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Case Report

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Case Report on Double Primordial Uterine and Vaginal Atresia with Torsion of Left Ovarian Cyst Pedicle

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ABSTRACT

Background: A 51-year-old woman had left lower abdomen pain for 18 hours with nausea and vomiting. Prior CT scans suggested pelvic neoplasms. Our hospital's emergency CT showed an enlarged uterus with cystic shadows, right adnexal cysts, and stomach fluid. Physical examination revealed left lower abdomen discomfort. A gynaecological examination revealed a painful, firm pelvic mass of 151210 cm. Further diagnosis is underway.

Method: The patient underwent emergency exploratory laparotomy, discovering a twisted, swollen left ovary with a 540° rotation, classified as a benign cyst. It was found that the patient had congenital upper vaginal atresia and bilateral initial uteri. Pain was reduced after surgery, thanks to symptomatic treatment. An abnormal karyotype of 46, XX,1qh+ was found during genetic testing.

Result: Fallopian tubes, uterus, and vagina develop from the embryonic accessory mesonephric duct. MRKH syndrome is caused by bilateral accessory mesonephric duct dysplasia and disappearance of the uterus or vagina. MRKH has three types, with Type 1 lacking uterus or vagina. Due to ovarian cyst torsion, this Type 1 MRKH with double initial uterus and upper vaginal atresia needed left adnexa resection. Genetic testing showed a typical female karyotype. MRKH's complex aetiology incorporates chromosomal abnormalities, emphasizing early cytogenetic evaluation for personalized treatment and fertility assistance.

Conclusion: Early cytogenetic testing for MRKH syndrome patients is crucial for determining the underlying cause and guiding personalized treatment plans to restore reproductive function and improve quality of life.

Key-words: Double primordial uterus; MRKH syndrome; Upper vaginal atresia; Torsion of left ovarian cyst pedicle

INTRODUCTION

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is a rare congenital disorder affecting the female reproductive system [1]. An underdeveloped or absent uterus and vagina characterize it. Affected individuals typically do not experience menstrual cycles. MRKH syndrome occurs in about 1 in 4,500 females at birth. External genitalia and secondary sexual characteristics, like pubic hair and breasts, develop normally.

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Treatment for MRKH syndrome focuses on creating a functional vagina and managing any associated symptoms [2,3].

Case Presentation

General Information- The patient, a 51-year-old female, presented with "persistent pain in the left lower abdomen for more than 18 hours." She was admitted to our hospital on January 07, 2022. At 10:30 on January 6, 2022, the patient developed persistent pain in the left lower abdomen after changing position, which was alleviated in the left decubitus position and aggravated in the right decubitus position, and was accompanied by nausea and vomiting, and the vomit was the stomach contents. Before admission, CT examination of the whole abdomen in the local hospital showed: 1. Multi-locular cyst-like changes on the left side of the pelvic cavity,

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considered as possible neoplastic lesions (increased density of the left anterior edge of the lesion), and the uterus was unclear. Enhanced CT examination of the middle and lower abdomen was recommended. 2. The right accessory area is circular with low density and was suggested for clinical correlation; 3. A plain scan of the liver, bile, pancreas, spleen, and kidneys showed no apparent abnormalities. On admission, emergency abdominal CT of our hospital (Figure 1) showed: 1. Uterine volume was slightly increased, and multiple cystic low-density shadows were shown; Small cystic hypodense foci in the right adnexal area; Nature was to be determined. 2. Abdominal and pelvic cavity contain a small amount of fluid. 3. A little calcification in the abdominal aorta. 4. No definite abnormal density shadow was found on CT plain scan of the liver, bile, pancreas, spleen and both kidneys.

