Complete Bicornuate Uterus with Complete Longitudinal Vaginal Septum

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ABSTRACT

Bicornuate Uterus is a type of lateral fusion disorder of the mullerian ducts. According to the American Fertility Society Classification of Mullerian Anomalies, bicornuate uterus is a class IV anomaly. Incidence of this anomaly varies. This may affect a woman's obstetric, as well as her gynecologic outcome. Here we present a 23 year primigravida at 38 weeks and 5 days gestation with footling breech presentation. She was identified, during cesarean section, of having complete bicornuate uterus with complete longitudinal vaginal septum.

Keywords: *bicornuate, pregnancy, uterus, vaginal septum*

INTRODUCTION

Congenital anomalies of the uterus are often asymptomatic and therefore unrecognized. They occur in 2-4% of fertile women with normal reproductive outcomes.[1] The bicornuate uterus is one of the symmetric-unobstructed disorders of lateral fusion of the Mullerian Ducts with 26% frequency.[2-3] It refers to a uterus with indented fundus (arbitrarily defined as ≥1cm) and generally normal vagina but a longitudinal vaginal septum may be associated.[4] It may be complete (with two cervices) and partial (with one cervix).[3] Pregnancy outcomes are close to those of the general population. However, complications such as pregnancy loss, preterm labor, or malpresentations may occur.[5]

CASE REPORT

A 23 year primigravida at 38 weeks gestation came with per vaginal leaking for 18 hours and pain abdomen for 10 hours. She had antenatal checkups elsewhere. She had ultrasonography done twice during her antenatal visits but no abnormality of the genital tract was detected. She was married for two years and had not used any contraception. Her menstrual history was regular with dysmenorrhoea and without menorrhagia. She did not give the history of dyspareunia.

On per abdomen and vaginal examinations, footling breech presentation was diagnosed and emergency cesarean section was planned. After all basic investigations, cesarean was performed.

Fetus was found to be in double footling presentation during the surgery. After exteriorizing the uterus, an indentation over the fundus and two endometrial cavities with a complete septum was identified. Right horn was the one containing the fetus and the placenta. The uterine septum was excised. Per speculum and per vaginal examinations after operation revealed two cervices and a complete vaginal septum.

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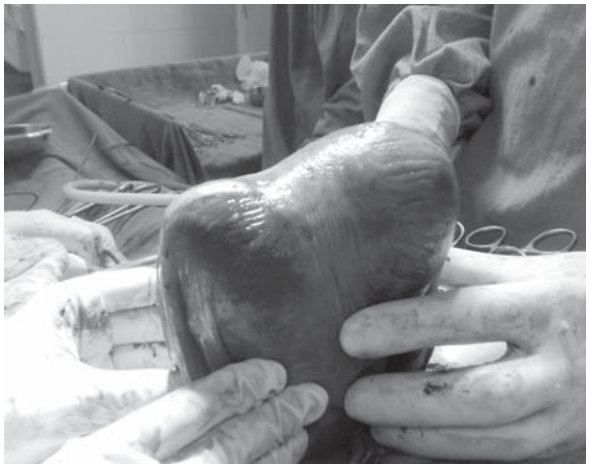

Figure 1. Indentation over the fundus of the uterus

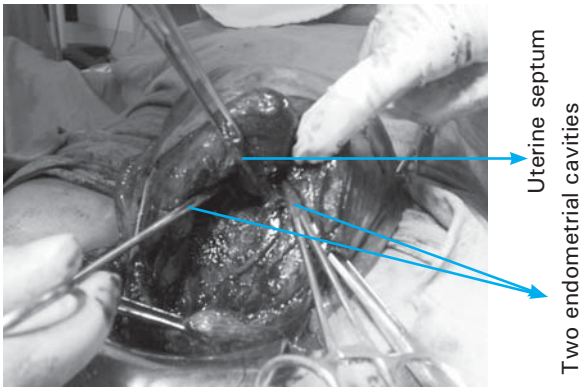

Figure 2. Two endometrial cavities divided by a septum

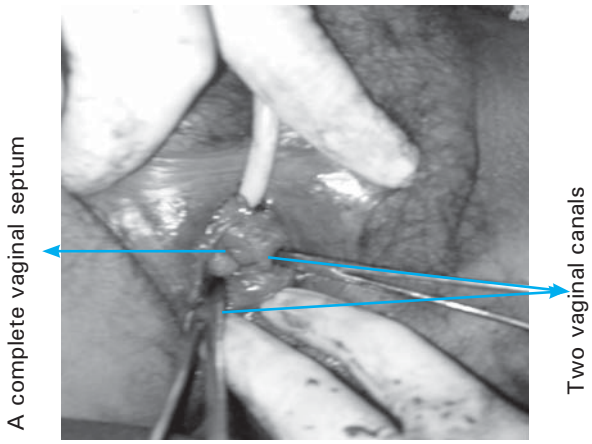

Figure 3. A complete vaginal septum with two vaginas

DISCUSSION

The bicornuate uterus and vaginal septum are congenital defects of the female genital tract. Although some uterine anomalies can cause infertility, most patients are able to conceive without difficulty. However the incidences of spontaneous abortion, premature birth, fetal loss, malpresentation, and cesarean section are clearly increased when a uterine anomaly is present. It is impossible to predict which patients with uterine anomalies will have these problems.[3]

