Failure of development or fusion of the ducts gives rise to a variety of genital tract anomalies (1).

According to the American Fertility Society (AFS), classification of müllerian anomalies (1988) is as follows:

Class I: Müllerian Agenesis/Hypoplasia – in this, agenesis and hypoplasia may involve the vagina, cervix, fundus, fallopian tubes, or any combination of these structures.

Class II: Unicornuate uterus with or without a rudimentary horn – there is failure of development of one müllerian duct. When an associated horn is present, this class is sub-divided into communicating (continuity with the main uterine cavity is evident) and non-communicating (no continuity with the main uterine cavity). The non-communicating type is further sub-divided on the basis of whether an endometrial cavity, with functional endometrium, is present in the rudimentary horn.

Class III: Didelphys uterus – there is complete or partial duplication of the vagina, cervix, and uterus.

Class IV: Bicornuate uterus – Complete bicornuate uterus is characterised by a uterine septum that extends from the fundus to the cervical os. The partial bicornuate uterus demonstrates a septum, which is located at the fundus. In both variants, the vagina and cervix each have a single chamber.

Class V: Septate uterus – in this, a complete or partial midline septum is present within a single uterus.

Class VI: Arcuate uterus – A small septate indentation is present at the fundus.

Class VII: Diethyl stilboestrol (DES) related abnormality – is due to DES exposure during intrauterine life. A T-shaped uterine cavity with or without dilated horns is evident (2,3).

In the prepubertal period, normal external genitalia and age-appropriate developmental milestones often mask abnormalities of the internal reproductive organs. After the onset of puberty, young women often present to the gynaecologist with menstrual disorders, for example menorrhagia, dysmenorrhoea and cryptomenorrhoea. Late gynaecological clinical manifestations include infertility and dyspareunia (4,5).

Expected obstetrical complications include mid trimester abortions, cornual pregnancy, malpresentation, preterm labour, obstructed labour, retained placenta and post-partum haemorrhage where the placenta is implanted over the uterine septum (6).

Ultrasonography (three dimensional), Hysterosalpingography (HSG) and Magnetic Resonance Imaging (MRI) form the mainstay of diagnostic imaging. In young unmarried patients in whom HSG is best avoided, laparoscopy is performed instead (7,8).

This case is presented to highlight other possibilities of uterine anomalies.

CASE REPORT

Miss B.M., a 23 year old nulliparous lady who presented with complaints of severe left iliac fossa pain for four days necessitating use of analgesics. One week prior to this, she had experienced per vaginal spotting. Her last normal menstrual period had been

eight weeks prior. In the last cycle she had been seen with complaints of post coital bleeding and on off per vaginal bleeding, treated with progesterones and this resolved.

On examination she had marked left iliac fossa tenderness and some suprapubic and right iliac fossa tenderness.

Investigations done – urine PDT was negative. Pelvic ultrasound showed a solid hypoechoic, hypervascular mass measuring 3.6cm by 3.43cm with an impression of solid left ovarian mass most probably an ovarian fibroma or thecoma. Tumour markers done – CA-125 was elevated at 68.3 u/ml

(N: <35u/ml), Carcino-embryonic antigen was 1ug/l (N: <4ug/l) and Alpha-feto proteins were 4.9 ng/ml (N: <10.9 ng/ml). In the hormonal profile, the results were as follows – Estradiol was elevated at >3000pg/ml and progesterone was normal at 0.79 nmol/l.

Impression going into surgery was an ovarian thecoma. Laparotomy was done and the intra-operative findings were as follows – a tubal mass linked to the uterus by a fibrous conduit 7mm long and 5mm in diameter. Uterus was normal. The right ovary was healthy with the left ovary smaller than the expected size. No endometriosis was seen in the pelvic cavity.

