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Case Report
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Mayer-Rokitansky-Küster-Hauser Syndrome Complicated with Pedicle Torsion of Ovarian Serous Cystadenoma: A Case Report

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Introduction

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is caused by either hypoplasia or caudal dysplasia of the Müllerian duct system. It is characterized by congenital vaginal agenesis, with a prevalence of 1/4000 to 1/5000 [1]. Patients with MRKH typically either have no uterus or only rudimentary uterus. Some of them may also show urologic anomalies and, in rare cases, spinal malformations. They all have a normal female karyotype of 46 XX with normal ovarian functions [2]. Ovarian cyst pedicle torsion is a common gynecological acute abdomen with pedicle torsion occurring in approximately 10 percent of ovarian cysts.

However, MRKH syndrome with ovarian cyst torsion is rare. This paper summarizes the clinical data of a MRKH syndrome case complicated with pedicle torsion and necrosis of a giant ovarian serous cystadenoma.

Case Presentation

The patient is a 23-year-old female, who presented with 2-week history of irregular abdominal pain on the right lower quadrant, but no nausea, vomiting, fever, or urinary symptoms. This recurrent symptom initially started over a year ago and had been treated as "pelvic inflammation" with antibiotic in another hospital for multiple episodes. During that period, no ultrasonography was performed. The patient has a known history of MRKH syndrome and underwent vaginoplasty and ultrasonography two years prior. A postoperative follow-up ultrasound scan showed a right ovarian cyst of 4 cm.

Gynecological examination: normal development of vulva, narrow vagina, smooth mucosa, difficulty in accommodating 2 fingers, depth of about 7 cm; absence of cervix and uterine body; a cystic mass of 10 cm × 9 cm × 6 cm next to the right adnexal area with mild tenderness, unclear boundary and not easy to move; no abnormal observations on left adnexa.

Laboratory data: White blood cell count: 8.5 × 10^9/L. Neutrophil percentage: 66.9%. Serum amyloid A: 11.5 mg/L. High-sensitivity C-reactive protein: 5.1 mg/L. Fibrinogen: 4.04 g/L. D-dimer: 1.15 mg/L FEU. Anti-Mullerian tube hormone: 0.8 ng/mL (normal range 1.52-9.95). Sex hormones and thyroid function were normal. Karyotype was reported as 46 XX.

MRI showed a triangular soft tissue between the giant cyst and the bladder. The structure of vagina was visible, while the boundary was not clear. Multiple cysts were found in the bilateral ovarian areas. The cross section of the right ovarian lesion was about 5.3 cm × 2.9 cm, posterior to this lesion there was a nodular signal of obvious enhancement at the size of 2.3 cm × 1.3 cm (Figure 1). The diameter of the largest left ovarian cyst was 1.9 cm. Next to it, there was nodular soft tissue of obvious inhomogeneous enhancement, with the size of 3.1 cm × 1.9 cm (Figure 2). MRI also demonstrated a large irregular tube-shaped cystic in the pelvis, at the size of 9.8 cm × 4.6 cm × 10.1 cm, with clear boundary, inhomogeneous wall and no obvious enhancement on contrast-enhanced scan (Figure 3). The kidneys, bladder, intestine, liver, all-bladder and lymph nodes were reported normal.

Given her medical history of MRKH, the triangular soft tissue above the bladder with bilateral nodules was consistent to an undeveloped primordial uterus. Bilateral ovarian structures were not clearly shown. There were multiple cystic foci in the ovarian region and the right cystic focus showed signs of hemorrhage. The preliminary diagnosis was ovarian cyst. However, because the

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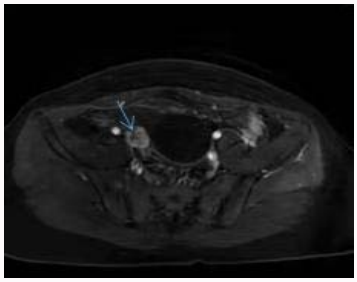
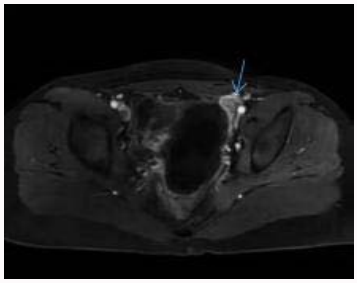


Figure 1:



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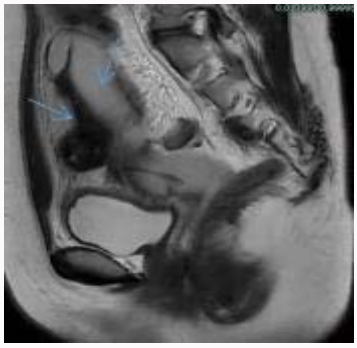

Figure 3:

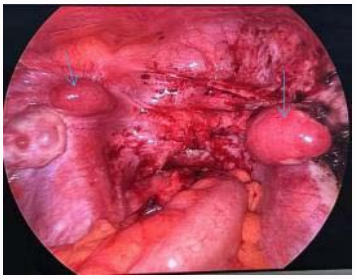

Figure 4:

Figure 5:

giant cystic focus of the pelvis extended to the right ovarian region the possibility of it being the pedicle torsion of the right ovarian cyst couldn't be eliminated.

