In the most recent publications on MRKH syndrome, the uterine remnants and ovaries in patients may develop uterine remnant leiomyoma, adenomyosis, or ovarian tumor, and this can lead to problems in differential diagnosis. Here we summarize the diagnosis methods and available interventions for ovarian tumor in MRKH syndrome, with emphasis on the relevant clinical findings and illustrative relevant case. According to the clinical findings and illustrative relevant case, with the help of imaging techniques, ovarian tumors can be detected in the pelvis in patients with MRKH syndrome and evaluated in terms of size. Laparoscopy could further differentiate ovarian tumors into different pathological types. In addition, laparoscopic surgery not only is helpful for the diagnosis of MRKH combined ovarian tumor, but also has a good treatment role for excising ovarian tumor at the same time. Moreover, laparoscopic removals of ovarian tumor can be considered as a safe and reliable treatment for conservative management.

1. Introduction

The Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is characterized by congenital hypoplasia of the uterus and the upper part of the vagina. The incidence of MRKH syndrome has been estimated as 1 in 4500 women [1]. The principal character is a primary amenorrhea in women presenting with normal development of secondary sexual characteristics and normal external genitalia, but congenital vaginal or a shallow concave nest in the vaginal mouth, congenital uterine, or uterus aplasia. The ovaries are normal and functional as well as the endocrine status. Karyotype is 46, XX, with no visible chromosome modification. At present, most of the studies suggest that MRKH syndrome has been considered as a genetic disease, and genes such as the HOXA7, HOXA9–13, HOXD9–13, and WNT4 have been considered as possible offenders [2].

In the most recent publications on MRKH syndrome, we could found some cases report about uterine remnant leiomyoma, or adenomyosis, but the ovarian tumor is rare in MRKH syndrome and is difficult to be diagnosed [3–8]. Although most publications about pelvic masses in MRKH are about uterine remnants, adenomyosis, or fibroids, the occurrence of ovarian tumors in MRKH could not be ignored as these patients do have ovaries.

Ovarian tumors in patients with MRKH are difficult to examine, especially if no vaginal reconstruction has been performed. The aim of this review is to describe the diagnosis methods and available interventions for ovarian tumor in Mayer-Rokitansky-Küster-Hauser (MRKH) Syndrome, with emphasis on the relevant clinical findings and illustrative relevant case.

2. Definition and Prevalence

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome was first characterized by Mayer, Rokitansky, Küster, Hauser, and Schreiner, which was estimated to complicate 0.2% of births annually [9, 10]. MRKH syndrome is usually present in the form of primary amenorrhea and abnormalities of internal...
Table 1: Case reports of MRKH syndrome with ovarian tumors.

| Study                  | Published year | Study location | Age | Diagnosis methods                        | Pathological type            | Treatment                        |
|------------------------|----------------|----------------|-----|-----------------------------------------|------------------------------|----------------------------------|
| Fukuda et al. [22]     | 2010           | Japan          | 50  | MRI, laparotomy and histological analysis | Ovarian mucinous cystadenoma  | Laparoscopic resection            |
| Huepenbecker et al. [23] | 2017           | United States  | 64  | CT, laparotomy and histological analysis | Serous ovarian adenocarcinoma | Laparoscopic resection            |
| Juusela et al. [24]    | 2017           | United States  | 72  | Laparotomy and histological analysis     | Bilateral ovarian Sertoli cell tumors | Laparoscopic resection            |
| Mishina et al. [25]    | 2007           | Moldova        | 35  | Ultrasound and histological analysis     | Ovarian dysgerminoma          | Oophorectomy                      |
| Nusrath et al. [26]    | 2016           | India          | 65  | CT, laparotomy and histological analysis | Ovarian endometrioid carcinoma | Laparoscopic resection and cytoreductive surgery |
| Tsaur et al. [27]      | 1995           | China          | 4   | Ultrasound, CT and histological analysis | Ovarian teratoma              | Oophorectomy                      |

Magnetic resonance imaging; CT: computed tomography.

After a review of the literature, we find that benign tumors have a majority in the 6 case reports and most studies use laparoscopy to remove the tumors.

3. Differential Diagnosis

The differential diagnosis of MRKH syndrome combined pelvic mass mainly includes pelvic mass originated from female genital tract (MRKH syndrome combined uterine fibroids, MRKH syndrome combined uterine adenomyosis, MRKH syndrome combined ovarian tumor, etc.) and derived from other pelvic organ (intestinal tract, mesentery, and retroperitoneal tumor). We need to choose the diagnostic method carefully to differentially diagnose the MRKH syndrome combined pelvic mass. The medical diagnosis of MRKH syndrome is based on the history of primary amenorrhea and the gynecological examination where patients have no vagina and no palpable uterus. The three most common methods of diagnosing MRKH syndrome are by magnetic resonance imaging (MRI), ultrasound, or by laparoscopy.

3.1. Illustrative Relevant Case. A 29-year-old young woman, who complained of primary amenorrhea and pelvic mass over 1 year, was used to as the illustrative relevant case. She presented with a cystic pelvic mass 10 cm in diameter on ultrasound and magnetic resonance imaging that could not be differentiated between polycystic ovary and ovarian cystadenoma. The patient was laparoscopically operated on, and the left ovarian tumor was detected and removed. Histology confirmed a benign ovarian serous cystadenofibroma.

