Case Report

Challenges in Diagnosing Obstructed Hemivagina and Ipsilateral Renal Anomaly Syndrome: Case Report of a Rare Condition

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Abstract

Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome is a very rare type of Müllerian duct anomaly. Cases of OHVIRA syndrome have varied clinical manifestations. We report the case of a 15-year-old virgin with endometrial cyst and OHVIRA syndrome. Magnetic resonance imaging confirmed the diagnosis of OHVIRA syndrome. A salpingo-oophorectomy laparotomy was performed, but the patient refused to perform a septotomy or vaginal septal resection. We compared the cases of OHVIRA syndrome that we found with several other reports. The complaints of patients with OHVIRA syndrome may vary from one another, with different histories. Imaging investigations can confirm the diagnosis. The main treatment in OHVIRA syndrome is intended to alleviate symptoms that appear.

Keywords: *Herlyn–Werner–Wunderlich syndrome, Müllerian duct anomaly, obstructed hemivagina and ipsilateral renal anomaly syndrome, rare*

Introduction

Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome or Herlyn–Werner–Wunderlich (HWW) syndrome is a very rare congenital anomaly of the female urogenital tract. OHVIRA syndrome is a variant of the Müllerian duct anomaly, which has a triad of uterine didelphys, hemivaginal obstruction and ipsilateral renal agenesis.[1]

Müllerian duct anomalies occur in 0.5%–6.7% of the total population, and the incidence of OHVIRA syndrome ranges from 2% to 3% of all Müllerian duct anomalies.[1,2] Although there is a typical triad of OHVIRA syndrome, the clinical manifestations of this syndrome are highly variable.[3]

Case Report

In this case report, we discuss a rare case of OHVIRA syndrome in a 15-year-old girl with the main complaint of an enlarged abdomen. These complaints were also accompanied by pain, particularly during menstruation. The pain was more intense a few days before admission. Physical examination revealed a cystic mass as high as two fingers above the umbilicus with a well-demarcated, smooth surface and no tenderness. Transabdominal ultrasound examination detected that the uterus was didelphys and that there was a cystic mass measured ± 16 cm × 10 cm with firm boundaries, unilocular and no solid area that originated from the adnexa. Further examination of the tumour marker Ca-125 showed a value of 362 and a Risk Malignancy Index value of 1086. Other pre-operative evaluations are within the normal range. We diagnosed the patient with a suspected malignant cystic ovary neoplasm and prepared for a Veriscope laparotomy (frozen section).

Intraoperative findings revealed a cystic mass measuring ± 20 × 15 cm that originated from the right ovary [Figure 1].

We decided to perform a right salpingo-oophorectomy, and the VC result showed a haemorrhagic benign cyst.

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Further exploration found that didelphys uterus, normal left ovary and right kidney were absent [Figure 2].

Post-operative recovery was uneventful, and she was discharged 3 days after the surgery. Histopathology examination (paraffin block) of the mass showed a right ovarian endometrial cyst. Postoperatively, a magnetic resonance imaging (MRI) scan of the abdomen and pelvis was performed [Figure 3]. It revealed the structure of the cervical duplex, and uterine duplex was accompanied by right hemivaginal obstruction and right renal agenesis. These findings confirmed the diagnosis of HWW syndrome, also known as OHVIRA syndrome.

Since the primary treatment of OHVIRA syndrome is to relieve the obstruction,[4] we suggested performing septotomy, but the patient and her family refused to perform the procedure because the patient was still a virgin. The patient had no complaints related to the previous condition at least until 6 months of observation.

Discussion

OHVIRA syndrome is a very rare type of Müllerian duct anomaly.[1] This syndrome, also known as HWW syndrome, was first reported in 1922 with a collection of typical symptoms of uterine abnormalities (uterus didelphys or uterine septate) and abnormalities kidney (renal agenesis, multicystic dysplastic kidney and renal duplication).[4] This condition results from the failure of the two Müllerian tubes to fuse by the 10th week of gestation.[5]

In this discussion, we compare our case with other cases of OHVIRA syndrome reported by Mabuchi *et al.* (Japan), Malarazhagan *et al.* (Australia), Zeba *et al.* (Bangladesh), Pillai *et al.* (Qatar) and Al Ghafri *et al.* (Saudi Arabia, 2018).[1,3-6]

From these case reports, we found that they ranged from 13 to 17 years old when sought treatment, except for one case reported by Mabuchi *et al.* (25 years).[6] In accordance with the case we reported, the patient was 15 years old.

Patients may present with moderate-to-severe complaints (emergency services) to mild complaints (outpatient services). The complaints vary from primary amenorrhoea, foul-smelling vaginal discharge, abdominal-pelvic masses, to severe abdominal pain and urinary retention. In the case presented above, the patient came to seek treatment in an outpatient service with complaints of a mass in the abdomen accompanied by pain. The menstrual history in the five cases was also different, there were cases with primary amenorrhoea and, in other cases, they had regular menstruation. Similarly, the interval between menarche and chief complaints ranges from a matter of months to a dozen years after menarche. In the case we reported, the interval between menarche until symptoms appear and coming for treatment is 2 years.