The operation began with a diagnostic laparoscopy, which revealed a dysplasia-uterine-like muscular tissue of about 3 cm × 4 cm ×3 cm in the left pelvic cavity near the iliac fossa (Figure 4). Adjacent to it was an oviduct-like tubal structure with part mucosal structure at the umbrella end. Below it was the ovary of about 3 cm × 4 cm × 2.5 cm, with no obvious abnormality in appearance nor obvious vegetative growth on the surface. The uterine-like muscular tissue and the left adnexa were stuck together with the left pelvic wall. There was a cystic mass in the right pelvic cavity, partially dark purple and covered with omentum, densely adhered to the right pelvic and anterior abdominal wall. After the separation of the covered omentum and membranous adhesins, a dumbbell-shaped cystic-solid mass emerged. The larger part was about 11 cm × 6 cm × 8 cm, the other part was about 6 cm × 7 cm × 6 cm (Figure 5). There was no obvious normal ovarian tissue and no obvious fallopian tube-like structure. No abnormal lesions were found in the omentum, stomach, liver, spleen and diaphragmatic roof. A partial resection of the right mass was sent for intraoperative frozen biopsy and was reported as right ovarian serous cystadenoma. Intraoperative diagnosis was pedicle torsion and necrosis of right ovarian serous cystadenoma with bilateral dysplasia uterine. Therefore, laparoscopic right ovariectomy and pelvic adhesion lysis were performed. The surgery went smoothly, and the patient recovered well. Postoperative pathological report was the same as the intraoperative frozen biopsy. On the follow-up, 2 months after operation, the AMH was 1.13 ng/mL, and the sex hormones were still in normal range.

Discussion

MRKH syndrome is characterized by congenital absence of the vagina and uterus, which is generally divided into 2 types: Type I, more common, only vagina and uterus are affected; type II: In addition to no uterus nor vagina, patients also suffer from malformations of renal and skeletal systems. It should be distinguished from hymen atresia [3], vaginal atresia [4] and Complete Androgen Insensitivity Syndrome (CAIS) [5]. Studies suggest vaginal dilatation as the first-line treatment for MRKH syndrome [6-8], while surgery is also an option. The surgery is called vaginoplasty, including biological patch and peritoneal method. Improved surgical methods are reported constantly to benefit patients [9-11]. The mental health of postoperative patients is also critical and shouldn't be overlooked [12]. For patients with MRKH syndrome, the risk of pelvic complications should not be ignored in clinical practice, and routine gynecological physical examination should also be performed. There are reported cases of MRKH syndrome complicated with uterine leiomyoma [13-15], adenomyosis [16] and so on, which could have resulted in misdiagnosis.

MRI is an important imaging examination method for diagnosis of MRKH syndrome [17,18], which helps to determine the presence of uterus or primordial uterus. A recent retrospective analysis showed that MRI results in determining the degree of reproductive system involvement and multi-system related malformations are highly consistent with the final clinical diagnosis, proving the potential value of MRI in shortening preoperative diagnosis time [19]. Clinically,

ultrasound and MRI can be collaboratively used to reach the diagnosis of pelvic complications in patients with MRKH syndrome.

In regards to the choice of surgical methods [20], considering this patient's clinical history, and the fact that the right ovarian cyst had been twisted for too long, resulting in extensive hemorrhage and necrosis which led to the right ovarian function loss, right ovariectomy was suggested and performed. As for whether the uterus should be retained, given 1) the patient did not have periodic abdominal pain, 2) her uterus was considered as the primordial uterus, and 3) resection might affect the blood supply to the other healthy ovary and eventually lead to the decrease of ovarian reserve function, resection was not recommended for the contralateral (left) uterus. As for the affected side (right) uterus, since it may be complicated with uterine fibroids, adenomyosis and other related complications, we had suggested resection to her family members but the suggestion was rejected. After the surgery we reiterated the relevant risks to the patient and suggested routine follow-up visits.

For this case, due to congenital dysplasia, the distance between the ovarian suspensory ligament and the ovarian proper ligament was relatively short, so the pedicle torsion was more likely to happen anatomically. Given her MRKH history and repeated abdominal pain symptoms, if the imaging examination was performed sooner, a timelier surgery could have been performed to avoid adverse consequences such as cyst necrosis and extensive hemorrhage.

To sum up, MRKH syndrome is rare in clinical practice, yet, there is still a probability of complication of ovarian torsion due to the malformation of the ovarian ligaments. For such cases, physicians should be aware of this syndrome and pay more attention to the timely diagnosis and treatments.

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