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Congenital Vaginal Atresia: About an Uncommon Case

Dr. Najwa LOUKILI ; Dr. Hanae CHIMI ; Dr. Amal BENCHINE
Fatima HASSOUNI ; Mounia EL YOUSFI; Samir BARGACH
Department of Obstetrics and Gynecology, Oncology and High-Risk pregnancies, Maternity Hospital Souissi, University Hospital Center IBN SINA, University Mohammed V, Rabat, Morocco

Abstract:- Congenital vaginal atresia is an uncommon developmental defect in the female reproductive system. In this review, we discuss a rare case involving a 19-year-old woman who presented with primary amenorrhea and recurrent pelvic pain. Upon clinical evaluation, a cup-shaped structure was noted in place of the vaginal opening. Imaging studies, including ultrasound and MRI, revealed a complete vaginal atresia along with a right unicornuate uterus, a non-functional rudimentary left horn, and significant accumulation of menstrual blood in the uterus. Diagnosed with vaginal aplasia and an abnormal uterus, the patient consented to undergo a subtotal hysterectomy, with preservation of the adnexa. Congenital vaginal atresia generally presents with primary amenorrhea and chronic pelvic pain. Diagnosis involves a thorough clinical assessment coupled with imaging techniques, where MRI plays a key role in determining the severity of the atresia and informing the surgical strategy. The goal of surgery is to restore the integrity of the utero-vaginal tract and improve reproductive outcomes for those affected.

Subject Areas:- Gynecology – Obstetric

Keywords:- *Congenital Vaginal Atresia, Cyclical Pelvic Pain, IRM, Primary Amenorrhea , Malformed Uterus.*

I. INTRODUCTION

Congenital vaginal atresia is a rare developmental anomaly in females, resulting from incomplete canalization of the urogenital [1]. Often associated with Mayer-Rokitansky-Küster-Hauser syndrome, which also involves uterine aplasia, this condition can occur in about 9% of cases with an intact uterus [2]. It affects roughly 1 in 4,500 female births [3]. The Müllerian ducts, responsible for forming the fallopian tubes, uterus, and upper vagina, are typically involved in this condition, while the lower vagina develops from the urogenital sinus. Isolated vaginal atresia is due to defects in the formation of the distal paramesonephric ducts [1]. This review explores the clinical presentation and treatment approach for a case of vaginal atresia with an abnormal and non-functional uterus.

II. CASE REPORT

We present the case of a 19-year-old woman with no significant medical history .She presented to our department with a primary amenorrhea associated with chronic and cyclic pelvic pain that had been evolving for five months. Her clinical examination was normal including a good size and well-developed secondary sexual characteristics, the external genitalia appeared also normal, with well-defined labia majora, labia minora, and clitoris. However, an imperforate hymen was noted, with no continuation to a vaginal cavity. On abdominal palpation, a very paintful pelvic mass of 4cm width and 3cm height was noted**.** Her ultrasound revealed a bicornuate uterus with a right horn measuring 4x3 cm, showing a cavitary thickening and heterogeneous retention of 4x3 cm, in the left horn, only the corpo-fundal region was distinct and measured 3x2 cm; the rest of the ultrasound was normal, with present ovaries and no evidence of pelvic effusion (Figure 1).

