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Case Report

Successful term pregnancy in an unrecognized complete septate uterus with double cervix and longitudinal vaginal septum diagnosed during caesarean delivery

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ABSTRACT

Congenital Müllerian anomalies are uncommon developmental defects of the female genital tract and may remain undetected until pregnancy. Complete uterine septum associated with double cervix and longitudinal vaginal septum is an unusual anomaly with significant obstetric implications. We report the case of a 19-year-old primigravida at 39 weeks gestation with cephalic presentation, in whom vaginal examination revealed a complete longitudinal vaginal septum with two cervices. Elective caesarean section was performed in view of the obstructive vaginal anatomy. Intraoperatively, a thick complete uterine septum extending from the uterine fundus to the vagina with two distinct cervices was identified. A healthy live baby was delivered uneventfully. This case highlights the importance of meticulous pelvic examination and awareness of rare Müllerian anomalies during antenatal care and delivery planning.

Keywords: Müllerian anomaly, Septate uterus, Double cervix, Vaginal septum, Caesarean section, Congenital uterine anomaly

INTRODUCTION

Müllerian duct anomalies occur due to abnormal fusion or resorption of the paired Müllerian ducts during embryogenesis. The reported prevalence in the general population ranges between 0.5% and 5%, although many cases remain asymptomatic and undiagnosed.[1] Septate uterus is considered the most common Müllerian anomaly and is associated with infertility, recurrent pregnancy loss, malpresentation, preterm labour, and increased caesarean delivery rates.[2]

The coexistence of complete uterine septum, double cervix, and longitudinal vaginal septum represents a rare complex anomaly that does not fit into traditional classification systems.[3] This type of rare complex mullerian malformation is under class U2b C2 V1 by ESHRE/ESGE classification. Such anomalies may remain clinically silent until gynaecological examination during pregnancy or labour.

We present a rare case of complete septate uterus with double cervix and complete longitudinal vaginal septum identified during elective caesarean section in a primigravida at term.

CASE REPORT

A 19-year-old primigravida presented at 39 weeks gestation for delivery planning. She had been married for one year and conceived spontaneously without any history of infertility or assisted conception. There was no history suggestive of dyspareunia, dysmenorrhea, chronic pelvic pain, or menstrual obstruction, although she reported

irregular menstrual cycles since menarche. The patient had not undergone first trimester dating or nuchal translucency scans because of personal reasons. She first presented to our institution at 25 weeks gestation with a TIFFA scan performed elsewhere, which was reported as normal. No uterine anomaly had been identified antenatally. This may have been due to progressive enlargement of the gravid hemi-uterus causing lateral displacement and compression of the septum, thereby obscuring the underlying anomaly on ultrasonography.

Her antenatal period remained otherwise uneventful with no episodes of threatened preterm labour or antepartum haemorrhage. Ultrasound at term demonstrated a single live fetus in cephalic presentation with adequate liquor volume and normal fetal growth parameters.

On per-vaginal examination performed prior to delivery, a complete longitudinal vaginal septum was identified dividing the vaginal canal into two compartments. Two distinct cervices were visualized separately on either side of the septum. In view of the complex vaginal anatomy and anticipated difficulty with vaginal delivery, an elective lower segment caesarean section was planned.

Intraoperatively, after opening the abdomen in layers followed by uterine incision, a live term baby boy weighing 2.8 kgs with satisfactory APGAR scores was delivered uneventfully. The placenta was noted to be attached predominantly to the fundal and posterior uterine wall and was removed completely.

Inspection of the uterine cavity following placental delivery demonstrated a thick complete septum measuring approximately 1.5 cm in thickness. Uterus exteriorised and through inspection of outer surface and inner cavity was done. The septum appeared muscular and vascular, extending from the uterine fundus through the endometrial cavity into two separate cervical canals and continuing inferiorly as a complete longitudinal vaginal septum. No communication was noted between the two uterine cavities. Both adnexa appeared normal. The intraoperative pictures are shown below (Figure 1-3).

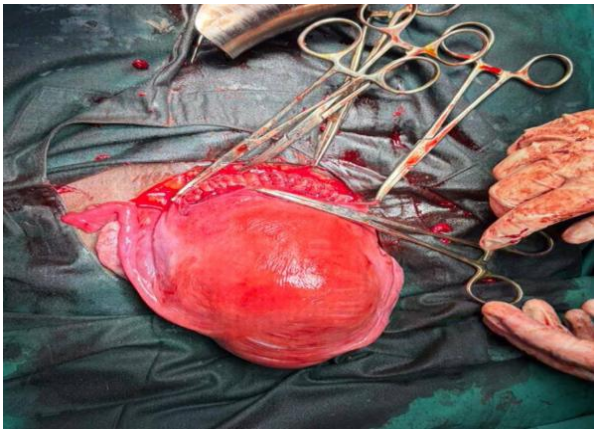

Figure 1: Intra-operative uterine shape.

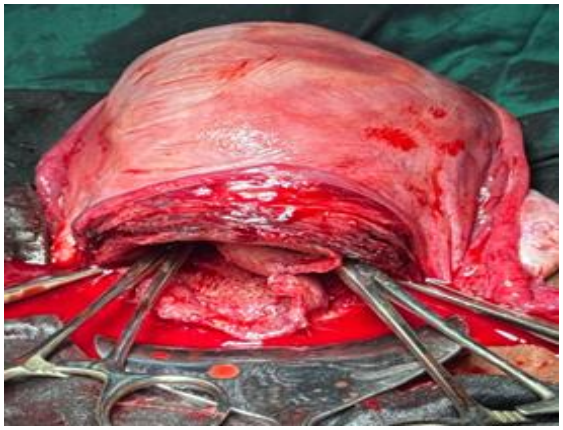

Figure 2: Uterine cavity with septum following placental delivery.