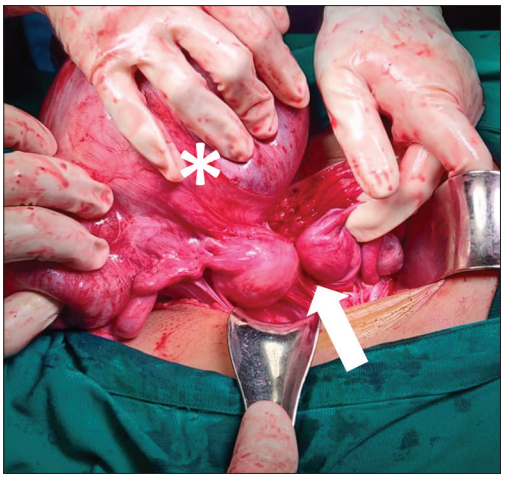

Figure 1: Intraoperative findings. Right ovarian cyst (*) and uterine didelphys (arrow)

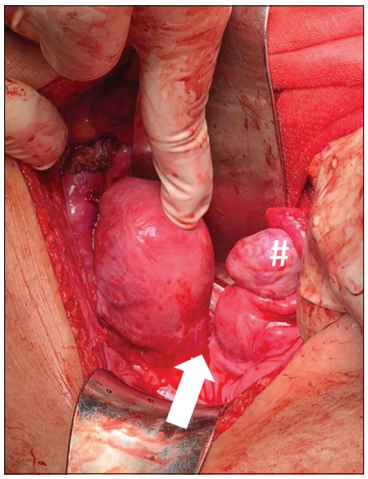

Figure 2: Post-right salpingo-oophorectomy. Uterus didelphys (arrow). Normal left ovary (#)

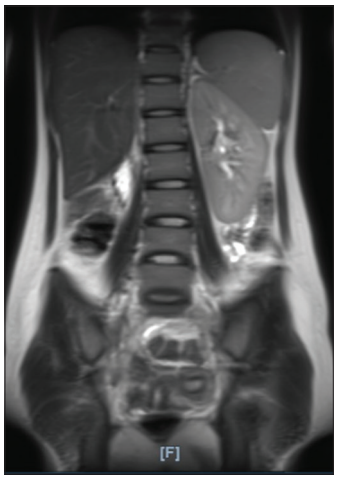

Figure 3: Magnetic resonance imaging revealed right renal agenesis, uterine and cervical duplex and right hemivaginal obstruction

On physical examination, generally, the patient is hemodynamically stable, and if it is possible to do an examination (married/coitarche), there is a bulge in the vagina on the side of the abnormality. Supportive imaging performed on the patient can confirm this OHVIRA syndrome. Ultrasound examination of the uterus didelphys and absence of unilateral kidney with normal or slightly enlarged contralateral kidney (compensatory hypertrophy) may be found. Ultrasound examination is used as an initial examination because of its wider availability and relatively lower cost.[4] Even though abnormalities were found on ultrasound, all cases reported were still confirmed with computed tomography scan or MRI modalities. Before imaging studies, OHVIRA syndrome may have gone undiagnosed and may mimic other gynaecological disorders such as pelvic inflammatory disease, endometriosis or endometrial cysts. In the case we reported, the patient was diagnosed early with an ovarian cyst, and therefore, we decided to perform a Veriscope laparotomy.

Although the complaints that arise in patients with OHVIRA syndrome vary, the majority of these complaints are the result of hemivaginal obstruction. Therefore, the main treatment in cases of OHVIRA syndrome is a septotomy. In the case we presented, she complained of an abdominal mass, and with the salpingo-oophorectomy procedure, the complaint subsided. After deep counselling, the patient and family refused to perform a septotomy because the patient was still a virgin.

Huseyin *et al.* reported a case of OHVIRA syndrome that was complicated by pyometra and managed by a laparoscopic hemihysterectomy procedure.[7] This option should be considered regarding OHVIRA syndrome treatment, especially in a virgin patient.

Conclusion

This case report presents a 15-year-old virgin female patient with an intra-abdominal mass. A salpingo-oophorectomy procedure was performed to remove the mass with the histopathology result of an endometrial cyst. The result of the MRI confirmed the diagnosis of OHVIRA syndrome. As an optimal management, the patient has been recommended a septotomy of the vaginal septum.

Patients with OHVIRA syndrome may present with different complaints. Although there is a triad of OHVIRA syndrome, the main complaints that make patients seek treatment vary from primary amenorrhoea, foul-smelling vaginal discharge, abdominal-pelvic mass, to severe abdominal pain and urinary retention. Patient profiles (age, menarche status and coitarche) at the time they came for treatment were different. Careful imaging studies can confirm the diagnosis of OHVIRA syndrome. The management of OHVIRA syndrome is aimed at overcoming the complaints that arise.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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