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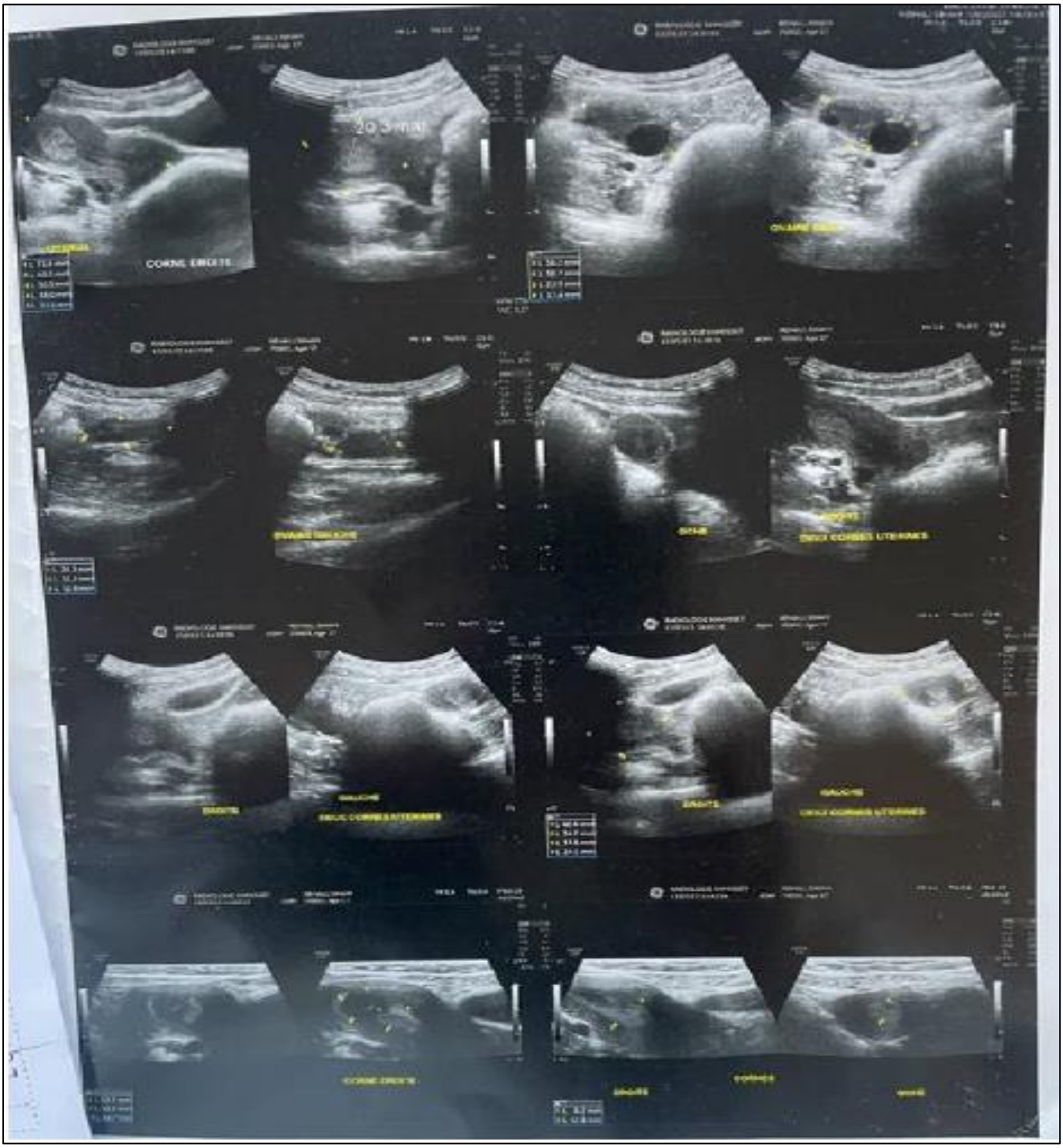

Fig 1. Echography Showing a Bicorne Uterus with Hematometry.

Magnetic resonance imaging (MRI) showed A bicornuate uterus with a right horn containing significant hematometra of 4x3 cm and a rudimentary, non-functional left horn, the vaginal cavity was absent, the ovaries were normal with no other associated malformations (Figure 2).

