Case Report

Didelphys Uterus Associated to Ipsilateral Vaginal and Renal Agenesis in Mother of Two Kids: Unusual Case Report

Hafsa Elouazzani1*, Ydou Salah2, Fouad Zouaidia1, Ahmed Jahid1, Amine Saouli2, Kaoutar Znati1, Zakiya Bernoussi1, Najat Mahassini1

1Department of Pathology, Ibn Sina University Hospital Center Rabat, Morocco
2Department of Urology, Ibn Sina University Hospital Center Rabat, Morocco

*Corresponding author: Hafsa Elouazzani, Department of pathology, Ibn Sina University Hospital Center Rabat, Morocco. Tel: +212674556975; Email: hafoussa02@hotmail.fr


Received Date: 13 February, 2018; Accepted Date: 22 March, 2018; Published Date: 30 March, 2018

Abstract

A didelphys uterus is one of the least common of Mullerian duct anomalies that arise from abnormal embryological development of the Mullerian ducts [1]. Embryologic anomaly of one Wolffian ducts - causing unilateral renal agenesis- is additionally seen in 15-30% of uterine didelphys [2]. In this paper, we report a first presentation–in our knowledge- of didelphys uterus with complex malformations in mother of two kids. It’s about a 30-year-old Moroccan mother who successfully conceived and carried her pregnancies to term without complications. She suffered from chronic pelvic pain. The computed tomography scan revealed a left pelvic mass that was diagnosed as a mute pelvic kidney. The discovery of a didelphys uterus was fortuitous in pathological examination of the presumed nephrectomy. Indeed, we received an open cystic mass with thickened myomatous wall. Within the wall we found a rounded cavity with chocolate like contents. In histological examination, this mass corresponded to a uterine and endo-cervical wall, fallopian tube, residual ovarian parenchyma ovary and old hematosalpinx. There was no vagina. Thus, the final diagnosis was didelphys uterus, associated to ipsilateral vaginal and renal agenesis in mother.

Keywords: Mother; Renal Agenesis; Uterus Didelphys; Vaginal Agenesis

Introduction

Uterine anomalies are an uncommon Müllerian malformation with unknown etiology. Many hypotheses and theories involving genetic, environmental and pharmacologic issues have been suggested [3]. The incidence of uterine anomalies is believed to be 0.5-2.0% of reproductive-age women, with didelphic uterus accounting for approximately 10% [3]. Embryologic anomaly of one Wolffian ducts causing unilateral renal agenesis is additionally seen in 15-30% of uterine didelphys [3]. In the majority of the cases, it is associated to obstructed hemi-vagina which is referred to as Herlyn-Werner-Wunderlich syndrome [2,3]. In our paper, we report a first case - in our knowledge- of didelphys uterus, associated to ipsilateral renal agenesis and complete vaginal agenesis in mother of two kids. Through this presentation we discuss the clinic-pathologic features of this uncommon entity.

Case Report

It’s about a 30-year-old Moroccan mother of two kids, who successfully conceived and carried her pregnancies to term without complications. She attained menarche at the age of 13 years and had regular menstrual cycles. She suffered from chronic pelvic pain without any other functional sign.

General physical examination showed no abnormalities. Laboratory tests, including complete blood count and urinalysis were normal. Vaginal and speculum examination revealed a normal vaginal opning. No rudimentary hemivagina, transverse, or vertical septum was identified. In the pelvic ultrasound, the left kidney was not visualized. Computed tomography scan revealed a left pelvic mass that was diagnosed as a mute pelvic kidney. The
left renal lodge was empty with a compensatory hypertrophy of the right kidney (Figure 1). Thus, an open radical nephrectomy was arranged. The discovery of a didelphys uterus was fortuitous in pathological examination of the presumed nephrectomy.

![Figure 1: (a) Pelvic Computed tomography scan shows a solid left latero-uterine mass (11 cm in diameter) with cystic component. (b) The left renal lodge * is empty with a compensatory hypertrophy of the right kidney.](image)

In our laboratory, we received an open cystic mass with thickened myomatous wall (11 cm in diameter). Within the wall we found a rounded cavity measuring 4 cm in diameter with chocolate like contents (Figure 2).

![Figure 2: Grossly, the pelvic mass corresponds to a thickened myomatous wall of uterus (11 cm in diameter) with the presence within the wall of a rounded cavity measuring 4 cm in diameter with chocolate like contents corresponding to a hematosalpinx and a section of fallopian tube.](image)

In histological examination, this mass corresponded to a uterine and cervical wall with fallopian tube and residual ovarian parenchyma (Figure 3).