Figure 1

A photo of the unicornuate uterus with the rudimentary horn

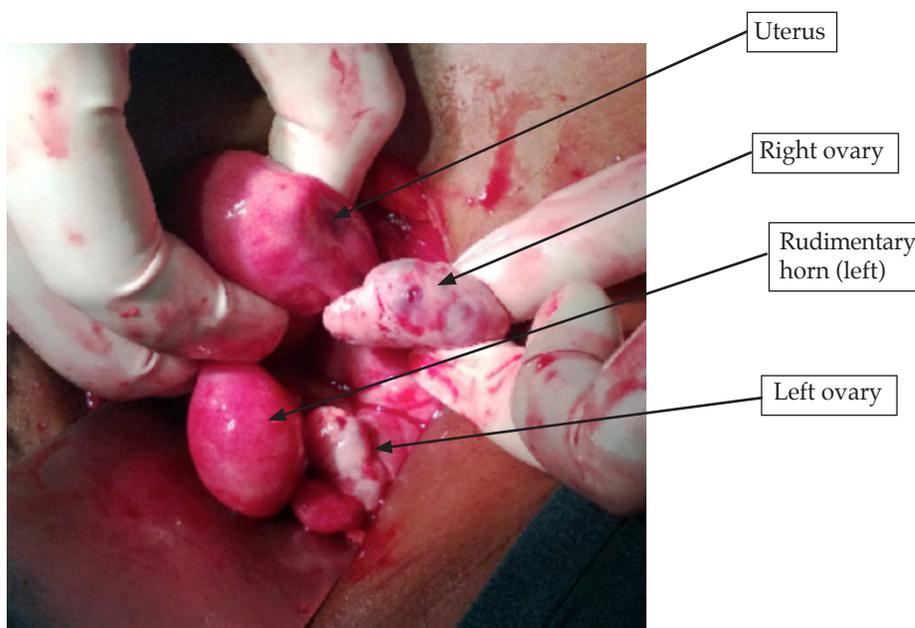
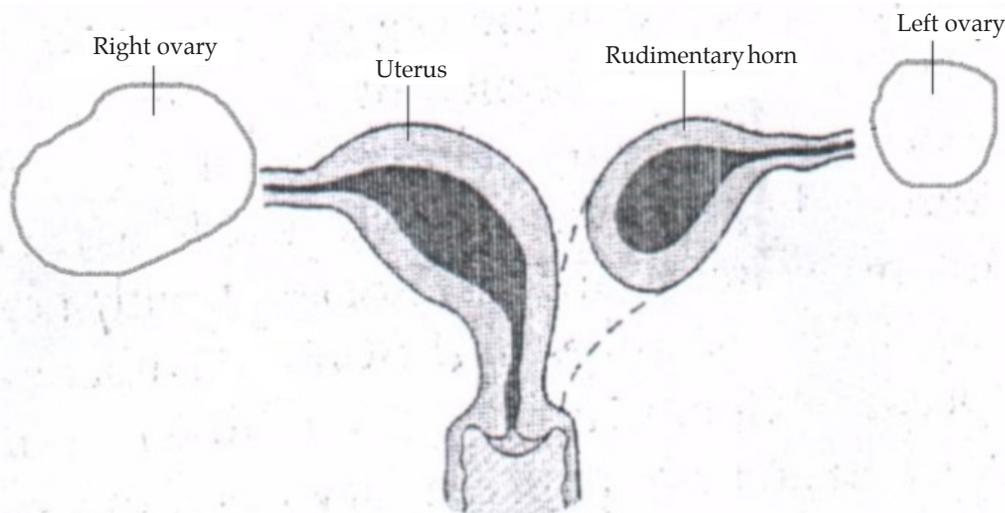


Figure 2

A drawing showing the relationship of the unicornuate uterus with the rudimentary horn

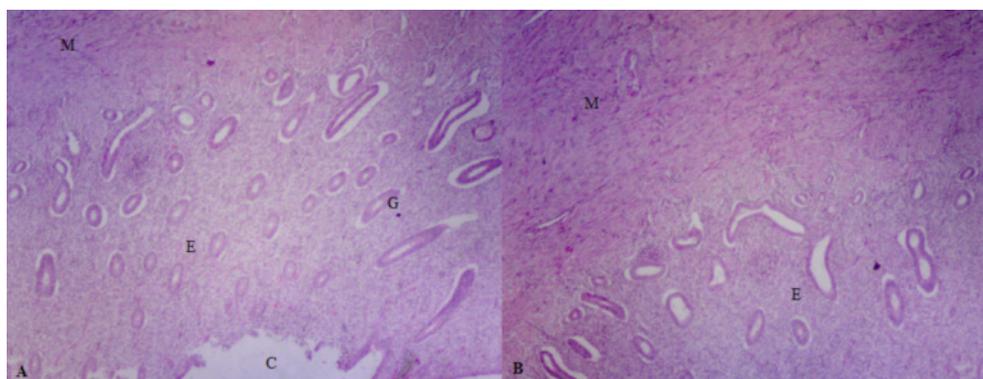


Left salpingectomy was performed and mass taken for histology. Due to the mass being tubal, a serum BhCG was performed but the result was <5 miu/ml ruling out an ectopic pregnancy. Histology results showed

myometrium, serosa and active proliferative phase endometrium confirming uterine wall tissue. This was done at a reputable laboratory and collaborated by another reputable laboratory.