The patient was reported to be in good health previously. History of allergy to penicillin. History of menstruation, marriage, and childbearing: never menstruated, married at 18, remarried at 29, husband in good health. On admission, temperature: 37.2℃; pulse: 79 per minute; respiration:19 times per minute; blood pressure: 139/80 mmHg. General conditions can be, God Qing language, into the ward, physical examination cooperation. No yellowing and bleeding spots on skin and mucous membrane. There was no swelling in the superficial lymph nodes. Neck is soft and the thyroid gland is not palpable. Bilateral breast development was normal. No chest deformity, both lungs breath sounds clear, no wet and dry rales. There was no prodrome elevation, the heart rate was 79 times per minute, rhythm was homozygous, and no pathological murmurs were heard in the auscultation area of each valve. The abdomen was flat, the left lower abdomen was tender, the liver and spleen were not palpable under the ribs, and the lower limbs had no oedema. There were no abnormalities in the spine or limbs. Physiological reflexes were present, but pathological reflexes were not elicited. Examination of the department of gynaecology: vulva marriage type, visible hymen membrane margin, pointing to not into, not to explore the vagina. Triad diagnosis: pelvic palpable mass of about 15*12*10 cm, hard, poor mobility, tenderness, tenderness in the left adnexal area, no obvious tenderness in the right adnexal area. The rest of the tests showed no obvious abnormalities.

Preliminary diagnosis

1. Abdominal pain cause to be investigated (pelvic inflammatory disease? Ovarian cyst pedicle torsion?)
2. Genital atresia (vaginal atresia?)
3. Pelvic mass properties to be investigated
4. Pneumonia.

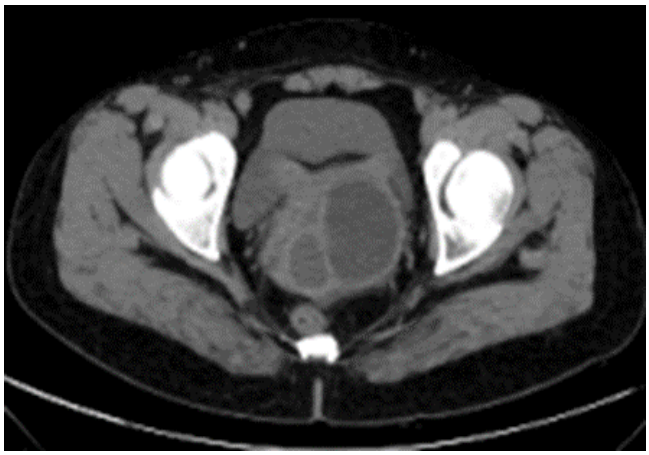

Fig. 1: Total abdominal CT

Diagnosis and Treatment Process- Emergency "exploratory laparotomy" was performed immediately after admission. The left ovary was significantly enlarged during the operation, forming a mass of about 11*10*8 cm, purple-brown with a 540° twist at the root. The left oviduct was congested, edema and thickened, with a diameter of about 1.2 cm. Two primordial uteri of about 3*2*1 cm (Fig. 2 and 3) were seen in the pelvic cavity, with smooth surfaces. The lower part of the primordial uterus on both sides was connected by an elongated tube with a diameter of about 0.3 cm. A cystic cervix of about 1*1*0.5 cm was formed at the junction, and the upper vaginal atresia could be explored downward. Intraoperative B-mode ultrasound was used to explore the bilateral initial uterus (Fig. 4 & 5), and solid muscularity echoes like normal uterine echoes could be detected, but uterine body and cervical structure were not smooth, and uterine cavity and endometrial echoes were not seen. The appearance of the right ovary and right fallopian tube was normal. The left adnexa were removed and frozen intraoperatively. The results showed a benign cyst in the left adnexa. Postoperative pathology examination showed (Fig. 6) serous cystadenoma with extensive cyst wall haemorrhage (left ovary)—ipsilateral oviduct acute, chronic inflammation with hemorrhagic infarction.

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Postoperative diagnosis

1. Torsion of pedicle of left ovarian benign cyst;
1. Double initial-base uterus;
2. Congenital upper vaginal atresia;
3. Pneumonia.

