Adenomyosis in a patient with the Rokitansky-Kuster-Hauser syndrome

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Endometriosis remains an enigma despite extensive clinical investigations and experience. Several theories have attempted to explain the puzzling and controversial pathogenesis of endometriosis. Sampson’s implantation theory is the most commonly accepted. Early endometriotic lesions have been found in the Douglas cavity, where retrograde menstruation was confirmed in 90% of women. These findings support Sampson’s theory. In contrast, endometriosis occurs in some patients who have no functional endometrium, such as those with the Rokitansky-Kuster-Hauser syndrome, a clinical condition that supports Iwanoff and Mayer’s coelomic metaplasia theory.

Most recent experimental studies support both the implantation theory and the coelomic metaplasia theory (1, 2). Of the published case reports of endometriosis associated with the Rokitansky-Kuster-Hauser syndrome, none have reported adenomyosis in a patient with this syndrome. We report a rare case of adenomyosis in a patient with the Rokitansky-Kuster-Hauser syndrome and discuss its implications in the pathogenesis of adenomyosis.

A 27-year-old Japanese woman came to Tottori University Hospital for evaluation of lower left abdominal pain that occurred every month. She reported cyclic pain lasting 3–5 days since she was 16 years of age. We first examined her for primary amenorrhea at 19 years of age. At that time, we diagnosed the Rokitansky-Kuster-Hauser syndrome by means of laparoscopy and performed vaginoplasty by using the McIndoe procedure. During the first laparoscopic visualization of the pelvis, we confirmed the presence of rudimentary uterine horns on the apex of bilateral normal tubes and ovaries. The left uterine horn, which was about 3 cm in diameter, was larger than the right. The patient did not seek further care at our hospital until she was 27 years of age. When she returned at age 27, she reported that she had had monthly abdominal pain since she was last seen here. Increasingly severe pain caused her to again seek care.

Pelvic examinations revealed normal external genitalia, a blind vagina of almost normal length, and a tender hen’s-egg–sized tumor in the left pelvic cavity. The basal body temperature chart was biphasic. Laboratory examination showed normal gonadotropin level (LH, 1.80 mIU/mL; FSH, 5.38 mIU/mL) and an elevated CA125 level (170.2 U/mL). Transvaginal ultrasonography showed a 4.9 × 4.6 × 4.4 cm tumor in the lower left abdomen. Magnetic resonance imaging showed a tumor 5 cm in diameter in lower left abdomen, with irregular intensity and no myoma nodules. We suspected myoma or adenomyosis arising from the left rudimentary uterine horn.

We administered an injection of GnRH agonist (leuprolein acetate, 1.8 mg [Takeda Pharmaceutical Co., Tokyo, Japan]) to control the patient’s pain. After obtaining informed consent, we decided to perform laparoscopic tumor resection.

Laparoscopic visualization of the pelvis revealed no adhesions, ascites, or endometriotic lesions. Bilateral fallopian tubes and ovaries were normal; however, rudimentary uterine horns were seen on the apex of each tube. The horn on the right was small, while that on the left measured 5 cm in diameter. The attachments were the round and uteroovarian ligaments.
inserted in the anterior lateral and posterior medial aspects, respectively. Using a bipolar forceps, the left round ligament was grasped and coagulated, then cut with a scissors forceps. The proximal fallopian tube was grasped, elevated, and coagulated. The müllerian remnant was then released from its tubal attachment. The uteroovarian ligament was similarly coagulated and cut.

The tumor was then mobilized medially with traction by using an atraumatic forceps. The ureter was visualized well below the attachment of the medial leaf of the peritoneum. The tumor was carefully removed at the anterior fibrous band of the midline. A mini-Pfannenstiel incision was made to remove the tumor. The tumor had no myoma nodules; however, there were lesions like blueberry stains, containing dark-brown bloody fluid. Serial sections through the tumor showed no endometrial cavity. Histologic examination revealed adenomyosis (Fig. 1). The patient was uneventfully discharged from the hospital. She has been free of pain since the surgery.

To our knowledge, this is the first case report in the literature of adenomyosis in a woman who had the Rokitansky-Kuster-Hauser syndrome and therefore no functional endometrium. She had severe intermittent lower left abdominal pain lasting 3–5 days every month for 10 years. Histologic examination revealed the presence of endometrial glands and stroma in the myometrum. This case strongly suggests the coelomic metaplasia theory in its histogenesis.

Other authors have reported laparoscopic removal of müllerian remnants in a woman with congenital absence of the vagina (3). This earlier report, in which patients complained of monthly recurrent pain, did not present the histopathology. In one of the reports, the authors used a morcellator to reduce the size of the tumor. To make a definitive histopathological diagnosis, we removed the tumor through a mini-Pfannenstiel incision.

Uterine adenomyosis is a benign entity characterized by the heterotopic growth of endometrial glands or stroma (or both) into the myometrium, as well as myometrial hypertrophy and hyperplasia. Most histologic criteria for diagnosing adenomyosis have been based on Cullen’s proposal that adenomyomas arise through direct invasion of the uterine mucosa into the uterine musculature. Some pathologists define adenomyosis as glands and stroma invading the myometrium to the depth of at least one third of the thickness of the uterine wall. In contrast to the established criteria, we found endometrium-like tissues containing both surface epithelium and stroma in myometrium of our patient, who did not have functional endometrium. The histogenesis of adenomyosis in this patient may be a mechanism other than direct invasion.

Recently, it was proposed that three types of endometriotic lesions—peritoneal, ovarian, and rectovaginal endometriotic lesions—must be considered separate entities with different pathogenesis (4). The authors suggest that the rectovaginal endometriosis nodule, whose histogenesis is related to metaplasia of müllerian remnants in the rectovaginal septum. The findings in this report may support this hypothesis. The present case suggests that adenomyotic lesions may develop in the myometrium of müllerian remnants, supporting the metaplasia theory in its histogenesis.

References