Case report

Hematocolpos in women with uterus didelphys, imperforate hemivagina and ipsilateral renal agenesis: A case report and review of the literature

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ABSTRACT

Introduction and importance: Hematocolpos can be part of a rare syndrome OHVIRA or Herlyn-Werner-Wunderlich syndrome that requires special management (Brousse et al., n.d. [1]). The diagnosis can be delayed, yet an early diagnosis is important in order to elaborate an adequate strategy management and prevent the patient from complications.

Case presentation: We report the case of a 14-year-old girl suffering from an hematocolpos due to this syndrome who has underwent a surgery in our hospital.

Clinical discussion: Hematocolpos, the main symptom, is due to a retention of retained menstruation blood in women with obstructed hemivagina. Therefore, it’s painful and associated with increasing dysmenorrhea (Brousse et al., n.d. [1]). The main treatment consists in its drainage with resection of vagina septum, preventing the patient from complications like endometriosis, infection, and infertility.

Conclusion: A good knowledge of this anomaly can lead to an adequate diagnosis and more importantly an adequate treatment based on surgery to prevent the bad outcomes.

1. Introduction

Hematocolpos is due to a retention of retained menstruation blood in women with obstructed hemivagina. It represents the main symptom. Therefore, it’s painful and associated with increasing dysmenorrhea [1]. Obstructed vagina, uterus didelphys and homolateral renal agenesis are part of a syndrome called obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) or Herlyn-Werner-Wunderlich syndrome [1–5]. We report the case of a 14-year-old girl suffering from a hematocolpos. Due to the rarity of the disease, the diagnosis isn’t well-known and can then be delayed, yet an early diagnosis is important in order to elaborate an adequate strategy management and prevent the patient from complications like endometriosis and infection.

2. Case report

A 14-year-old girl from low socio-economic level, presented by herself to the emergency department, with complaints of a massive vaginal bleeding and pelvic abdominal pain. Menarche had occurred 3 months before the consultation and had regular menstrual cycles (every 28 days). Moreover, there is no familial or personal medical history such as genetic abnormalities, similar symptoms, or drug history.

Abdominal examination showed a pelvic palpable mass. Laboratory tests revealed a mild anemia with a negative pregnancy test. The ultrasound demonstrated a hematosalpinx without peritoneal effusion and a cystic mass. It also showed the absence of the right kidney. The uterus and the ovaries appeared to be normal. The diagnosis was not made at that point. Magnetic resonance imaging (MRI) revealed a uterus didelphys bicornus bicervical with a unilateral right renal agenesis, a hematocolpos and hematosalpinx, leading to the diagnoses of OHVIRA, or Herlyn-Werner-Wunderlich syndrome (Figs. 1-3). A surgery under general anesthesia was then performed in order to relieve the patient and treat the cause by the chief of surgery. Before admitting the patient in the OR, a transfusion was necessary. Antibiotics were given during the surgery and continued for one week. The first step consists in seeing the hematocolpos and draining it. In the second step, we had to proceed carefully to the resection of the hemi-vaginal septal. A foley catheter was left in the uterus for 5 days. No infection or any other complication was described post-operatively. The follow-up in many consultations didn't notice any problem.

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3. Discussion

The obstructed vagina is a congenital anomaly of the Mullerian ducts (paramesonephric) and Wolffian structures (mesonephric) [2–4], consequence of a urogenital absence of fusion of these ducts. It's responsible for a double uterus more or less complete depending on the height of the defect: uterus didelphys (type III, American Society for Reproductive Medicine classification) or rarely a complete septate uterus (type V) [1]. We report a case report of this malformation following the recent scare guidelines [6].

This genital malformation is always associated with homolateral renal agenesis [1,4] because the reproductive system develops close to the urinary tract and kidneys from the same structure mesoderm [4]. The association-obstructed hemivagina/homolateral renal aplasia and the syndrome were respectively described for the first time in 1922 and 1950 [4,5].

Obstructed hemivagina is a rare disease. It happened in 0.1 % in all women if including hymenal imperforation and vaginal septum and 12 % if including women with congenital malformation of the muller duct [1]. The prevalence of congenital Mullerian duct anomalies is about 1 % [4].

