A case of unicornuate uterus with ipsilateral ovarian and renal agenesis

Bulent Haydardedeoglu, M.D., Erhan Simsek, M.D., Esra Bulgan Kilicdag, M.D., Ebru Tarim, M.D., Erdogan Aslan, M.D., and Tayfun Bagis, M.D.

Baskent University Faculty of Medicine, Department of Obstetrics and Gynecology, Adana, Turkey

Objective: To present a case of a unicornuate uterus with ipsilateral ovarian and renal agenesis.

Design: Case report.

Setting: Baskent University Faculty of Medicine, Department of Obstetrics and Gynecology, Adana, Turkey.

Patient(s): A 48-year-old gravida 3 (para 3 with term deliveries) woman with a 6-month history of menometrorrhagia was admitted to our clinic. Our diagnosis was a pedunculated submucous leiomyoma that protruded into the vagina.

Intervention(s): The patient chose to have a total abdominal hysterectomy and unilateral salpingoopherectomy.

Main Outcome Measure(s): During laparotomy, a unicornuate uterus with a noncommunicating horn, together with ipsilateral ovarian agenesis, was observed. A total abdominal hysterectomy and unilateral salpingoopherectomy were performed successfully. Because we could not detect the left ovary and left ureter during the operation, we planned an abdominal ultrasonography and intravenous pyelography (IVP) postoperatively to demonstrate possible urinary tract abnormalities.

Result(s): We detected left renal agenesis by IVP.

Conclusion(s): We presented a very rare clinical condition that demonstrates a unicornuate uterus with a noncommunicating horn, and ipsilateral ovarian and renal agenesis concomitantly. The absence of one ovary and one kidney in our case may be explained by the abnormal development of organs derived from a unilateral urogenital ridge. (Fertil Steril 2006;85:750.e1–4. ©2006 by American Society for Reproductive Medicine.)

Key Words: Unicornuate uterus, ovarian agenesis, renal agenesis

The incidence of Müllerian anomaly in the reproductive age group is 3%–5% and 5%–10% in recurrent miscarriages (1). The unicornuate uterus is an infrequent type of Müllerian anomaly, which is due to a failure of development of one Müllerian tract. The unicornuate uterus represents 6.3% of congenital uterine anomalies (2), which have significant problems with reproductive outcome secondary to abnormal uterine vasculature and decreased muscle mass. Approximately 40% of patients with unicornuate uteri have urinary tract anomalies, usually of the kidney (3). The occurrence of Müllerian anomalies with concomitant gonadal developmental abnormalities is very rare. Case reports about the unicornuate uterus with ovarian abnormalities are very limited in the literature. We present a case of a unicornuate uterus together with ipsilateral ovarian and renal agenesis.

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Reprint requests: Bulent Haydardedeoglu, M.D., Obstetrics and Gynecology Department, Cemalpasa Mah 5 Sok, Ferah Apt Kat:9 Daire:18, Seyhan, Adana, Turkey (FAX: +90 322 327 12 73; E-mail: bulenthaydar@yahoo.com).
During laparotomy, the unicornuate uterus with a non-communicating horn, together with ipsilateral ovarian agenesis, was observed (Fig. 1, Fig. 2, and Fig. 3). A Y-shaped dissection was performed on the uterus, and a fibroid-like mass originating from the cavity of unicorn
site and passing down through cervix was observed. During laparotomy, the left ovary and left ureter could not be visualized, which indicated a potential left renal agenesis. Postoperative urine output was normal; therefore, an abdominal ultrasound and intravenous pyelography (IVP) were performed to reveal urinary tract abnormality. The results indicated left renal agenesis (Fig. 4). We suspected an ectopic ovary, thus postoperative serum FSH and E₂ levels were measured to rule out an ectopic ovary. Serum FSH and E₂ levels were 77 IU and 14 pg/dL, respectively, which confirmed that no ectopic ovarian tissue was present.

Pathologic examination of the uterus and right ovary indicated a unicornuate uterus with a noncommunicating horn having a submucous fibroid and a normal cervix, as well as a proliferative endometrium together with a normal right ovary.

**FIGURE 4**

Intravenous pyelography (IVP) revealing left renal agenesis.
A classification based on the degree of failure of normal development of the female genital tract was proposed in 1979 by Buttram and Gibbons (4) and modified in 1988 by the American Society for Reproductive Medicine. According to this classification, the unicornuate uterus represents class II. This malformation is the result of defective development of one of the two Müllerian ducts and is frequently associated with a contralateral rudimentary uterine horn (5). Four subtypes of this anomaly have been described: communicating horn, noncommunicating horn, horn without cavity, and a unicornuate uterus without a horn. Our case was a non-communicating one.

Urinary tract anomalies occur frequently in association with all types of uterine anomalies, except diethylstilbestrol-related abnormalities (class VII). In the original study, Buttram and Gibbons (4) reported that 31% of their patients with uterine abnormality have urinary anomalies in which congenital absence of a kidney was the most common. Urinary tract abnormalities occur more frequently in class I and class II uterine anomalies than with those in classes III, IV, or V (6). Because of this knowledge, we found congenital unilateral renal agenesis in our case.

Pedro Acien, in his report in Human Reproduction in 1992, suggested that a unicornuate uterus may also be caused by complete agenesis of all the organs derived from one urogenital ridge, resulting in a unicornuate uterus and, on the contralateral side, no uterine horn or ovaries and renal agenesis or hypoplasia (5). Therefore, the absence of one ovary and one kidney in our case may be explained by the abnormal development of organs derived from unilateral urogenital ridge. However, to our knowledge, a very limited number of case reports are available in the literature to describe concomitant occurrence of gonadal and uterine developmental abnormalities, especially a unicornuate uterus. We could find only two case reports related to this condition: one with unicornuate uterus and ectopic ovary and the other with a unicornuate uterus with ovarian agenesis together with a pelvic kidney (7–8).

Another interesting finding in this case is the presence of three successful term deliveries in our patient’s history. Buttram and Reiter have demonstrated that spontaneous abortion and premature delivery rates are very high in patients with unicornuate uterus (48% and 17%, respectively) with a total live birth rate of 40% (6). We think that the competence of the unicornuate cavity may differ from patient to patient and may explain the three consecutive term pregnancies with one ovary in this case. This case could help with reproductive counseling for patients with unicornuate uteruses. It must also be remembered that patients with noncommunicating functional horns are at increased risk of morbidity secondary to the development of hematometrium, hematosalpinx, endometriosis, and rudimentary horn gestation, and they should have the horn excised.

In conclusion, we presented a very rare clinical condition that demonstrates a unicornuate uterus with a noncommunicating horn, and ipsilateral ovarian and renal agenesis concomitantly. To our knowledge, this is the first case report that involves agenesis of organs derived from a unilateral urogenital ridge.

REFERENCES