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Abdominopelvic Pain in Patient with Uterus Didelphys and Unilateral Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA Syndrome)

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Abstract

Introduction: Uterus didelphys with obstructed hemivagina associated with ipsilateral renal agenesis (OHVIRA syndrome) is a rare female urogenital malformation and delay in its diagnosis could lead to several complications.

Case presentation: A 21-year-old virgin woman was admitted to the emergency department (ED) with severe abdominal pain, without fever and vaginal discharge. She reported a history of cyclic abdominopelvic pain and dysmenorrhea for 5 years. The primary diagnosis (OHVIRA syndrome) was made using ultrasonography, spiral computed tomography (CT) and magnetic resonance imaging (MRI). In addition, laparoscopy was performed to confirm diagnosis and drain hematosalpinx. Then, hysteroscopy was carried out for septum resection and catheter insertion. At one-month follow-up the ultrasonography showed normal left hemicavity of uterus associated with significant decrease in dysmenorrhea.

Conclusion: Being aware of OHVIRA syndrome and clinical suspicion of this rare anomaly are essential for making a timely diagnosis, preventing complications, relieving symptoms, and preserving future fertility.

Key words: Abdominal Pain; Dysmenorrhea; Gynatresia; OHVIRA Syndrome

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INTRODUCTION

Uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome is a rare complex female urogenital malformation. It is classified as Mullerian dysgenesis class III (1). The total prevalence of Mullerian dysgenesis is 5% (2). This syndrome presents with a wide variety of clinical symptoms, and having information about this anomaly and strong doubt are important for accurate diagnosis. Difficulty in diagnosis, owing to infrequency of the syndrome and need for specific imaging, may delay the diagnosis. Complications due to delayed diagnosis are frequent (3). Here, in this case report, we presented a 21-year-old virgin woman who had OHVIRA syndrome.

CASE PRESENTATION

A 21-year-old virgin woman was admitted to the emergency department (ED) due to abdominal pain during menstruation period since 3 days before. She did not present with any fever or any vaginal discharge. She reported a history of cyclic abdominopelvic pain and dysmenorrhea since she was 16 years old. She had not been admitted for reconstructive surgery. An abdominopelvic MRI had been performed 5 years ago, which had shown double uterus with left hematocolpometra and left renal agenesis (Figure 1).

On general examination, the patient was conscious, secondary sexual characters were normal, pulse rate was 96 beats/minute, blood pressure was measured 100/60 mmHg with negative orthostatic hypotension. Physical examination revealed a cystic mass arising from the pelvis and there was left lower abdomen tenderness without rebound or guarding.

Ultrasonography of her abdomen and pelvis showed left ipsilateral renal agenesis and uterus didelphys with obstructed hemivagina leading to hematocolpometra in the left hemicavity. The right uterus was normal with 8.7 mm endometrial thickness. The left endometrial cavity was distended with fluid containing low-level internal echoes with a diameter of 128 mm up to the distal part of pelvic. A large tubular cystic mass, suggestive of possible hematosalpinx, was seen adjacent to the left uterus. Both ovaries were normal.

After receiving gynecology consultation, the

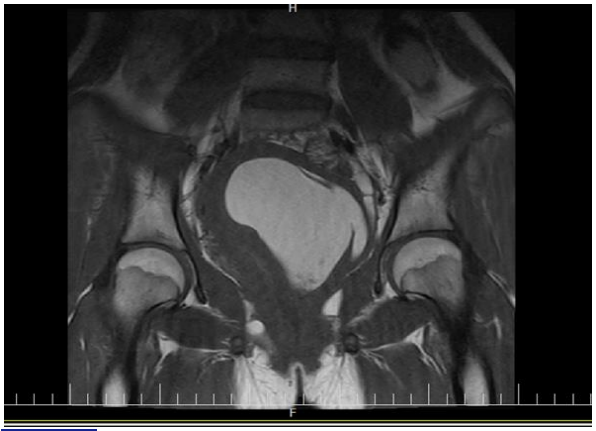

Figure 1: The coronal cut of the abdominopelvic magnetic resonance imaging (MRI) that had been performed 5 years before admissin showed double uterus with left hematocolpometra and left renal agenesis.

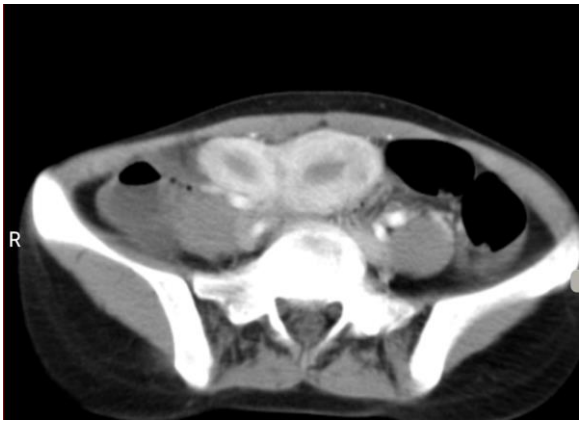

Figure 2: The axial cut of spiral computed tomography (CT) scan showed left ipsilateral renal agenesis and uterus didelphys with obstructed hemivagina leading to hematocolpometra in the left hemicavity.