Symptomatic and supportive treatment, such as fluid replacement, was given after the operation, and the patient's pain symptoms were significantly relieved. During hospitalization, peripheral blood samples were collected for genetic testing. Karyotype analysis showed that 46, XX,1qh+ (GTG); Conclusion: Chromosome G banding increased the length of the easy chromatin region on the long arm of chromosome 1, suggesting genetic counseling (Fig. 2 & 3).

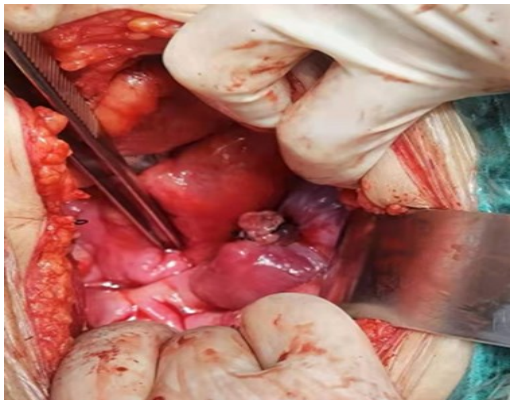

Fig. 2: Left uterus

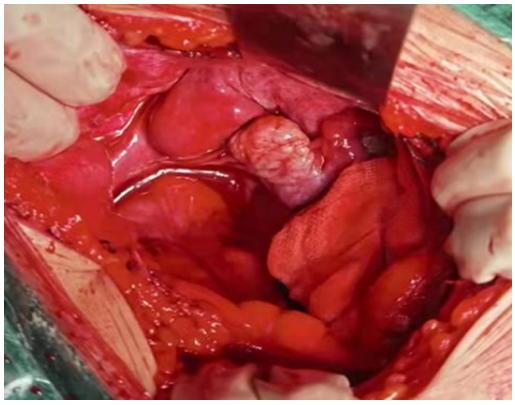

Fig. 3: Right uterus

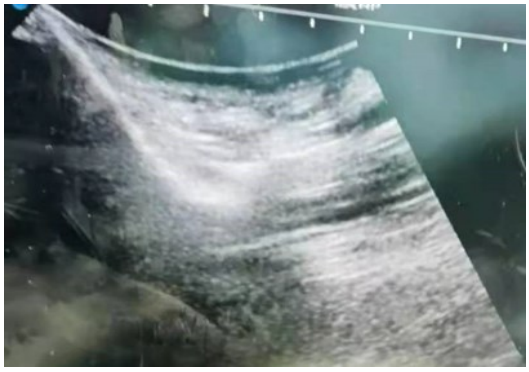

Fig. 4: Left uterus

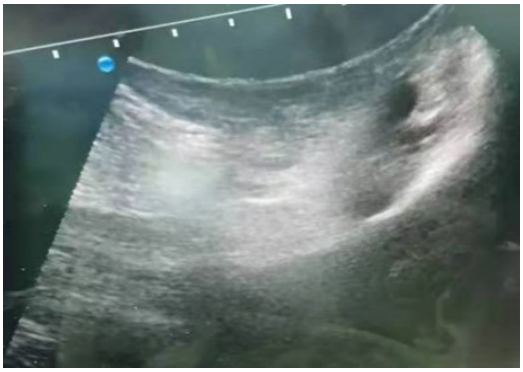

Fig. 5: Right uterus

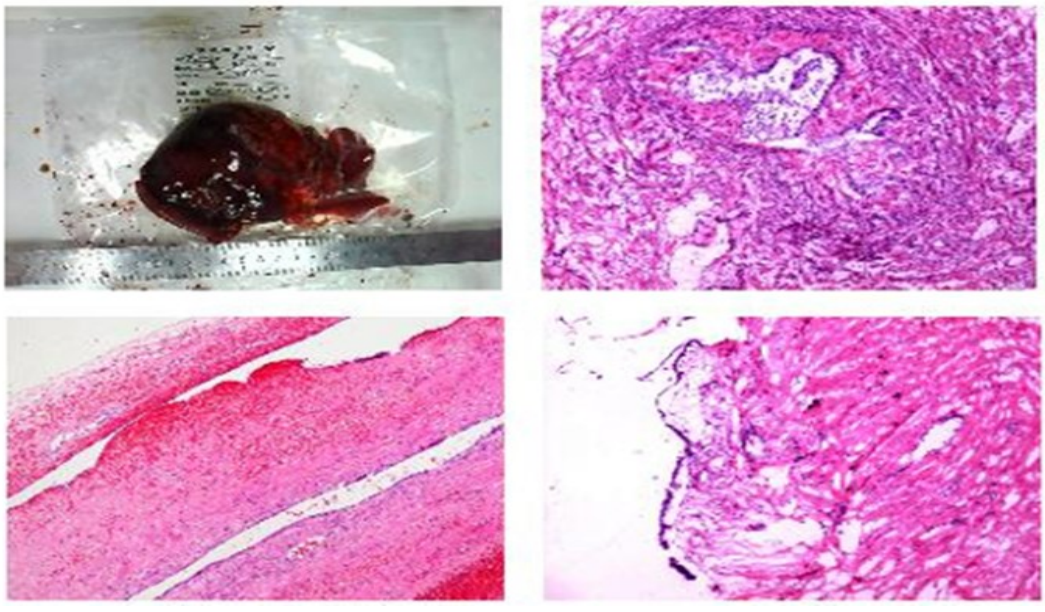

Fig. 6: Postoperative examination (left ovary)

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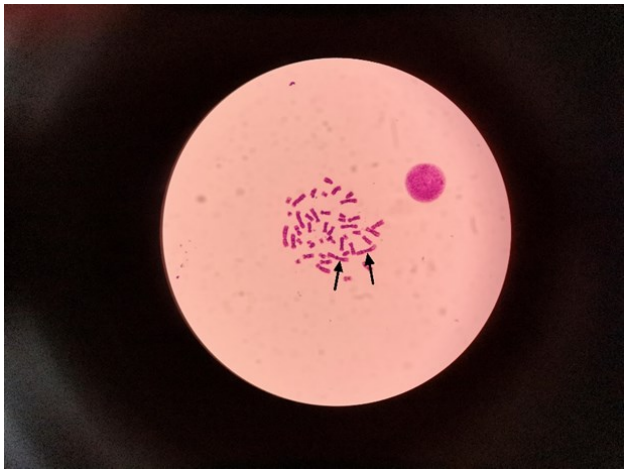

Fig. 7: Karyotype of peripheral blood, black arrow shows thickened chromosome 1

DISCUSSION

The development of female genitalia is formed in the embryonic stage, and the fallopian tube, uterus and vagina are developed from the accessory mesonephric duct [1]. The accessory mesonephric duct is divided into head, middle and tail segments. At the 10th week of the embryo, the middle and caudal end turn inward and downward, meet and fuse with the contralateral side in the midline to form the uterus and cervix, and the caudal end reaches the dorsal side of the urinary reproductive sinus [1]. The unfused head develops into the fallopian tube [1]. At 12 weeks, the septum between the two accessory mesonephric tubes fused to form a single lumen, which developed into the uterine and vaginal upper segments [1]. If bilateral accessory mesonephric duct dysplasia or bilateral accessory mesonephric duct caudal dysplasia is called MRKH syndrome (Mayer-Rokitansky-Kuster-Hauser syndrome). The main manifestations are the absence of the uterus or only the initial uterus and the congenital absence of the vagina, with an incidence of about 1/4000-1/5000 [2,3]. The main diagnostic point was the normal development of female secondary sexual characteristics. Sexual life is normal, but there may be genital obstruction, showing no vaginal opening or only in the posterior vestibule to see a shallow concave, occasionally shallow blind vaginal end. It can be accompanied by other organ malformations and abnormalities, especially urinary system malformations [4]. Some were associated with spinal abnormalities. The chromosome karyotype was 46XX, and the blood endocrine examination was normal female level [2,3].

