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CASE REPORT

Laparoscopic management of a unicornuate uterus with two cavitated, non-communicating rudimentary horns

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An 18 year old nulligravid woman presented with severe dysmenorrhoea secondary to stage IV (revised American Fertility Society) endometriosis, right haematosalpinx, right endometrioma, unicornuate uterus and two cavitated, non-communicating rudimentary uterine horns. To our knowledge, this is the first reported case of a unicornuate uterus accompanied by two rudimentary horns. Operative video-laparoscopy proved a successful approach for treating this previously unreported variant of congenital Müllerian anomaly. A review of the world literature confirms the uniqueness of this case while demonstrating laparoscopy to be a viable alternative to laparotomy for management of congenital Müllerian anomalies. The case presented may help to elucidate Müllerian duct embryology further.
Key words: congenital Müllerian anomaly/laparoscopy/rudimentary horn

Introduction

The reported incidence of congenital Müllerian anomalies has ranged from 1:500 to 1:2000 in one report (Semmens, 1962) to as high as 3.2% in another (Simon et al., 1991). A partial or complete lack of development of one Müllerian duct during weeks 7–8 of gestation may result in the formation of a unicornuate uterus. In a retrospective longitudinal review of 3181 patients of reproductive age desiring to conceive, the incidence of unicornuate uterus was found to be 0.06% (Raga et al., 1997).

The functioning unicornuate uterus may or may not be associated with a rudimentary uterine horn. The rudimentary horn may be cavitated or non-cavitated, and may or may not communicate with the functioning unicornuate uterus (Falcone et al., 1997). An estimated 75–90% of unicornuate uteri with rudimentary horns are non-communicating (O'Leary and O'Leary, 1963; Buttram and Gibbons, 1979; Rock and Schlaff, 1985).

In the case of a blind non-communicating, cavitated rudimentary horn with a functioning endometrium, cryptomenorrhoea may lead to dysmenorrhoea soon after menarche and result in

haematometra (Thompson and Rock, 1992). Retrograde menstruation from a functioning rudimentary horn through a patent ipsilateral Fallopian tube may result in the development of haematosalpinx and endometriosis (Olive and Henderson, 1987). Transperitoneal migration of spermatozoa very rarely may result in a rudimentary horn pregnancy (Dicker et al., 1998). Rudimentary horn pregnancies have been associated with a 70% risk of uterine rupture occurring before 20 weeks of gestation, accompanied by potentially massive and life-threatening intraperitoneal haemorrhage (Kadir et al., 1996).

Given these potential complications, it is advisable to remove a rudimentary horn when first recognized. In recent years, laparoscopy has become a viable alternative to laparotomy for management of the rudimentary horn. To date, 16 cases of laparoscopic resection of rudimentary uterine horns have been published in the world literature (Tables I and II). Three of these cases involved laparoscopic treatment of rudimentary horn pregnancies (Table II). We report here a case of laparoscopic management of a unicornuate uterus with severe peritoneal endometriosis [revised American Fertility Society (AFS) stage IV], endometrioma, haematosalpinx and two cavitated, non-communicating rudimentary horns. To our knowledge, this represents the first reported case of a unicornuate uterus accompanied by two rudimentary horns. This variant is not described by the AFS classification of Müllerian anomalies (American Fertility Society, 1988) and may allow for some elucidation of the embryology of the Müllerian system.

Case report

An 18 year old nulligravid woman presented with severe dysmenorrhoea since menarche in May 1995 which was only minimally relieved with medical management [oral contraception and non-steroidal anti-inflammatory drugs (NSAIDs)]. In April 1998, the patient experienced an episode of severe pain in the right lower quadrant. Pelvic ultrasound by her paediatrician revealed a large irregular complex mass in the right hemi-pelvis with multiple cystic and solid components. The uterus and left ovary were thought to be normal. The right ovary could not be identified.