![Figure 3: Histological examination shows a uterine wall with endometrium and myometrium (a), endocervical wall (b), fallopian tube (c) and ovarian parenchyma (d) (Hematoxylin-eosin, original magnification x 10).](image)

The rounded cavity corresponded to an old hematosalpinx composed of chronic hemorrhage with hemosiderin laden macrophages, fibrosis, foci of necrotic tissue surrounded by histiocytes and areas of chronic inflammation (Figure 4).

![Figure 4: Histological examination shows also a hematosalpinx that appears as a cavity with chronic hemorrhage (a) foci of necrotic tissue *, hemosiderin laden macrophages, fibrosis, and chronic inflammation(b) (Hematoxylin-eosin, original magnification x 10).](image)

No squamous epithelium, which is normally found in transformation zone, exocervix, and vagina, was identified. Thus, the final diagnosis was a left didelphys uterus with hematosalpinx,
associated to ipsilateral vaginal and renal agenesis. The postoperative course and follow up were uneventful.

**Discussion and Conclusion**

Complex malformations of the female genital tract are rare and often incorrectly identified, inappropriately treated, and sometimes incorrectly reported [2]. The incidence of uterine anomalies is believed to be 0.5-2.0% of reproductive-age women, with didelphic uterus accounting for approximately 10% [3]. In most cases, the etiologic factors are unknown. A didelphys uterus is one of the least common of Mullerian duct anomalies that arise from abnormal embryological development of the Mullerian ducts [1]. In the embryo, the Wolffian (mesonephric) ducts and the Mullerian (paramesonephric) ducts are the two paired urogenital structures from which the internal genital organs and the lower urinary tract derive. The fallopian tubes, uterus, and the upper two-thirds of the vagina develop from the bilateral Mullerian Ducts [2]. As a result of any interruption in the development of this embryological event, different types of uterine anomalies can arise, such as agenesis, hypoplasia, unicornuate, didelphys, bicornuate, arcuate, and septate uterus.

Ipsilateral renal and Mullerian duct anomalies were produced following the destruction of the caudal portion of the mesonephros:

- On the affected side, Mullerian ducts cannot fuse, resulting in a didelphic uterus, and cannot come into contact with the urogenital sinus centrally, forming two separate vaginas with one obstructed, which is referred to as Herlyn-Werner-Wunderlich syndrome [2,3]. It is characterized by the triad of didelphys uterus, obstructed hemivagina, and ipsilateral renal agenesis. Our patient had a vaginal agenesis and not obstructed hemivagina. This presentation, would it be a form of Herlyn-Werner-Wunderlich syndrome? We did not find an answer in the literature since there is only one similar case reported [4].
- Embryologic anomaly of one Wolffian ducts may cause unilateral renal agenesis. This anomaly is additionally seen in 15-30% of uterine didelphys [2].

The clinical presentation of didelphic uterus varies in patients with uterovaginal anomalies and renal agenesis; they present at a mean age of 17 years (range, 11–38). Typically, the onset of symptoms occurs 1–2 years after menarche. Symptoms at initial presentation include lower abdominal pain, dysmenorrhea, foul and mucopurulent vaginal discharge, and irregular menstruation [5]. The literature available on the didelphys uterus is quite limited at the present time. Therefore, more studies are needed in order to better determine the reproductive and gestational outcomes [1]. The fertility of women with untreated didelphys uterus has been shown by some sources to be better than those with other Mullerian duct abnormalities but still less than women with normal uterine anatomy. There is also an increased risk of spontaneous abortion, fetal growth retardation, and prematurity with an estimated 45% (or lower) chance of carrying a pregnancy to term in comparison to a normal uterus [4,5]. Finally, in our case, there was no problem of fertility or postoperative complication but we emphasize the importance of the early detection of Mullerian duct abnormalities in women with urogenital anomalies for avoiding the misdiagnosis and for appropriating the management.

**Acknowledgements**

The authors would like to acknowledge the help of Dr Lassri in the management.

**Funding**

No external funding sources are relevant to this submission.

**Authors’ contribution**

HE retrieved clinical information, wrote the manuscript and performed the literature review. FZ first identified this case, proposed the study and revised the manuscript for important intellectual content. YS acquired photomicrographs. AJ, KZ, AS and ZB provided valuable insight during manuscript preparation. All authors read and approved the final manuscript.

**Consent for publication**

Written informed consent for publication of their clinical details and/or clinical images was obtained from the patient.

**Competing interests**

The authors declare that they have no competing interests.

**References**