At present, MRKH syndrome is mainly divided into three types in the world. Type 1 is the most common, which only shows no uterus or vagina, and the development of fallopian tubes, ovaries and other organ systems is normal. Type 2 with ovarian or urinary malformation; Type 3 is also called MURCS syndrome (Mullerian duct aplasia, renal aplasia, and cervicothoracic somite dysplasia) in addition to the reproductive and urinary system malformations, also with cervical thoracic segment of malformations. These include cardiac malformations and muscle weakness, as well as skeletal system abnormalities [4].

This case is a case of double initial uterine and upper vaginal atresia (MRKH syndrome type 1) with left ovarian cyst pedicle torsion. The patient had persistent left lower abdominal pain, which was suspected to be ovarian cyst pedicle torsion. After laparotomy, it was confirmed that the pedicle of the ovarian cyst was twisted and necrotic. The larger the torsion Angle and the longer the time, the more serious the attachment ischemia and the more severe the abdominal pain. Because the patient has a double initial uterus, the support force around the annex is weakened, the center of gravity is easy to shift, and torsion can quickly occur when the position is changed. During the operation, the initial uterus on both sides of the pelvic cavity had a smooth surface. The lower part of the bilateral initial uterus was connected by a slender tube with a diameter of about 0.3 cm. A cystic cervix with a size of about 1*1*0.5 cm was formed at the junction, and the upper vaginal atresia could be explored downward. In addition, B-mode ultrasound showed muscle solid nodules of bilateral initial uterus without endometrium, and the patient had not menstruated and had no periodic abdominal pain. Combined with the patient's clinical manifestations, gynaecological examination, and intraoperative findings, it is precisely because the patient's bilateral accessory mesonephric duct caudal dysplasia in the embryonic development period, which shows the characteristics of MRKH syndrome type 1. Considering the patient's age and lesion condition, bilateral uterus and right adnexa were retained, and only "left adnexa resection" was performed without "vaginoplasty".

Past sexual differentiation and developmental abnormalities were classified into true hermaphroditism, female pseudo hermaphroditism and male pseudo-hermaphroditism [5]. According to the research of Ge

Qinsheng et al., human sex can be divided into ① Sex chromosome sex: normal male is XY, normal female is XX; ② Gonadal sex: normal male testis, female ovary; ③ Internal and external genital gender: male vas deferens, epididymis, seminal vesicle, prostate, penis and scrotum; Women for the fallopian tube, uterus, cervix, vagina, size labia and clitoris; (4) Sex hormone gender: androgens enable male sexual development and maintain male sexual function; estrogen enables female sex development and maintains female characteristics; ⑤ Gender: when an individual lives in society as a male or female since childhood, it is called social gender; ⑥ Psychological gender: people's personality, hobbies, behaviours, thoughts, sexual desire, etc. conform to male or female psychological gender [6].

Many factors influence sexual development of the reproductive tract. Normal sexual development depends on the synergistic coordination of activators and repressors interacting in a precise spatiotemporal pattern [7]. In this process, if abnormal, the above six gender can cause a variety of inconsistencies. The fertilized egg determines chromosome sex: XY chromosome develops into the testis, XX chromosome develops into ovary [6,8]. The abnormal number or structure of sex chromosomes results in the increase, deletion, or position change of genes so that the genes related to sexual differentiation cannot be expressed in an orderly and coordinated manner, which affects the gonadal development and the formation of sexual characteristics, resulting in abnormal sexual development (ASD) [9,10].

