Successful pregnancy outcome in an untreated case of concomitant transverse complete vaginal septum with unicornuate uterus

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ABSTRACT

Transverse vaginal septum is a result of faulty canalization of embryonic vagina. Septum may be complete but usually has laterally placed tiny hole giving an impression of vaginal vault without cervix. We described a case of untreated transverse vaginal septum with small central aperture diagnosed during labor and unicornuate uterus diagnosed intraoperatively, with successful pregnancy outcome.

KEY WORDS: Cesarean section, mullerian ducts, pregnancy, unicornuate uterus

INTRODUCTION

Incidence of transverse vaginal septum varies from l: 21000 to 1:72000 and may be present in lower, middle, and upper thirds in 19%, 35%, and 46% of patients, respectively.[1] Vertical fusion/canalization disorder of mullerian ducts and urogenital sinus result in partial/complete vaginal septum of various thickness (0.5–6 cm).[2] Septum may be complete but usually has laterally placed tiny hole giving an impression of vaginal vault without cervix. Concomitant presence of unicornuate uterus is very rare in term pregnant woman. Till date, only one case of concomitant transverse vaginal septum with unicornuate uterus has been reported in young non-pregnant girl.[3]

CASE REPORT

A 24-year-old primigravida at 38 + 5 weeks came to labor room in emergency hours with chief complaints of pain in abdomen since morning. She was an unbooked patient with no investigations except for her ultrasonography done at 26 weeks of gestation which revealed no abnormalities. She was married since 11 months. Her menstrual history was regular with minimal flow lasting for 2–3 days. On general examination, she had masculine appearance with acne over face and back. There was no pallor, icterus, cyanosis, or edema. On per abdominal examination, uterine height was term with breech presentation. Liquor was less than period of gestation, uterus irritable, fetal heart sound was regular (144/min). On local examination, labia majora and minora were well-formed with abundant pubic hair. On per speculum examination, vagina was blind pouch with 2 mm opening present in centre [Figure 1]. Blood-stained mucoid discharge was seen coming out of that minute opening. Urethral opening was normal. On per vaginal examination, vagina was blind-ended pouch; but behind that small opening, a fibrous ring-like structure felt which was around two fingers loose and through which bag of members were also felt. So diagnosis of transverse vaginal septum was made and as patient was in labor with breech presentation, she was shifted for emergency lower segment cesarean section, after all necessary investigations. Abdomen was opened by transverse incision, uterus by lower segment transverse incision. Baby extracted out as breech. The liquor was almost absent. Baby cried immediately. On exteriorizing uterus it was found that uterus was unicornuate with only one tube and ovary on left side, the right sided tube and ovary was absent [Figure 2]. Uterus and abdomen closed in layers after achieving complete hemostasis. Post-operative period was uneventful and whole abdomen and pelvic ultrasonography done on 7th day of lower (uterine) segment

cesarean section (LSCS) revealed absent right kidney and ureter [Figure 3]. Mother and baby were discharged in good condition and advised to come for follow-up after 6 weeks for septum resection.

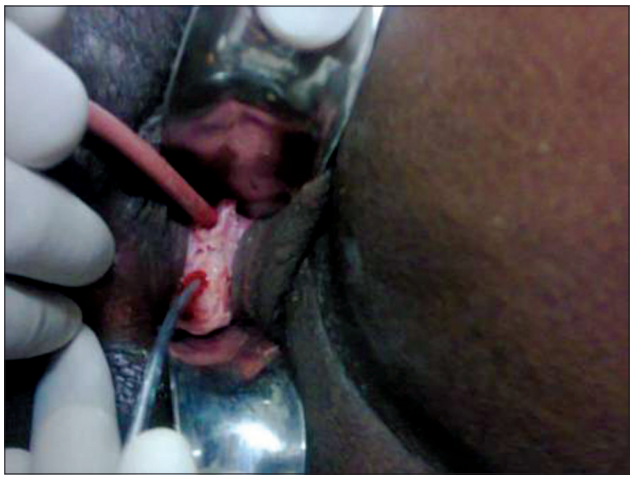

Figure 1: Showing normal uretheral opening with red rubber catheter and a small aperture in the transverse vaginal septum with a small dilator gently introduced in it