The patient underwent diagnostic laparoscopy performed by her primary gynaecologist in May 1998. The findings included severe endometriosis involving the left ovary, posterior cul-de-sac and the subdiaphragmatic surface. Additionally, fibrous adhesions involving the ascending colon, small intestine and right pelvic sidewall and layering of haemosiderin throughout the pelvis were described. Inspection of the right adnexa revealed

Table I. Laparoscopic resection of rudimentary horn (adapted from Amara *et al.*, 1997)

Author	Year	Age	Sex	Preop Rx	IVP	Endometriosis	Anomaly
1. Canis *et al.*	1990	30	Dysp Dysm	GnRH agonist 3 mo	Normal	Yes	AFS IIB
2. Yeko *et al.*	1992	18	Dysm	None	Normal	No	Absent vagina w/bilat Müllerian remnants
3. Nezhat *et al.*	1994	28	Dysm	Danazol 6 mo	Bilat sponge kidneys	Yes	AFS IIB
4. Mais *et al.*	1994	26	Dysm Pelvic Pain	GnRH agonist 6 mo	Normal	No	AFS IIB
5. Schattman *et al*	1995	29	Dysm Infert	None	Normal	Yes	AFS IIB
6. Amara *et al.*	1997	13	Dysm	None	Vaginal Urethra	No	AFS IIB
7. Chapron *et al.*	1995	15	1^0Amen pelvic pain	None	Bifid right ureter	Yes	Hypoplastic vagina Bilat rud. horns
8. Kadir *et al.*	1996	34	Pelvic pain	None	Single left kidney/ureter	Yes	AFS IIB
9. Falcone *et al.*	1997	23	Dysm Vaginismus	None	Not reported	Yes	AFS IIB
10. Giatras *et al.*	1997	29	Dysm Infert	None	Normal	Yes	AFS IIB
11. Giatras *et al.*	1997	25	Infert Pelvic pain	None	Single left kidney/ureter	No	AFS IIB
12. Giatras *et al.*	1998	30	Pelvic pain	None	Normal	No	Hypoplastic vagina Bilat rud horns
13. Present case	1998	18	Dysm. Pelvic pain	GnRH agonist 3 mo	Single left kidney/ureter	Yes	Unicornuate uterus with two rud. horns

Dysp = dyspareunia, Dysm = dysmenorrhoea, Infert = infertility, 1^0Amen = primary amenorrhoea, Bilat. rud. = bilateral rudimentary, Preop Rx = preoperative medication, IVP = intravenous pyelogram.

right haematosalpinx and a 7–8 cm endometrioma involving the entire right ovary which was drained. The senior author was consulted intraoperatively. On our advice, the procedure was terminated to allow further work-up prior to definitive surgery.

A subsequent i.v. urogram revealed a contoured deformity of the distal left ureter suggesting possible encroachment by endometrial tissue. An enlarged left kidney and absence of the right kidney were also observed. An abdominopelvic magnetic resonance imaging confirmed the congenital absence of the right kidney and mildly enlarged left kidney. A unicornuate uterus was seen projecting to the left aspect of the pelvis with a normal-appearing endometrial stripe. A serpentine tubular structure measuring 4.8×4.7 cm in the largest dimension seen in the right lower pelvis and thought to be haematosalpinx was observed adjacent to the unicornuate uterus. A 5.5×5.0×5.0 cm complex mass was noted at the right posteriolateral aspect of the pelvis. A 2–3 cm lesion on the right ovary suggesting an endometrioma and a normal-appearing left ovary were described.

Following 3 months of gonadotrophin-releasing hormone agonist (GnRHa) therapy, the patient underwent operative video-laparoscopy in August 1998. Examination under anaesthesia revealed a normal vulva and vagina. Hysteroscopy confirmed evidence of a unicornuate uterus projecting to the left with a left tubal ostium. No ostium was seen on the right. Next, via an intra-umbilical incision through direct trochar entry (Nezhat *et al.*, 1995), a 10 mm trocar and video-laparoscope were introduced. Pneumoperitoneum was obtained and three 5 mm trocars were introduced into the lower abdomen for the purpose of the introduction of ancillary instruments.

Table II. Laparoscopic management of rudimentary horn pregnancy

Author	Year	Age (years)	Gestational age (weeks)	Anomaly[a]
1. Dulemba *et al.*	1996	26	13 weeks	AFS IIB[a]
2. Dicker *et al.*	1998	31	8 weeks	AFS IIB
3. Yahata *et al.*	1998	22	7 weeks	AFS IIB

[a]cf American Fertility Society (1988).