The sex chromosome of this patient is XX, and the normal number is 46, showing that the patient is normal female. The female gonad and internal and external genitalia are differentiated, and the sex hormone level is normal, which maintains the second female characteristic, which is in line with the gender classification of people in the study of Ge Qinsheng *et al.* [6]. We agree with Zhang *et al.* [11] study; in patients with autosomal chromosome (1), there may be some of the genes involved in development in [10-12], chromatin accessibility to chromosome 1 long arm length increases, eradicating the structure of the chromosomes, affecting its gene expression, it affected by the gene function is related to the development of the uterus. As a result, patients have clinical manifestations of abnormal uterine development [12,13]. Whether genes related to gonad development exist on autosomes and the relationship between these genes and abnormal uterine development need further study [13,14].

CONCLUSIONS

MRKH syndrome is a dysplasia congenital reproductive system disease. Key factor and one of the essential reasons are chromosomal abnormalities, so patients with clinically diagnosed uterine dysplasia cytogenetics inspections should be early, so early clear etiology, giving patients the best treatment plan. In addition, according to different clinical manifestations and different types of patients, various treatment programs should be adopted to restore the normal physiological function of patients and improve their quality of life.

CONTRIBUTION OF AUTHORS

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Research design- Wenkang, Tao
Supervision- Jifang Shi
Materials- Wenkang, Tao
Data collection- Wenkang, Tao
Literature search- Wenkang, Tao
Writing article- Wenkang, Tao
Critical review- Jifang Shi
Article editing- Wenkang, Tao
Final approval- Jifang Shi

REFERENCES

[1] Zhao D. A case report of ultrasound diagnosis of congenital bilateral basal uterus. J Practical Gynecol Endocrinol., 2018; 5(10): 73-74.
[2] Huang H, Yang Q. Three cases of congenital absence of vagina and basal uterus. J Gannan Med Col, 2007; (04): 638-39.
[3] Xie X, Gou W. Obstetrics and Gynecology. 8th edition. People's Medical Publishing House, 2013; 2: 1-8.
[4] Zhou H, Zhu L. Diagnostic characteristics and clinical management of MRKH syndrome. Chinese Department Family Planning Obstetrics Gynecol., 2017; 9(9): 12-14.
[5] Tian Q, Ge Q. New classification of sexual differentiation and developmental abnormalities. Chinese J Pract Gynecol Obstetrics, 2001; 4: 7-9.
[6] Ge Q, Gu C, Ye L, Lin S, He F, Yu Q. Recommend a classification of sexual development abnormalities. J Reprod Med., 1994; 2(03): 131-34.

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DOI: 10.21276/SSR-IIJLS.2023.9.6.11

[7] Selma FW. Disorders of sex development. Best practice & research. Clin Obst Gynaecol., 2018; 48: 90-102

[8] Liu P, Wu Q, Shi H. Research progress on the genetic causes of 46, XY female sexual development abnormalities. Int J Reprod Health/ Family Planning, 2017; 36(06): 492-97.

[9] Li J, Huang Y, Yang X, Bi L, Mei X, et al. Analysis of genetic characteristics and clinical phenotypes of patients with sexual development abnormalities. Chinese J Health Inspection, 2021; 31(02): 191-94.

[10]Huang J, Zhang Y, Zhang X, Li Y. Cytogenetic analysis of patients with primary amenorrhea. Tianjin Med., 2007 (10): 770-71.

[11]Zhang Y, Wu L, Li X, Hu J. Cytogenetic analysis of patients with uterine dysplasia. Chinese J Prenatal Diagn., 2019; 11(04): 63-66.

[12]Luo G, Pan H. Treatment and effects of MRKH syndrome. J Practical Obstetrics Gynecol., 2018; 34(9): 648-50.

[13]Patton PE, Novy MJ, Lee DM, Hickok LR. The diagnosis and reproductive outcome after surgical treatment of the complete septate uterus, duplicated cervix and vaginal septum. Am J Obstet Gynecol., 2004; 190(6): 1669-75.

[14]Huang H, Yang Q. Three cases of congenital absence of vagina and basal uterus. J Gannan Med College, 2007; (04): 638-39.

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