CASE REPORT

Double cervix and vagina with a normal uterus: an unusual Mullerian anomaly

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A 34 year old woman presented with primary infertility and duplication of the cervix and vagina. Laparoscopy demonstrated a normal uterus and hysterosalpingography revealed a normal uterine cavity communicating with both cervices. This rare Mullerian anomaly is inconsistent with our current understanding of Mullerian development. An alternative embryological mechanism is reviewed to account for this and other anomalies which do not fit into existing classification systems.

Key words: cervical duplication/embryology/Mullerian anomaly/vaginal septum

Introduction

A classification system exists to categorize the various types of Mullerian anomalies (American Fertility Society, 1994). We encountered a case, and reviewed others in the literature, which could not be accommodated by this system and are also incongruous with the accepted mechanism of Mullerian embryogenesis.

Case report

A 34 year old white nulligravida presented with an 8 year history of primary infertility. Two years earlier, she had undergone laparoscopic cauterization of mild endometriosis and excision of a complete longitudinal vaginal septum. The uterus, tubes, and ovaries were described as being grossly normal. An intravenous pyelogram was also normal. Her history was otherwise unremarkable and her physical examination was entirely normal with the exception of cervical duplication. Her infertility evaluation included a normal semen analysis, post-coital test, and endometrial biopsy. Hysterosalpingography was performed by injecting water-soluble contrast into the left cervical os. The uterine cavity was normal and the Fallopian tubes were patent. In addition, contrast was seen to exit the right cervical os on fluoroscopy (Figure 1). The patient was subsequently lost to follow-up.

Discussion

This case represents a unique Mullerian malformation. Two similar cases of cervical and vaginal duplication with a normal uterus have been previously described; however one cervical canal ended blindly and did not communicate with the uterus (Kletz et al., 1994; Candiani et al., 1996). These anomalies do not conform to our classical understanding of Mullerian development (Crosby and Hill, 1962). This theory holds that the Mullerian ducts cross ventrally over the mesonephric ducts and their medial walls fuse in the midline at the caudal-most aspect, known as the Mullerian tubercle. Fusion continues cranially to the junction of the round ligaments. The fused medial walls are then resorbed, leading to a unified uterine cavity. The Mullerian tubercle induces the proliferation of the adjacent urogenital sinus to form the sinovaginal bulbs. These give rise to the solid vaginal plate which later canalizes creating a patent outflow tract.

Recent reports of a septate uterus with cervical and vaginal duplication (McBean and Brumstead, 1994), a communicating septate uterus with a double cervix and normal vagina (Lev-Toaff et al., 1992), and a double cervix with a normal vagina and uterus (Tavassoli, 1977) also fall outside the current classification system of Mullerian anomalies (American Fertility Society, 1988) and suggest an alternative embryological mechanism. Musset et al. (1967) proposed that fusion
first occurs at the level of the uterine isthmus and simultaneously proceeds in both directions. Midline resorption also begins at the isthmus and is first directed caudally, unifying the cervix and vagina, and later cephalad to eliminate the uterine septum. This theory can account for the standard classifications of Mullerian anomalies in addition to the above noted exceptions. It is unknown whether it is the mechanism for normal Mullerian development or only an aberration, aetio logically unique to these most unusual anatomical variants.

References


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