3.2. Imaging Features. According to the literature review, ultrasound, MRI, and CT are the major imaging tools to diagnose the MRKH syndrome with ovarian tumors. The ultrasound image of MRKH syndrome is characterized by no normal uterus to be found in either longitudinal or cross-cutting image in the back of the filling bladder, but normal volume ovary on both sides [17]. Moreover, it is essential to check the abdominal cavity and the groin area to find the heterotropic uterus, ovary, and urinary system malformation by transabdominal ultrasound. Therefore, Ultrasonography is the most basic test for patients with MRKH syndrome and is helpful in finding the ovarian lesions. The method of ultrasonography plays an important role in the preliminary diagnosis [18]. But for surgical interventions, ultrasound
MRI and ultrasound imaging are valuable tools to diagnose the MRKH syndrome, as well as evaluate patients for concurrent renal anomalies, endometrioma, and tubal disease. Although CT is not a common method of diagnosing MRKH syndrome, it should be kept in mind that CT is useful in finding the ovarian tumors in MRKH syndrome.

From the above, imaging tools as ultrasound, MRI, and CT are valuable to find the ovarian tumors in MRKH syndrome and distinguish it from intestinal tract, mesentery, and retroperitoneal tumor. However, it is hard for imaging tools to diagnose the pathological type of ovarian tumors before operation.

3.3. Laparoscopy Features. Laparoscopy is the gold standard for evaluation of MRKH syndrome. But laparoscopy used in MRKH diagnosis is not an attractive method due to its invasive nature. Laparoscopy is more expensive than MRI and should be reserved for patients undergoing surgical intervention or guiding the process [11]. For MRKH patients with ovarian tumors, laparoscopy offers the possibility of diagnosing and treating at the same time. From our illustrative relevant case, laparoscopy was performed and revealed a large pelvic mass about 10 cm in diameter, which was located in left ovary lateral margin and was a multiple cystic clear boundary ovarian tumor with complete capsule. Suspensory ligament of left ovary, left proper ligament of ovary, and fallopian tube were 180 degrees of torsion. We thought the reason of the tumor torsion is associated with the heterogeneity of tumor. Left fallopian tube was normal, connected with left uterine nODULES. The right attachment was normal, connected with right solid nodules (Figure 3). The complete resection of the left ovary tumor was taken along the left ovary pole, resetting the left ovary and the fallopian tube. The pathological examination showed ovarian serous papillary cystadenofibroma (CAF) (Figure 4).

4. Treatment

Laparoscopy is the ideal technique to identify and treat ovarian benign tumor, so it may also be able to treat the ovarian benign tumor in MRKH syndrome. From our illustrative relevant case, we treated a rare case of large ovarian serous papillary cystadenofibroma in a young woman with the MRKH syndrome with laparoscopic surgery. After 6 months of postoperative follow-up, the patients recovered well. To the best of our knowledge, our illustrative relevant case describes the fifth case in which ovarian tumor in MRKH syndrome was removed under laparoscopy confirming that laparoscopy is a powerful tool for treatment as well as diagnosis of these tumor. In addition, cytoreductive surgery and oophorectomy are further needed to treat the ovarian malignant tumor in MRKH syndrome.

5. Conclusion

With the help of imaging techniques, ovarian tumors can be detected in the pelvis in patients with MRKH syndrome and evaluated in terms of size. Laparoscopy could
Figure 2: A 29-year-old female patient with primary amenorrhea. (a) Well developed breasts. (b) Vulva. (c) Vaginal vestibule. (d) T1W1 coronal view: yellow arrow: bilateral primordial uterus and red arrow: ovary tumor. (e) T1W1 axial view: yellow arrow: bilateral primordial uterus and red arrow: ovary tumor. (f) T1W1 sagittal view: yellow arrow: vagina, blue arrow: bladder, and red arrow: ovary tumor.

Figure 3: (a) The right attachment was 180 degrees of torsion. (b) The right attachment. (c) After left ovarian neoplasm resection. (d) Left ovary tumor.

Further differentiate ovarian tumors into different pathological types. In addition, laparoscopic surgery not only is helpful for the diagnosis of MRKH combined ovarian tumor, but also has a good treatment role for excising ovarian tumor at the same time. Moreover, laparoscopic removals of ovarian tumor can be considered as a safe and reliable treatment for conservative management. From the above, we think women should be inspected regularly, especially adolescents without menstruation, to check genital tract malformation and discover the pelvic diseases, such as ovarian tumors, leiomyoma, and attachment mass.

Disclosure
Jirui Wen is a co-first author.

Conflicts of Interest
There are no conflicts of interest related to this paper.

Authors’ Contributions
Yali Miao and Jirui Wen made an equal contribution to the paper.
Figure 4: A 29-year-old female patient with primary amenorrhea. (a) Microscopic findings of H&E staining (×10). (b) Microscopic findings of H&E staining (×40). Morphology of calcification in ovarian cancer.

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