Findings at laparoscopy revealed a developed left unicornuate uterus with a patent Fallopian tube and a normal left ovary. Two Müllerian remnants were seen on the right side of the pelvis. One was ~5 cm in diameter and the other was 6 cm in diameter and each contained copious amounts of 'chocolate' fluid comprising two distinct haematometra. These remnants did not communicate with the uterus or each other. A 7.0×2.0 cm right haematosalpinx, and a 4.5×3.0×2.5 cm endometrioma involving the right ovary were noted. The right ovary itself measured 5.0×3.0×2.0 cm and contained a small follicular cyst measuring 0.2 cm in maximum dimension. The right ovary did not appear to receive its blood supply from the right infundibulopelvic ligament. Its only clear vascular supply was from the utero–ovarian attachments between it and the most superior rudimentary horn. Approximately 20–30 spots of endometriosis involving the right and left hemi-diaphragms were also discovered. There was evidence of complete posterior cul-de-sac obliteration.

Due to the above mentioned anatomical attachments of tube

and ovary, it was not possible to save the right ovary. Thus, the trocar site above the bladder dome was changed to a 12 mm trocar and using an endovascular stapling device (Ethicon, Cincinnati, OH, USA) multiple bites were taken from the right infundibulopelvic ligament and mesosalpinx allowing for removal of the right ovary and right Fallopian tube (Kovac *et al.*, 1990). Next, left ureterolysis using a CO_2 laser and hydrodissection was performed (Nezhat *et al.*, 1995). Posterior cul-de-sac adhesions and bowel adhesions were lysed using a CO_2 laser. Rectovaginal examination was performed delineating a plane to dissect the rectum off the posterior cul-de-sac peritoneum, uterosacral ligaments and rudimentary horns (Nezhat *et al.*, 1992).

The retroperitoneum on the right side was entered and the most superior rudimentary horn was freed from adjacent structures and vasculature, enucleated from the right pelvic sidewall, morcellated (Karl Storz, Culver City, CA, USA) and removed. All major blood vessels feeding the rudimentary horns were identified at their origin to the right hypogastric artery desiccated and cut. This allowed for safe removal of the second, most inferior rudimentary horn. Initial morcellation of the second rudimentary horn revealed multiple small endometrial polyps contained within it. This remnant was attached to the apex of the vagina with ~3 cm distance between it and the unicornuate uterus. While this remnant was noted to extend partially into the vagina, it did not communicate with the vagina. The vaginal cuff was closed with multiple interrupted Vicryl sutures (Ethicon). Further treatment of endometriosis spots on the bladder and pelvic sidewall was performed using laser excision and vapourization (Nezhat and Nezhat, 1989). Cystoscopy was performed revealing normal flow through the left ureter. There was no right ureteral orifice or bladder endometriosis. The total operating time was 4 h and 15 min.

The patient's hospital course was uneventful and she was discharged on the first postoperative day.

Pathology findings revealed proliferative phase endometrium and normal myometrium in one horn and fragments of attenuated endometrium and myometrium with changes suggestive of endosalpingiosis in the other. The right Fallopian tube showed changes consistent with haematosalpinx. The right ovary contained a haemorrhagic cyst consistent with endometrioma and otherwise normal ovarian tissue.

Discussion

The true incidence of unicornuate uteri is not known. Current estimates are based only on those few cases which are actually diagnosed and subsequently reported in peer review journals. It stands to reason then that yet undescribed variants of this anomaly exist. It has been reported that the 1988 AFS classifications of unicornuate uteri (the most widely accepted classification) does not completely describe anatomical variations of the rudimentary horn (Falcone *et al.*, 1997). His review of the literature suggested that anatomical variations of rudimentary horns included: cavitated versus non-cavitated, communicating or non-communicating, and attached to the unicornuate uterus by a band of tissue or the rudimentary horn as part of the unicornuate uterus. There is also no classification

Figure 1. Unicornuate uterus with two cavitated, non-communicating rudimentary horns. The inferior horn has endometrial polyps.

in the AFS system which accurately defines our patient's anatomy; and specifically, it does not include multiple rudimentary horns accompanied by a unicornuate uterus.

