CASE REPORT

Bicervical uterus and septate vagina: report of a previously undescribed Müllerian anomaly

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A case of non-septate uterus with double cervix and complete longitudinally septate vagina is described. This previously undescribed Müllerian anomaly has been detected by complete clinical and instrumental investigations (transvaginal ultrasonography, urography, hysterosalpingography, hysteroscopy and laparoscopy). Aetiological hypotheses are discussed and we suggest adding the uterovaginal malformation described in this report to the embryological classification recently proposed by Aden et al. (1991, 1992) which postulates isolated Müllerian anomalies as a consequence of a minor mesonephric defect.

Key words: congenital anomalies/Müllerian anomalies/uterine malformations

Introduction

The commonest congenital malformations of the Müllerian axis are developmental anomalies of the uterus, which frequently cause infertility. The aetiology is multifactorial, and one of the following events is involved: defective development of one or both paramesonephric ducts (which is responsible for unicornuate uterus, rudimentary horn, and uterine agenesis), failed or defective fusion (bicorne and didelphic uteri), and defects of canalization (septate uterus).

We describe a woman with bicervical non-septate uterus, longitudinal vaginal septum, and normal adnexa. To our knowledge such a malformation has not previously been described in the literature.

Case report

In 1985, a 31 year old woman with a 3-year history of primary infertility was hospitalized in a gynaecological department in North Italy. A longitudinal vaginal septum, double cervix, and apparently single uterus with bilateral palpable adnexa were observed at routine gynaecological examination, and complete ablation of the vaginal septum was performed (Figures 1 and 2).

Hysterosalpingography (HSG) through the right cervical canal showed a regular uterine cavity and bilateral tubal patency; the left cervical canal was 2 cm in length and ended blindly (Figure 3). Urography revealed the presence of bilateral megaloureter without functional alterations.

The patient underwent a curettage of the uterine cavity for post-abortion endometritis in 1987, and 2 years later she aborted spontaneously at gestational week 8.

In 1992 she was admitted to Istituto S.Raffaele, Milano, for an infertility investigation. Gynaecological examination confirmed the previous results. Transvaginal ultrasonography showed a single uterus, regular uterine cavity, and thin endometrial lining. Two external uterine orifices were seen but only the right cervical canal appeared to communicate with the uterine cavity. The ovaries were of normal size and morphology.

At laparoscopy no pelvic adhesions were seen, the adnexa and uterus were normal, and both uterine tubes were patent (Figure 4).

Hysteroscopy confirmed the presence of a regular single uterine cavity originating from the right cervical canal. The tubal orifices were visualized. A blind canal, 2 cm in length, originated from the left cervix.

The biopsy obtained from the left cervical canal confirmed the presence of unistratified columnar epithelium resting on a stroma with mucus-secreting glands.

Endocrine, microbiological and immunological investigations yielded normal results.

Discussion

The malformation described, which we have termed bicervical uterus and septate vagina, does not seem to fit into any of the
existing classifications (Jarcho, 1946; Jones, 1956; Musset et al., 1973; Buttram and Gibbons, 1985). The first gynaecological examination demonstrated a longitudinal vaginal septum, which suggested the presence of a didelphic-type malformation. This was excluded by subsequent instrumental investigations showing that the second (left) cervical canal was blind. HSG through the right cervix demonstrated that the entire corresponding Müllerian axis was normal. A single uterus and bilateral adnexa were observed at transvaginal ultrasonography and laparoscopy.

When didelphic and unicorneate bicervical uterus with rudimentary left horn were excluded, a developmental anomaly of the caudal portion of the left paramesonephric duct and Müllerian tubercle was postulated.

Recently, McBean and Brumsted (1994), describing a rare malformation, suggested that the presence of cervical duplication with a unified uterine fundus challenges the classically held view that a normal female apparatus results only when Müllerian fusion proceeds in a unidirectional, caudal or cephalad direction. Experimental studies on morphogenetic processes that induce malformations have identified four types of disturbances that can affect the evolution of the embryonic primordia: defective or hypertrophic processes, altered fusion and aberration due to movements.

However, in the case described, the development, fusion and canalization of the paramesonephric ducts were normal, resulting in a normal genital apparatus. It seems unlikely that the above mentioned events were responsible for the cervicovaginal malformation. It is probably an isolated Müllerian anomaly induced by a minor mesonephric defect. If close developmental relations exist between the Müllerian and Wolffian ducts, as postulated by Gruenwald et al. (1941), and if a deficient inductive function of a mesonephric duct can lead to a uterovaginal anomaly, it is possible that both a second isolated cervix and a longitudinal vaginal septum could derive from an abnormal effect of the left mesonephric duct on the corresponding caudal portion of the Müllerian duct and on the Müllerian tubercle could give rise to, respectively, a double cervix and a longitudinal vaginal septum.

In conclusion, we suggest adding the uterovaginal malformation described in this report to the embryologic classification recently proposed by Acien et al. (1991, 1992) which postulates isolated Müllerian anomalies as a consequence of a minor mesonephric defect.

References

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