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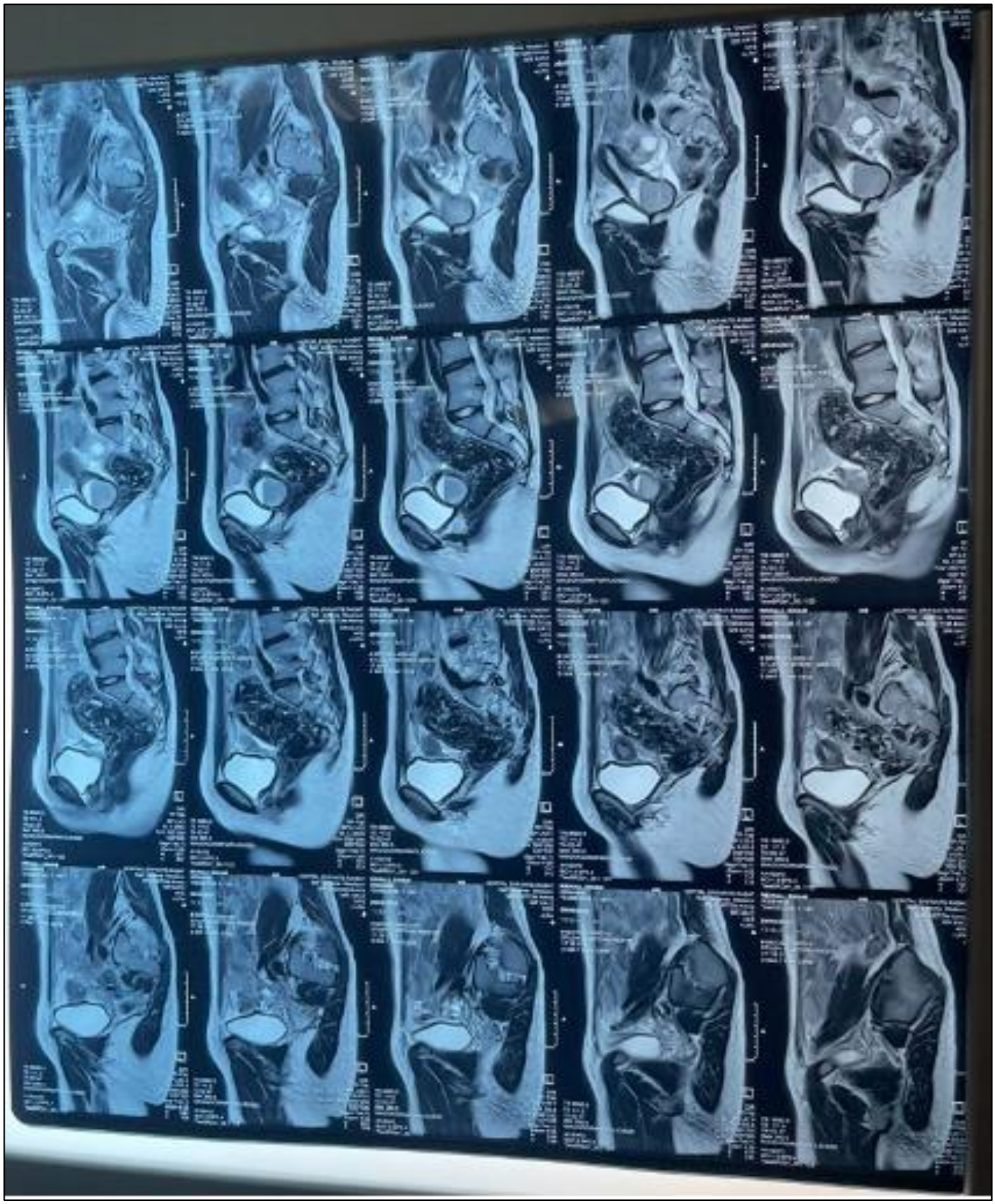

Fig 2. An MRI Showing Complete Vaginal Agenesis with a Bicorne Uterus and Significant Hematometry.

We therefore concluded a diagnosis of complete vaginal aplasia with a malformed, non-functional uterus containing hematometra. Following this diagnosis and after explaining all therapeutic options to the patient and her parents, we opted for a subtotal hysterectomy. During open surgical exploration, the uterus was enlarged due to an important hematometra. Vesicouterine detachment was performed, along with ligature and sectioning of the utero-ovarian ligaments. A subtotal hysterectomy was performed (Figure 3). The postoperative recovery was without complications.

