

Uterine didelphys with transverse vaginal septum: A case report and review of the literature

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Abstract

PURPOSE: To report a rare case of two co-existing Mullerian anomalies.

METHODS: Case report.

RESULTS: A 13-year-old G0 female presented with a one-day history of episodic, severe pelvic pain. Her menses had ended two days prior to her arrival to the hospital and was shorter than her usual five-day cycle and had heavier associated clotting. Menarche was four months prior. Past medical history was significant for a single left sided kidney and a family history of congenital kidney disease. Her mother reported that she herself had a uterine didelphys. Breasts and pubic hair were in Tanner stage 2 development. Her external anatomy was grossly normal. Internal exam was limited due to patient discomfort. An abdominal ultrasound showed a hypoechoic cystic structure with layering echogenic debris arising from the cervix. A provisional differential diagnosis of a Mullerian anomaly or a newly obstructed cribriform hymen was given. Therefore, a pelvic MRI was ordered, and an anesthetized pelvic exam was scheduled. MRI of the pelvis revealed uterine didelphys and dilated vagina with suspected cribriform versus imperforate hymen. Pelvic examination under anesthesia revealed a perforate hymen. Within the vaginal vault, a blue bulge with a thin yellow septum was noted. To the left, a small 0.5 cm opening was palpated without a cervix. A 1.5 cm incision was made into the fluid pocket and significant hematocolpos and suspected hematometra was drained. The tissue within the incision was vaginal epithelium by illumination, consistent with a transverse vaginal septum. A possible cervix was palpated through the incision. The final diagnosis for the patient was uterine didelphys with a partially obstructing transverse vaginal septum and an ipsilateral right pelvic kidney. Postoperatively, the patient reported resolution of abdominal pain and was discharged the following day. The patient was referred to pediatric adolescent gynecology at a tertiary care center for further evaluation and follow up. She was started on contraception to prevent recurrent hematocolpos and hematometra.

CONCLUSIONS: This case demonstrates many characteristic features of Mullerian anomalies including a history of a unilateral kidney, pelvic pain shortly after menarche, and a bulging mass on pelvic exam. However, the presence of regular menses for four months prior to presentation is unusual. A transverse uterine septum is considered one of the rarest anomalies of the female genital tract. On English language PUBMED search, there are only two other reported cases comprising of both uterine didelphys and a transverse vaginal septum. There are serious potential chronic complications resulting from obstructive Mullerian anomalies, including endometriosis, infertility, infection, spontaneous abortion, and premature birth. These possible long-term implications emphasize the importance of early and accurate diagnosis and management of these conditions.