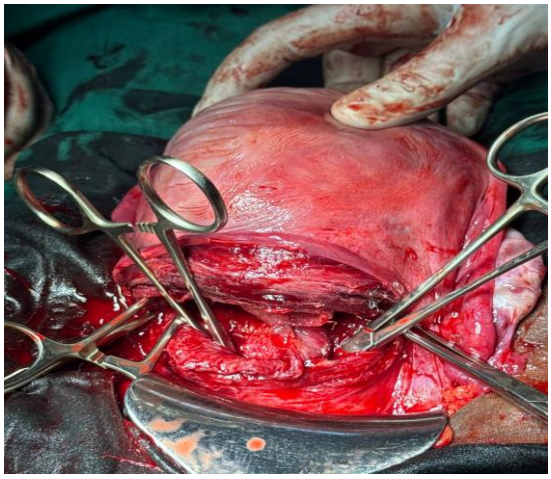

Figure 3: Intra-operative view of the septum extending into the lower uterine segment and cervical region.

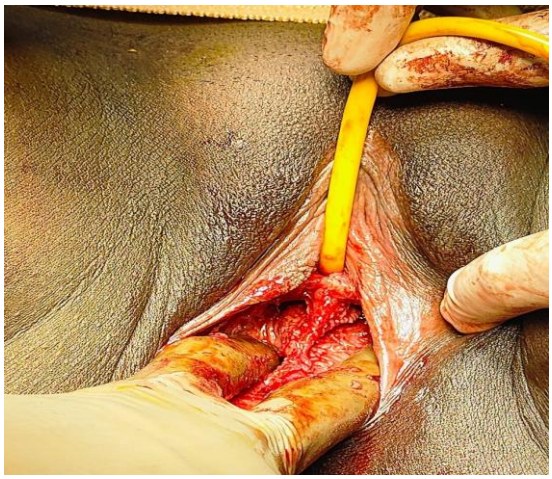

Figure 4: Local examination demonstrating complete longitudinal vaginal septum.

The postoperative period was uneventful, and the patient was discharged in stable condition with advice regarding further evaluation and follow-up.

DISCUSSION

Müllerian anomalies result from defective organogenesis, fusion, or resorption of the Müllerian ducts during fetal development.[4] Septate uterus specifically results from incomplete resorption of the midline septum after normal fusion of the Müllerian ducts.

The coexistence of a complete uterine septum with double cervix and longitudinal vaginal septum represents an unusual Müllerian anomaly that challenges the classical embryological theory of caudal-to-cranial fusion of the Müllerian ducts. According to the traditional concept proposed by Crosby and Hill, fusion of the Müllerian ducts begins caudally and progresses cranially, followed by resorption of the intervening septum.[5]

However, anomalies such as complete septate uterus with double cervix and longitudinal vaginal septum cannot be completely explained by this mechanism alone. The presence of duplicated cervices with a persistent uterine septum suggests a possible defect in both fusion and resorption processes occurring simultaneously.[6]

An alternative bidirectional fusion theory proposed by Musset describes Müllerian fusion beginning at the isthmic region and progressing simultaneously in both cranial and caudal directions. Failure of resorption at multiple levels may therefore result in complex anomalies involving the uterus, cervix, and vagina, as observed in the present case.[7]

Such anomalies are difficult to classify precisely within conventional American Society for Reproductive Medicine (ASRM) categories and highlight the spectrum and variability of Müllerian developmental defects.

The presence of double cervix and longitudinal vaginal septum associated with septate uterus is rare and has been debated embryologically because it challenges the classical unidirectional fusion theory.[5] Alternative embryological theories propose bidirectional fusion and resorption mechanisms to explain these complex anomalies.

Patients with longitudinal vaginal septum may present with dyspareunia, dysmenorrhea, infertility, or labour obstruction; however, asymptomatic cases are not uncommon.[6] In the present case, the anomaly remained undiagnosed until term pregnancy due to the absence of gynaecological symptoms.

Pregnancy in women with septate uterus carries higher risks of miscarriage, fetal malpresentation, preterm birth, intrauterine growth restriction, and operative delivery.[7] Although vaginal septum excision during labour has been described in selected cases, elective caesarean section was considered safer in our patient because of the complete septum and distorted vaginal anatomy.

Three-dimensional ultrasonography and magnetic resonance imaging are considered highly accurate modalities for diagnosing Müllerian anomalies.[8] However, antenatal imaging may occasionally fail to identify complex anomalies, especially in advanced pregnancy.

This case emphasizes the importance of careful pelvic examination in antenatal patients and the need for individualized obstetric management in women with complex Müllerian anomalies. Absence of infertility or dyspareunia despite complete longitudinal vaginal septum makes the present case clinically unusual and not necessitating the removal of septum during the surgery. The notable feature of the present case was not merely the presence of the anomaly itself, but the absence of expected reproductive and gynaecological symptoms. Despite a complete uterovaginal septum, the patient conceived spontaneously within one year of marriage, remained asymptomatic, and carried the pregnancy till term.

CONCLUSION

Complete septate uterus with double cervix and complete longitudinal vaginal septum is an uncommon Müllerian anomaly that may remain asymptomatic until pregnancy. Thorough clinical examination and intraoperative assessment are essential for accurate diagnosis and optimal obstetric management. Elective caesarean section can result in favourable maternal and fetal outcomes in such rare anomalies. It is for the obstetrician to decide whether to intervene in such cases i.e. to proceed with septal resection or not based on the patient history and further counselling.

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