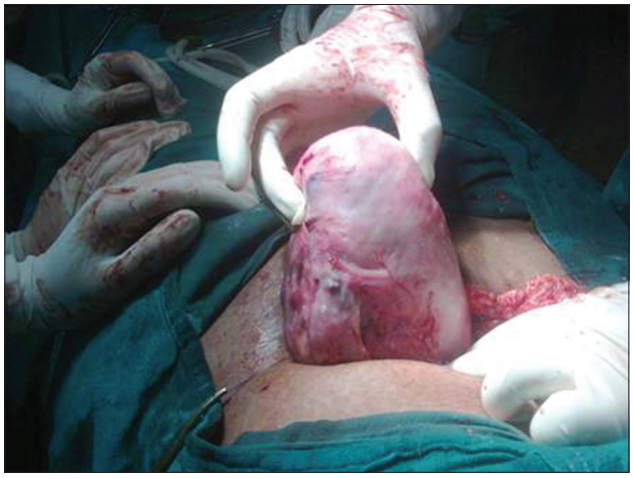

Figure 2: Showing unicornuate uterus with single ovary and fallopian tube on left side with absent fallopian tube and ovary on right side

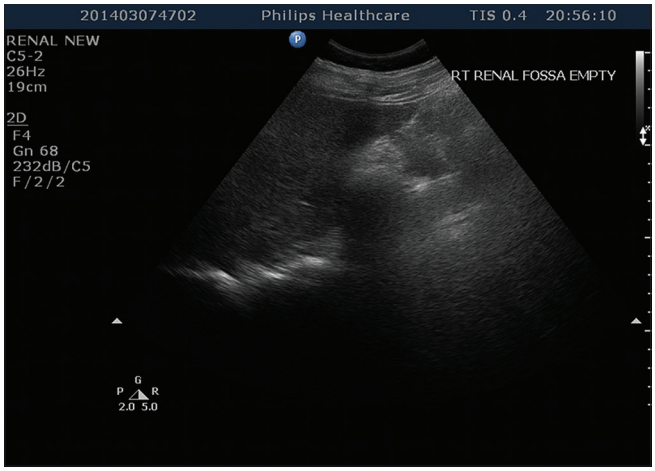

Figure 3: Empty renal fossa with absent kidney on right side

DISCUSSION

Müllerian ducts are primordial anlage of internal female reproductive organs and differentiate to form fallopian tubes, uterus, uterine cervix, and superior aspect of the vagina. Urogenital sinus gives rise to inferior and mid-vagina. When an interruption occurs in any of these dynamic processes of differentiation, migration, fusion, and canalization, wide spectrum of müllerian duct anomalies occur.

Transverse vaginal septum (TVS) is formed when tissue between vaginal plate and caudal aspect of fused müllerian ducts fails to reabsorb. This anomaly divides vagina into two segments, reducing its functional length.[4] Most of these septa are located in superior vagina at putative junction between vaginal plate and caudal aspect of uterovaginal primordium (46%).[4] It is one of most rare müllerian duct anomalies, with frequency of 1 case in 70,000 females.[5] Unlike other müllerian duct anomalies, TVS is occasionally associated with urologic defects. No evidence indicates that this disorder is genetically inherited, although a study of an inbred Amish community suggested that hydromucocolpos due to obstructive TVS was the result of a rare autosomal disorder.[5] In general, it remains undetected until time of menarche. Presentation after menarche varies depending on whether septum is complete/incomplete. If TVS is complete, patient commonly presents with primary amenorrhea and cyclic pelvic pain. Physical examination reveals palpable central lower abdominal or pelvic mass secondary to hematocolpos, hematometra, hematosalpinx, and hemoperitoneum. However, unlike an imperforate hymen, examination of genitalia reveals no evidence of bulging at introitus. Incomplete TVS allows menstrual flow to escape periodically. Diagnosis can be made on transvaginal sonography, computed tomography, and magnetic resonance imaging. Management consists of surgical correction depending on its location within vagina and its thickness. In high TVS, surgical correction is more difficult because septum is usually thick and extensive; dissection between bladder and rectum is required.[6] In low, mid, and thin TVS, septa are excised with multiple radial incisions.[4]

Unicornuate uterus results from abnormal/failed development of one of paired müllerian ducts accounting for 2.4–13% of müllerian anomalies.[7] This group of anomalies can be further subdivided into four variants according to the American Fertility Society. Isolated unicornuate uterus is most common, with frequency of 35%. When rudimentary horn is present, it is non-cavitary in 33% of cases, cavitary non-communicating in 22%, and cavitary communicating in 10%.[8] Unicornuate

uterus is associated with poorest fetal survival amongst all müllerian anomalies.[9] Cesarean delivery rates are high. Common obstetrical problems include malpresentation, intrauterine growth retardation, and preterm birth.

Anomalies of urinary tract are commonly associated and are more frequent with unicornuate uterus than other müllerian duct anomalies. Renal agenesis contralateral to main uterine horn is most common abnormality.[10]

Müllerian anomalies are diverse group of developmental disorders involving internal female reproductive tract. Establishing an accurate diagnosis is hence essential for planning treatment and management strategies.

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