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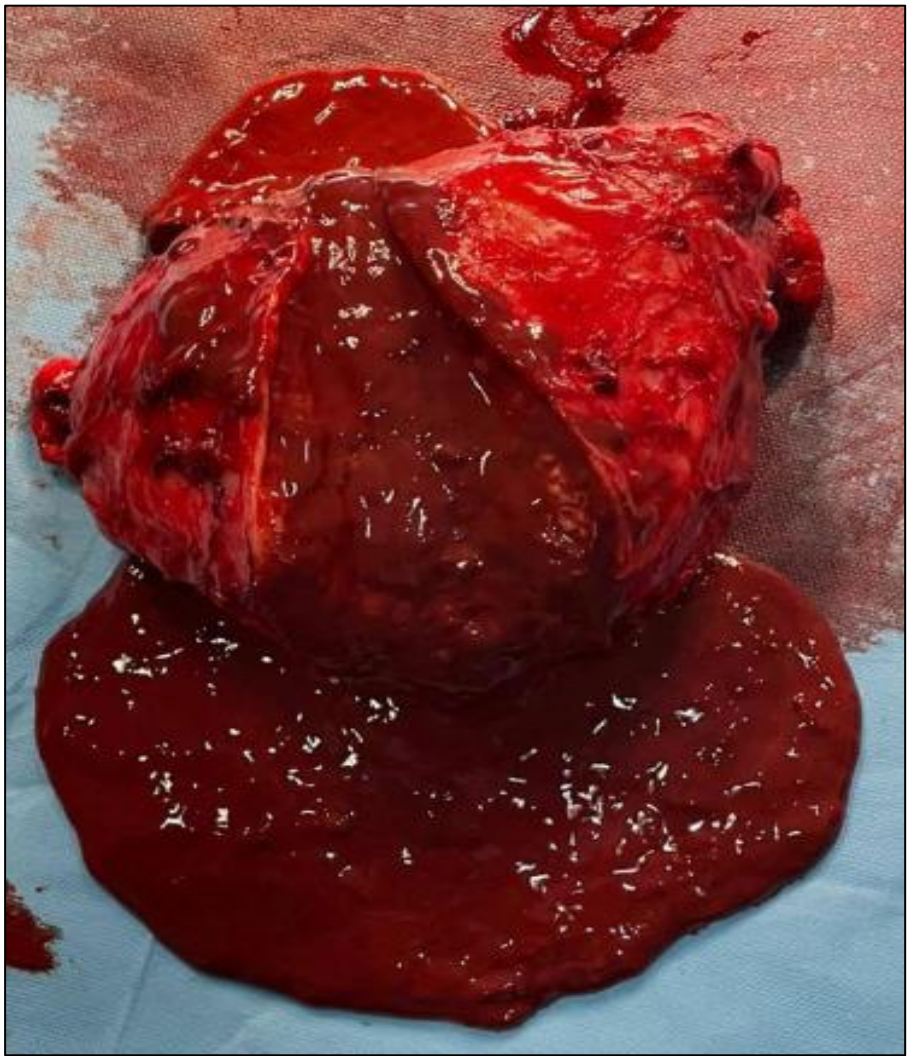

Fig 3. Hysterectomy Inter-Annexal with Hematometra.

III. DISCUSSION

Congenital vaginal atresia is a rare developmental disorder where the vaginal canal does not form correctly due to abnormalities in the paramesonephric ducts during embryonic growth. As a result, the vagina is replaced by fibrous tissue, affecting normal vaginal structure and function [4]. Typically, a pubertal girl with congenital vaginal atresia will present with primary amenorrhea and persistent pelvic pain. In some cases, diagnosis may be postponed until adulthood, especially if the primary symptom is difficulty with sexual intercourse due to inadequate vaginal penetration. Clinical examinations usually reveal normal findings, including appropriate height, well-developed breasts, and sexual characteristics, as observed in our patient. The gynecological examination reveals only a vaginal pouch measuring 3 to 4 cm. Pelvic ultrasound confirms the diagnosis by revealing the presence of an hematometra with either total or partial vaginal agenesis, as well as functional ovaries containing follicles. Renal ultrasound or intravenous urography will be used to check for any associated urinary malformations. MRI delivers a precise evaluation of vaginal aplasia, crucial for selecting the optimal surgical approach [5]. The most frequent cause of these agenesis cases is Mayer-Rokitansky-Küster-Hauser syndrome, which is associated with uterine aplasia [6, 7]. It is important to note that vaginal aplasia is often associated with other malformations, such as uterine abnormalities, malformative uropathies such as unilateral , renal agenesis, or renal dystrophy [8] . However, our patient did not present any of these associated. The management of these patients is surgical [9]. Therapeutic management aims to create a functional neovagina for sexual activity and menstrual flow [10, 11]. As surgical techniques, we find the Williams vulvovaginoplasty, using labia majora to form a perineal pouch, is outdated due to inadequate sexual penetration. The Vecchietti procedure expands the vaginal vault through gradual pressure. Other methods create a neovagina in the rectovesical space using tissues like skin, peritoneum or intestine [12]. In cases of complete vaginal atresia,a total or subtotal hysterectomy is often preferred by many gynecologists to prevent issues such as cervical or vaginal stenosis, adhesions, and pelvic inflammation resulting from poor menstrual blood drainage [13] as was the case for our patient.

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IV. CONCLUSION

Congenital vaginal atresia is a rare condition causing primary amenorrhea and pelvic pain. Diagnosis is confirmed through clinical assessment and MRI, which helps in planning surgery. The surgery's goal is to reconstruct the utero-vaginal tract, aiming to improve sexual function and quality of life.

- *Consent*

A consent was obtained from the patient to publish 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.

- *Ethical Approval*

This case report is exempt from ethical approval in our institute. Case reports are exempt from ethical approval in our University Hospital.

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