The etiology of reproductive failure in patients with uterine anomalies remains unclear. However, the implantation of the placenta in inadequately vascularized septum, associated cervical incompetence, luteal phase insufficiency, and distortion of the uterine milieu have all been implicated in the etiology of increased reproductive loss. Interestingly, it has been reported that the chance for a live born child increases with each pregnancy loss.[3]

The absence of mullerian-inhibiting factor results in persistence of mullerian ducts in the female. These ducts grow caudally and along with the mesonephric ducts it is enclosed in the peritoneal folds that later give rise to the broad ligaments of the uterus. At about 10 weeks' gestation, the two distal portions of the mullerian ducts approach each other in the midline and begin to fuse even before they reach the urogenital sinus. The fused ducts form a tube with a single lumen called the uterovaginal canal, which then inserts into the urogenital sinus at Mullerian tubercle. This canal forms the uterus and upper portion of the vagina. In a normal female, the uterine corpus and cervix differentiate, and the uterine wall thickens by 12 weeks' gestation. Initially, the upper pole of the uterus contains a thick midline septum that undergoes dissolution to create the uterine cavity which is usually completed by 20 weeks. The unfused cephalad portions of the mullerian ducts become the fallopian tubes. Any failure of lateral fusion of the two mullerian ducts or failure to reabsorb the septum between them results in separate uterine horns or some degree of persistent midline uterine septum. Moreover, vaginal agenesis is caused by failed caudal migration of these ducts. The distal third of the vagina develops from the bilateral sinovaginal bulbs, which arise from the urogenital sinus. The most inferior portion of the uterovaginal canal becomes occluded by a cellular mass derived from the sinovaginal bulbs, termed the vaginal plate. The cells of the vaginal plate desquamate during the second trimester, allowing for full canalization of the vaginal lumen. Defects in vertical fusion caused by incomplete canalization of this plate can lead to a persistent transverse vaginal septum. Septa can develop at different levels within the vagina and can be of various thicknesses.[6]

A bicornuate uterus is caused by incomplete lateral fusion of the mullerian ducts. It is characterized by two separate but communicating endometrial cavities. Failure of fusion may extend to the cervix, resulting in a

complete bicornuate uterus, or may be partial, causing a milder abnormality. Several studies suggest that women with a bicornuate uterus can expect reasonable success in delivering a living child (about 60%). Rock reported that a bicornuate uterus was present in 55% of women with an anomalous uterus who had a satisfactory reproductive history.[7]Only 14% of women with poor reproductive performance had a bicornuate uterus. As with many uterine anomalies, premature delivery is a substantial obstetric risk. Heinonen and colleagues reported a 28% abortion rate and a 20% incidence of premature labor in women with a partial bicornuate uterus. Women with a complete bicornuate uterus had a 66% incidence of preterm delivery and a lower fetal survival rate.[8]

Early pregnancy loss is significantly more common with a septate than with a bicornuate uterus.[9] Buttram and Gibbons noted pregnancy loss rates in the first 20 weeks of 70% for bicornuate uterus compared with 88% for septate uterus.[10]

A Y-shaped uterus on hysterosalpingography may represent either a uterine septum or bicornuate uterus. In these cases, the external contour of the uterine fundus must be evaluated using MR imaging, high resolution sonography, or laparoscopy. A smooth fundal contour is consistent with a diagnosis of uterine septum.

The differentiation between a bicornuate and septate uterus is less confidently achieved by traditional transvaginalsonographic techniques. Ideally, measurement of the angle between the two endometrial cavities and analysis of the fundal shape helps to differentiate between a bicornuate uterus (angle 105°) and a septate uterus (angle 75°).[11] Combining TVS findings with SIS provides accuracy up to 90% to distinguish the two anomalies. Three-dimensional sonography is considered by some to be the best noninvasive method for distinguishing between them as its sensitivity is 93% and specificity of 100% in experienced hands.[4,12] The technique allows improved delineation of the external uterine contour and uterine volume.

MR imaging is superior in differentiating septate and bicornuate uterus. In a bicornuate uterus, the dividing septum is composed of myometrium, and with MR imaging it is characterized by signal intensity of myometrium. The endometrium of a bicornuate uterus has a normal width and lines two uterine cavities that communicate, as demonstrated by their confluent increased signal intensity. The contour of the fundus is concave or flattened. Finally, the bicornuate uterus typically has a significant notch larger than 1cm in the fundus between the two horns, and the intercornual diameter is greater than 4cm.[13-5] Bicornuate uterus may be associated with urinary tract abnormalities.

Surgical reconstruction of the bicornuate uterus has been advocated in women with multiple spontaneous abortions and in whom no other causative factors are identified. Strassman described the surgical technique that was designed to unify equal-sized endometrial cavities.[16] Reproductive outcome after unification generally has been good. In 289 women, preoperative pregnancy loss was more than 70%. Following surgery, more than 85% of pregnancies ended in delivery of a viable infant. The actual benefit of metroplasty for a bicornuate uterus, however, has not been tested in a controlled clinical series.

Even in the era of operative hysteroscopy, transabdominal metroplasty remains the only approach in cases of bicornuate uterus.[17] Cesarean delivery is indicated following metroplasty to avert uterine rupture during labor. However, in Strassmann's series of 7,161 cases delivered vaginally there was no cases of uterine rupture during pregnancy or delivery. Despite the evidence, elective cesarean section in all patients who have undergone metroplasty is recommended.

A non-obstructed vaginal septum can be managed conservatively unless dyspareunia develops. Surgical treatment includes resection.[6]

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