A Complicated Hematometra and Fallopian Tubal Occlusion in Uterine Didelphys With Unilateral Cervical Atresia

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Uterine anomalies are uncommon Müllerian malformations without known etiology. Many hypotheses have been put forward to explain these anomalies such as the involvement of genetic, environmental, and pharmacological issues [1]. The incidence is reported to be between 0.5–5.0% of all women [2]. However, a congenital uterine anomaly involving cervical atresia is extremely rare. We report a patient, diagnosed with a uterine anomaly, who underwent a primary laparotomy for unilateral endometrioma without resectioning of the rudimentary horn. Postoperatively, the patient received 6 months of gonadotropin-releasing hormone agonist (GnRHa) treatment as a conservative therapy. After completion of the GnRHa therapy, endometriosis recurred and a hematometra developed. This occurred after her menstrual cycle had resumed for 3 months post GnRHa treatment. The patient underwent a second laparotomy and the rudimentary horn was resectioned. Recovery after the operation was uneventful. The final diagnosis of the uterine anomaly in this patient was uterine didelphys with unilateral cervical atresia.

A 23-year-old nulligravid woman had complained of dysmenorrhea since menarche at the age of 14 years in 1999 and this had been partially relieved with medical management. At the age of 22 years during April 2007, the patient presented with acute abdominal pain in the left lower quadrant, in which an abdominal mass was palpable. Pelvic ultrasonography by a primary gynecologist revealed a 10 × 10 × 10 cm complex mass of the left adnexa with multiple cysts and septa. A bimanual pelvic examination showed a single vagina and single cervix. Computed tomography (CT) revealed a multicystic mass associated with the left ovary, possibly endometrioma, and two uterine horns indicating the possibility of a bicornuate uterus (Figures A and B). The right kidney was enlarged and the left kidney was absent. An elevated serum CA-125 level of 8,640 U/mL was noted. The patient underwent exploratory laparotomy by the primary gynecologist. The findings included a 10 cm left ovarian endometrioma, severe endometriosis involving the cul-de-sac and the pelvis, as well as layering of hemosiderin throughout the uterine horns, the pelvis and the omentum. Fibrous adhesions involving the sigmoid colon, small intestine and pelvic sidewall were also noted. The right uterine horn and the right fallopian tube were normal in appearance; a right ovary could also be identified. The uterine anomaly was thought to be a bicornuate uterus. Left partial oophorectomy and lysis of the adhesions were performed. The pathological findings revealed ovarian endometrioma and endometriotic cysts that had endometrial epithelium and hemosiderin-laden macrophages. Postoperatively, the patient recovered quickly and was prescribed 6 months of GnRHa therapy.

In June 2008, the patient, who had resumed her menstrual cycles for 3 months after completion of the GnRHa therapy, presented at the hospital again with recurrent acute abdominal pain. Pelvic ultrasonography by a second gynecologist (Chi-Feng Su) revealed a 5 × 3 cm left adnexal complex with multiple cysts that favored a recurrent endometrioma together with a 6-cm left hematometra, a normal-appearing right uterus and a 4-cm right ovarian cyst. The findings of a CT study were compatible with the ultrasonographic results (Figures C and D). The serum CA125 level was 269.9 U/mL. A second laparotomy was performed. Intraoperatively, a severe pelvic adhesion was noted involving the sigmoid colon, left fallopian tube, left ovarian endometrioma and left pelvic sidewall. Lysis of adhesion was performed uneventfully. The left uterus, which was attached at the vaginal apex, was found to be separated from the right uterus by approximately 2 cm. The left uterine horn and left adnexa including the fallopian tube and remnant ovary were removed completely. An accumulation of menstrual debris was identified within the left uterine

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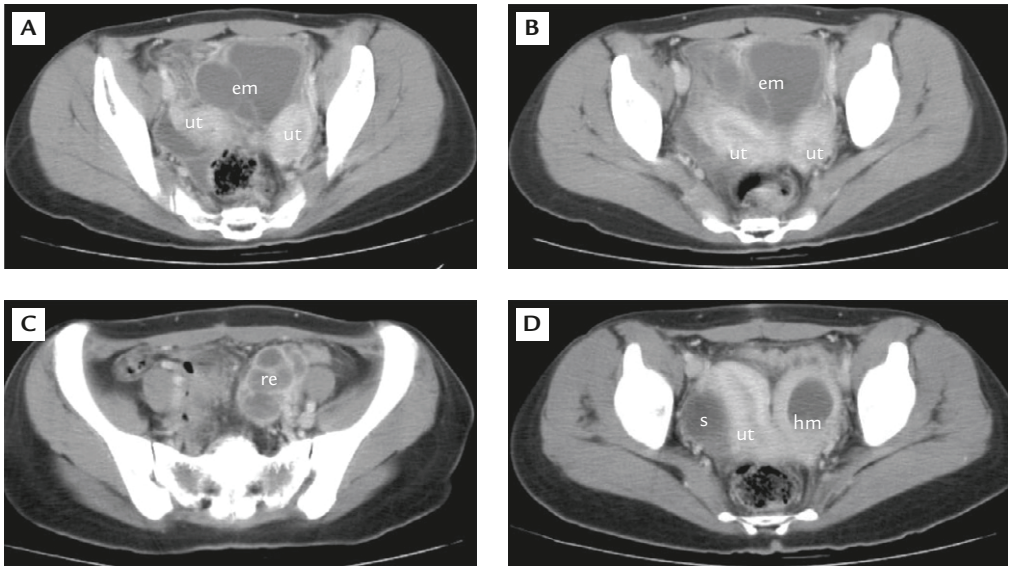

Figure. Computed tomography (CT) prior to the primary laparotomy. (A, B) Severe (stage IV)left endometrioma with multiple cysts (em) and a double uterus (ut). (C, D) CT prior to the second laparotomy. Recurrent left endometrioma (re) and a complicated left obstructed uterine horn with hematometra (hm) are shown. s = Right ovarian simple cyst.

horn on dissection. The right uterine horn and right fallopian tube appeared to be normal. A 4-cm right follicular cyst was noted and a cystectomy was performed. Pathological analysis of the left uterus showed an atresic cervix, inflamed endometrium and inflamed fallopian tube. The left remnant ovary contained multiple endometriosis. The right ovarian cyst was consistent with a simple cyst. The patient recovered well postoperatively.