The diagnosis is usually made right after the first menarche. Indeed, this syndrome leads to the formation of a hematocolpos [5]. Hematocolpos causes pelvic abdominal pain, acute dysmenorrhea [1], pelvic or vaginal mass or acute retention of urine [2]. We can also have vaginal discharge or intermenstrual bleeding when there is a communication between the two sides [2]. This patient could also suffer from vomiting or fever, infertility, endometriosis or complication during pregnancy and labor [4]. The diagnosis can be delayed. Indeed, the rarity of the pathology and the menstrual flow that comes from the patent unobstructed hemivagina gives the appearance of normal menses [4]. The most common clinical presentation is that of pelvic pain shortly after

![Fig. 1. An MRI showing a uterus didelphys.](image1)

![Fig. 2. An MRI picture showing the aplasia of the right kidney.](image2)
menarche in association with a vaginal or pelvic mass, and normal menstrual periods [5]. Some cases could be diagnosed during adulthood. Indeed, in case of communication between uterine horns, or when the presence of a hypofunctional horn on the side of obstructed hemivagina, the patient may consult lately for hematocolpos [1]. The diagnosis can then be misunderstood, yet an early diagnosis is important in order to elaborate an adequate strategy management [4]. An early management is also primordial because it allows preventing the patient from complications like endometriosis and infection [4,5].

Ultrasonography and MRI are used for the diagnosis. Even if the ultrasonography is more available, the diagnosis may be complicated but not impossible. It may show a pelvic mass and/or the presence of the vaginal septum. The 3D ultrasound is more effective in the diagnosis of uterine malformation [4]. However, the vaginal septum is difficult to visualize on ultrasound and is best shown on MRI [5]. The MRI is more sensitive with 100 % accuracy. It shows the shape of the intrauterine cavity, and the character of the septum [5]. It can also look for other signs like the hematocolpos infection, hematometry, or signs of superficial endometriosis and/or deep pelvic infiltrating due to a peritoneal reflux of menstruation, present in 23 % of cases of uterine malformation [1].

The recommended management approach for an obstructed hemivagina is to proceed with a septum resection [3]. The hematocolpos drainage without septum resection may cause fibrosis, the vagina stenosis, and the recurrence of the same symptoms [1]. It’s a vaginal surgical procedure under general anesthesia or spinal anesthesia according to the patient’s choice. The first step is the hematocolpos drainage. Vaginal exposure is ensured by anterior and posterior vaginal valves. The hematocolpos is then incised by a scalpel allowing the spontaneous evacuation of a chocolate-colored viscous liquid corresponding to old blood. An aspiration and an oxytocin infusion may be helpful. The second step is the vaginoplasty. It consists in looking for the second cervix. Indeed, after the hematocolpos evacuation, the initially hidden cervix is then visually spotted. Finger exploration of the “vaginal pocket” allows the evaluation of the thickness and dimensions of the vaginal septum. Once the two cervixes have been identified, the entire vaginal septum can be safely resected. At the end of the procedure, the two cervical orifices are visible on the vagina. Once both cervixes are identified, a hysteroscopy can be performed to look for the absence of an isthmic communication [1]. Moreover, we can suspect an infection of the hematocolpos if there is a fever with acute pain. Then, a laparoscopy should also be used [2].

Postoperatively, we can have stenosis, recurrence of hematometry or infection. A second operation can be processed. Some techniques can lower those complications like the use of vaginal molds, dilators and coated tracheobronchial stent [4].

One case of Mirena levonorgestrel intrauterine system insertion had been described in the literature in order to suppress the bleeding discomfort in the patient although a distorted uterine cavity is still a category 4 contraindication. King et al. insist that it should only be done in research and with specific criteria [3].

In conclusion, due to the rarity of the hematocolpos in didelphys uterus, it’s not well known. Therefore, the diagnosis and the management of the disease could be difficult. Surgery represents the main treatment. It consists in the evacuation of the hematocolpos and more importantly the vaginoplasty in order to improve the outcomes.

**Patient perspective**

The patient reports to be satisfied by the procedure even with her younger age. She was suffering before the surgery from anxiety because of the brutal hematometry and feels safer now.

**Sources of findings**

There are no funding sources to be declared.

**Ethical approval**

Ethics approval has been obtained to proceed with the current study. Consent to participate not applicable.

**Consent**

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

**Author contribution**

Oumaima Mhamdi made substantial contributions to conception and design, acquisition of data, analysis and interpretation of data; she has
been involved in drafting the manuscript and revising it critically for important intellectual content. Sarah Boujida and Ibtissam Benseghir made substantial contributions in the management of the patient. Aziz Baidada and Aicha Kharbach made substantial contributions to conception and design and acquisition of data; they have been involved in drafting the manuscript.

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The authors report no declarations of interest.

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