Figure 3

*Photomicrographs showing functional endometrium E, with glands G and surrounded by smooth muscle cells M, resembling myometrium. A cavity C is seen at the bottom in A. Magnification used is *40*



One year later patient remained asymptomatic. She was getting regular menses lasting for three days every 28 days. Pelvic ultrasound was normal, with no residual or recurrent mass seen, uterus normal in size with normal endometrial thickness 5.6 mm, ovaries normal in size with the right measuring 3.3 by 1.7 cm and the left 2.3 by 1.7 cm and the adnexae were clear. CA-125 was 31U/ml and Estradiol levels 511 pmol/L, both within normal limits.

DISCUSSION

While minor congenital uterine abnormalities escape attention, it is the moderate or severe forms which produce gynaecologic or obstetric problems (6). These congenital uterine anomalies may be more common than generally recognised. In a study by Nahum GG, he sought to establish the prevalence and distribution of uterine anomalies among the general population. Uterine anomalies were identified in 1 in 594 fertile women (0.17%) and 1 in 29 infertile women (3.5%). The prevalence of uterine anomalies in the general population was 1 in 201 women (0.5%). Their distribution was: 7% arcuate, 34% septate, 39% bicornuate, 11% didelphic, 5% unicornuate and 4% hypoplastic/aplastic/solid and other forms (9).

Heinonen P.K did a retrospective study on 42 women with a unicornuate uterus with or without a rudimentary horn. He found that a right unicornuate uterus with a non-communicating rudimentary horn was the most common type of uterine anomaly. Thirty four women produced 93 pregnancies; ectopic pregnancy occurred in 20 (22%) of the cases. The pregnant uterine horn ruptured in three of the seven cases. The foetal survival rate was 61%, prematurity

17%, foetal growth retardation 5%, and spontaneous intrauterine abortion rate was 16%. The high number of ectopic pregnancy indicates need for removal of the rudimentary horn and its tube when diagnosed (10). Our patient had a unicornuate uterus with a rudimentary horn and in this regard had a timely excision. The prognosis of intrauterine pregnancy is not impaired in the unicornuate uterus although prematurity threatens (10).

Women with non-communicating, functioning rudimentary horns may present with pelvic pain usually secondary to hematometra or endometriosis. In a study by Markham and Waterhouse it is stated that when the rudimentary horn becomes obstructed, hematometra can develop. There is also an increased risk of developing endometriosis, which usually resolves after excision of the horn, provided an early diagnosis is rendered (11). Our patient could have experienced blockage in the fallopian tube connecting to the rudimentary horn hence the sudden onset severe pelvic pain. There was no evidence of endometriosis in this patient.

A patient with a rudimentary uterus may have symptoms and physical findings that may suggest other gynaecological problems such as ovarian tumours, uterine myomata, missed abortion, abdominal pregnancy, ectopic pregnancy, pelvic inflammatory disease, and other neoplastic disease (1). As seen in our patient, she was diagnosed pre-operatively as having an ovarian tumour. The elevated CA-125 and Estradiol levels may have been aberrant.

If implantation of a pregnancy was to occur in a uterine component that does not communicate with the vagina, non operative delivery is impossible and the likelihood of rupture very high (1,9). Our patient

was nulliparous. But there is a real possibility that on trying to conceive, the conception could have occurred in the rudimentary uterine tissue.

A prophylactic cerclage, a simple and effective treatment, should be considered if the patient experiences recurrent mid trimester abortions (12). This should also be considered for our patient if she happens to experience that or other pregnancy complications, such as preterm labour, in future.

Right unicornuate uterus with a left rudimentary horn though uncommon, should be highly suspected in adolescent girls and young women who present with severe dysmenorrhoea, sudden onset pelvic pain or abdominal swelling; and upon diagnosis, it is important to have the rudimentary horn excised to avoid future complications.

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