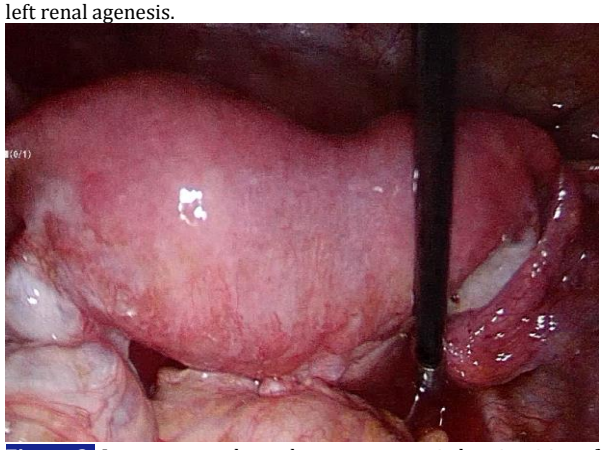

Figure 3: Laparoscopy showed two asymmetric hemicavities of didelphys uterus and left hematosalpinx

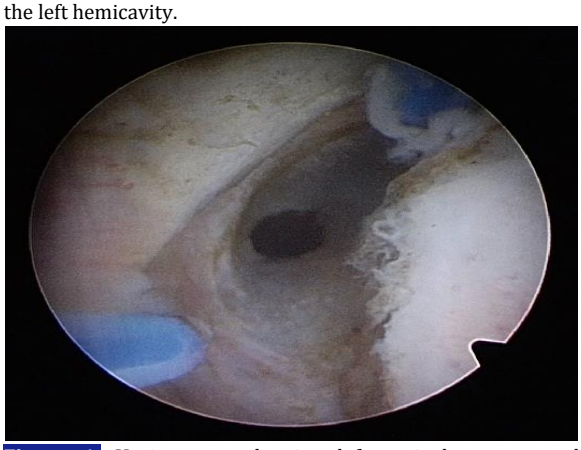

Figure 4: Vaginoscopy showing left vaginal septum and resection implemented with bipolar resectoscopy at fistulation site.

patient underwent preoperative evaluation and was admitted for surgical intervention. Another investigation performed for surgical planning was computed tomography (CT) scan, which confirmed the findings of ultrasonography (Figure 2).

The night before surgery, she reported a massive chocolate vaginal bleeding followed by decrease in abdominal pain, perhaps due to spontaneous drainage of hematoma. Then patient underwent reconstructive surgery under general anesthesia. Firstly, cystoscopy was performed to place right ureteral stent. Secondly, vaginoscopy and hysteroscopy of right hemicavity was carried out, which showed a small tubular cavity and a bulge in the left hemivagina. Thirdly, laparoscopy was implemented to confirm primary diagnosis and drain the hematosalpinx. Fourthly, septum resection was implemented with bipolar resectoscopy at fistulation site. Fifthly, a Foley catheter was inserted in the hemicavity and was left in place for a week to drain the blood (Figures 3 and 4).

At one-month follow-up, ultrasonography showed that the left sided uterus had returned to its normal size. The patient reported lower dysmenorrhea and was satisfied. She was advised follow-up visits for further management.

Discussion

OHVIRA syndrome presents with a wide variety of clinical symptoms and the most common one is dysmenorrhea, followed by non-specific complaints such as acute abdominopelvic or vaginal pain, urinary retention, dysuria, vaginal discharge, and infertility (4-6).

Previous studies have reported that in comparison to more frequent congenital anomalies of the female genital tract, such as imperforate hymen or vaginal atresia, OHVIRA syndrome is more often missed, as menses usually appear to be normal since there is regular menstrual flow from the unobstructed hemivagina. Associated

complications due to delay in diagnosis include infection, pelvic adhesions, endometriosis, infertility and chronic cryptomenorrhea (3, 5-7). Rarely, the obstructed vagina may spontaneously become perforated and lead to pyocolpos and pelvic inflammatory disease, and even in some situations, septic shock (1). Fortunately, in our case, although she had the syndrome for almost 5 years, she had not developed severe complications and only had abdominopelvic pain. It is likely that that fallopian tube obstruction prevented retrograde menstruation and endometriosis as complications. Spontaneous drainage of trapped hematoma and fluid in to the vagina can be another cause for absence of complications and delayed diagnosis as reported in a similar case (8).

Obstructive genital tract anomaly should be considered as a differential diagnosis in adolescent girls if they present with unilateral kidney agenesis accompanied by non-specific abdominal symptoms (9). Imaging studies such as ultrasonography and magnetic resonance imaging (MRI) are very useful in diagnosis and treatment of this syndrome. The case was diagnosed using abdominal ultrasonography and was confirmed via abdominopelvic MRI. Ultrasonography, with 90-92% diagnosis accuracy, has been deemed the gold standard for diagnosis and surgical planning (10). In addition, ultrasound device, as a laparoscopic probe intraoperative device, can show the origin of the pelvic mass, guide the surgeon in drainage of the hematocolpus and recognize the two different hemicavities during the hysteroscopic procedure (11). Using MRI can also help in finding more information about vaginal channels' continuity (obstructed/not obstructed) and uterine morphology (uterine horns disposition) (12).

previous studies reported that gold standard investigation for diagnosis and treatment of this complication is laparoscopy, specifically when MRI is not available or does not provide clear diagnosis (4, 5). Other suggested surgical techniques include simple vaginal incision, septoplasty, complete vaginal septum resection, hemihysterectomy, and puncture of the blood via infusion of saline through septum (1, 3, 4, 13, 14). Our case underwent laparoscopy and hysteroscopy for diagnosis and resectoscopy for septum resection. In this case, the advantage of resectoscopy was that it did not affect the integrity of the hymen. We think that this technique is associated with safe surgical outcome, does not cause intraoperative complications, and saves the integrity of hymen.

Conclusions

We believe that emergency specialists and internists must be aware of the obstructive uterine syndrome. Clinical suspicion of this rare anomaly is essential for making a timely diagnosis, preventing complications, relieving symptoms, and preserving future fertility.

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Authors' contribution

All the authors fulfil the criteria of authorship based on the recommendations of the International Committee of Medical Journal Editors (ICMJE).

Conflict of interest

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