During embryogenesis, the Müllerian ducts appear at ~7 weeks gestation. The site of invagination of the coelomic epithelium to become the Müllerian duct will ultimately be the abdominal opening of the Fallopian tube. Müllerian ducts then descend medially within the mesenchyme to fuse with the duct of the opposite side. The fused septum is ultimately resorbed producing a single cavity which meets the upward growing urogenital sinus. Müllerian abnormalities are therefore thought to result from failure of lateral fusion, failure of vertical fusion or failure of resorption. For the patient presented, the development of two separate rudimentary horns suggests both failure of lateral fusion and interruption of canalization (Figure 1), offering some insight into embryogenesis of the Müllerian system.

A case of congenital absence of the vagina with a grossly normal-appearing uterus has been reported (Davis *et al.*, 1992). Hysterotomy at the time of laparotomy revealed a slit-like cavity measuring 2.5 cm in length and 0.6 cm in width with patency to the right tube but no communication with the lower uterine segment. Examination of the post-hysterectomy specimen revealed a second 0.5 cm area of patency within the cervical tissue. These findings support the theory (Crosby and Hill, 1962) that canalization follows fusion and that it can begin at any location along the line of fusion and proceed in any direction. On the other hand (Muller *et al.*, 1967), it has been postulated that between 13 and 20 weeks gestation resorption occurs first in the isthmic region of the uterus and proceeds simultaneously in both directions (cranially and caudally). This would explain such anomalies as a double vagina and cervix with normal uterus (Goldberg and Falcone, 1996). Midline resorption has also been postulated to start at the isthmus and directed caudally unifying the cervix and vagina. The uterine septum is then thought to be resorbed in a cephalad direction. Failure of reabsorption of this septum can result in a septate uterus (Thompson and Rock, 1992). For our patient, the presence of two rudimentary horns stacked one on top of the other with the superior horn having a patent Fallopian tube suggests that one Müllerian duct, after failure of lateral fusion, underwent incomplete canalization initiated from two distinct sites supporting previous theories (Crosby and Hill, 1962; Davis *et al.*, 1992).

The entirely separate origin of the ovaries from the gonadal

ridges explains the infrequent association of uterovaginal anomalies with ovarian anomalies (Thompson and Rock, 1992). However, another perplexing aspect of this case is the tenuous communication of the right infundibulopelvic ligament to the right ovary. Instead the ovary seemed to dangle from the ovarian ligament on the right. At ~5 weeks of gestation, during the indifferent gonad stage of ovarian embryology, the paired gonads are structurally consolidated coelomic prominences overlying the mesonephros and forming the gonadal ridges. Seven to 10 days later, the mesonephros, together with the genital ridge, is called the urogenital ridge, indicating the close relationship of the urinary and reproductive systems. While the intimate relationship between Müllerian and renal development has been well publicized and while unilateral ovarian absence is usually associated with a failure of the ipsilateral tube, half of the uterus, kidney and ureter, a clear association between this patient's ovarian and Müllerian anomalies awaits future understanding of ovarian development.

In conclusion, the triad of dysmenorrhoea beginning at menarche, increasing severity of dysmenorrhoea with each menses and a unilateral pelvic mass is strong evidence for the presence of congenital Müllerian dysgenesis (Silber et al., 1990). Preoperative evaluation should include evaluation of the urinary tract. The reported incidence of major renal anomalies associated with incomplete uterine duplication with a non-communicating rudimentary horn ranges from 31 to 100% (Buttram and Gibbons, 1979; Silber et al., 1990). The most common anomaly is renal agenesis ipsilateral to the non-communicating rudimentary horn, with ipsilateral pelvic kidney the second most commonly reported anomaly (Silber et al., 1990).

Operative laparoscopy may be an alternative treatment for the symptomatic noncommunicating rudimentary horn (Amara et al. 1997; Falcone et al., 1997). We have previously reported two cases of laparoscopic excision of a non-communicating rudimentary uterine horn (Nezhat et al., 1994; Amara et al., 1997). Including the present case, 16 cases of laparoscopic resection of rudimentary horns have been reported to date , including three for management of rudimentary horn pregnancies (Tables I and II). Commonly accepted benefits of minimally invasive surgery such as enhanced visualization of the cul-de-sac, decreased adhesion formation, smaller incisions, reduced postoperative pain, and shortened hospital stay argue in favour of the laparoscopic approach. Surgical expertise, experience, and availability of proper instrumentation must also be considered.

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