The left hematometra had formed because of blocked menstrual outflow as a result of the unilateral cervical atresia. This occurred because of obstruction of the retrograde menstruation and ipsilateral tubal occlusion. The tubal occlusion may have occurred following the primary operation and of chronic inflammation and peri-tubal adhesion. The final diagnosis was a complicated hematometra and uterine didelphys with unilateral cervical atresia and postoperative ipsilateral tubal occlusion.

We categorized this case as uterine didelphys with unilateral cervical atresia (class III) according to the American Fertility Society Classification Scheme (1988) [3]. Our case could be distinguished from bicornuate uterus (class IV) because the present case had developed uterine horns and cavities. This case was also different from a unicornuate uterus with rudimentary horn (class II), because our patient presented with two uterine horns and both had an independent connection to the vagina. An explanation of the findings in the present case can be outlined as follows. The left cervical atresia resulted in a failure of menstrual blood outflow from the left uterine horn. The cyclic pain that the patient had suffered from the age of 14 years indicated retrograde menstruation from the left uterine horn to the peritoneal cavity along the patent left fallopian tube. The right uterus with a normal fallopian tube, a single cervix and a single vagina underwent normal cyclic menstrual flow. The patient's symptoms were relieved by medication. Therefore, this patient neglected the dysmenorrhea, which had continued for 6 years until the age of 22 years. The retrograde menstruation from a functioning left uterine horn resulted in the development of severe ovarian endometrioma and pelvic endometriosis. Based on the findings of a high level of CA-125 and a large ovarian endometrioma by CT, the patient underwent primary laparotomy for the removal of the left endometrioma. Initially, the diagnosis was thought to be bicornuate uterus with communication because of the regular cyclic menstruation in the patient and the presence of two uterine horns with a single cervix and vagina. The patient underwent conservative treatment, specifically a 6-month cycle of GnRHa therapy, and she was symptom-free during that period.

The cervical atresia of the left uterine horn and the lack of communication between the two uterine cavities could have been diagnosed if hysterosalpingography had been performed before the primary laparotomy. It also would have been advisable to remove the left uterus when it was first recognized. The potential complications could then have been prevented. Specifically, the occlusion of the ostium of the left fallopian tube developed because of tubal inflammation and adhesion after the primary operation, and this then resulted in left horn hematometra due to complete blockage of menstrual flow. Although the patient had a 6-month symptom-free period during the GnRHa therapy, acute abdominal pain and cyclic cramping recurred soon after menstruation

had resumed. After informing her family of the problem of hematometra in the obstructed uterus, a laparotomy was performed with informed consent to remove the left uterine horn and left adnexal endometriosis. One month postoperatively, the patient experienced normal menstruation without any abdominal cramping. In addition, the patient's CA-125 level returned to normal (16.07 U/mL). A hysterosalpingography was also performed 3 months later, which revealed a single uterine horn with a patent right fallopian tube. Thus, fertility was preserved in this patient.

The clinical presentation varies in patients with anomalies of the uterus and symptoms such as abdominal pain, dysmenorrhea and foul vaginal discharge may be present at a mean age of 17 years [5]. Typically, the onset of symptoms occurs 1 to 2 years after menarche [1,5]. Endometriosis and tubo-ovarian abscess have been reported to result from menstrual back flow and retrograde bacterial infection [1,5]. Removal of the functioning rudimentary horn with the obstruction and cervical atresia just after menarche should be able to prevent the development of endometriosis and hematometra [2,4]. An examination of the literature with regard to uterine didelphys, cervical atresia and hematometra using a PubMed computerized search was carried out. This indicated that our patient was a rare case and there are very few similar reports. Only two similar cases were found [6,7]. Knight and Birkinshaw [6] reported a 14-year-old young woman with lower back pain and a pelvic cystic mass who underwent laparotomy; she was found to have uterine didelphys with hematometra of the right uterus and unilateral cervical atresia. Sherer et al [7] reported a 24-year-old nulliparous woman with secondary dysmenorrhea who was diagnosed with uterus didelphys involving a distended obstructed right uterus and unilateral cervical atresia. Both patients presented as a primary case and were successfully treated by primary laparotomy and resectioning of the rudimentary uterus. However, they differed from the present case because our patient developed hematometra and an obstructed uterus after the primary surgery for endometrioma followed by GnRHa therapy. Failure of the above treatments gave rise to a need for a second operation.

In conclusion, our patient was an unusual case that has never been previously reported. We consider that a patient diagnosed with a uterine anomaly should undergo hysterosalpingography before the operation to identify the type of uterine anomaly that is present. Furthermore, endometriosis in patients with uterine didelphys and unilateral cervical atresia should undergo resectioning of the uterine rudimentary horn to prevent any subsequent complications. Moreover, treatment of such as case conservatively using GnRHa does not seem to be a useful approach.

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