Vaginal Aplasia with Functional Uterus: Two Cases and Review of Literature

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ABSTRACT

Maldevelopment of the vagina is usually combined with aplasia or hypoplasia of the uterus. **Objective:** To support the clinical peculiarities, imaging and surgical technique of this rare entity. **Materials and Methods:** This is a retrospective study conducted over 04 years. We collected two cases of vaginal aplasia with functional uterus. The diagnosis was based on clinical data, ultrasound, imaging. **Results:** Our patients had primary amenorrhea plus chronic, cyclic pelvic pain. The examination found partial vaginal aplasia for the first patient and complete vaginal aplasia for the second patient. Magnetic resonance imaging revealed atresia of the cervix with absence of the upper part of the vagina for the first case, an absence of the vagina for the second case. Both patients had vaginoplasty. The evolution was satisfactory for the first patient and the second evolution was marked by cervicovaginal stenosis, the patient had a hysterectomy. **Conclusion:** Whatever the surgical technique practiced, vaginoplasty allows in this rare malformation to restore the physical integrity and the sexual function, even if other case resulted in cessation to cervico-vaginal stenosis.

**Keywords:** Vaginal Aplasia, Functional Uterus, Ultrasound, Magnetic Resonance Imaging neovagin, skin graft, hysterectomy

1. INTRODUCTION

Isolated vaginal aplasia is a rare pathological entity due to abnormal development of the terminal part of Müller's canals. The vagina is formed during the third month of the embryo's life. The most common etiology of these agenesis is Mayer-Rokitansky-Kuster-Hauser syndrome associated with uterine aplasia(1). The poor development is usually combined with aplasia / hypoplasia of the uterus, resulting in a functional uterus in less than 10% of cases(2). The incidence of this anomaly is between 1: 4000 and 1: 20,000(3,4). Usually, vaginal aplasia is partial and the distal, medial or cranial third is affected. Vaginal aplasia may also be associated with cervical atresia. Total hysterectomy is recommended by some authors when canalization procedures are unsuccessful or impossible(5,6), Buttram, 1983(7). This latter management is certainly successful in relieving symptoms of cervical atresia, but has an irreversible effect on reproductive performance. We have reported two clinical cases of functional uterine vaginal aplasia, a literature review, and described the ultrasound diagnostic criteria, magnetic resonance imaging, and surgical techniques of this rare pathological entity.
2. METHODS

This is a retrospective and prospective study that was conducted on 2 patients from 2013 - August 2016, during which we collected 2 cases of vaginal aplasia on functional uterus. The diagnosis was based on clinical examination, ultrasound data and magnetic resonance imaging. Therapeutic management was a creation of a neovagin by a skin graft. The evolution was satisfactory with regular cycles for the first patient and the second patient was marked by cervico-vaginal stenosis, the patient had a hysterectomy.

3. CASE REPORT

Case one
A 17-year-old woman, single, with no notable pathological history who consulted for primary amenorrhea associated with chronic cyclical pelvic pain that had been evolving for two years. The clinical examination found a normal morphotype, a correct size, well developed secondary sexual characteristics, without abdominopelvic mass. At the vulvo-perineal inspection found a normal appearance of the external genitalia, labia majora, labia minora and a perfectly constituted clitoris. The rectal touch showed pelvic tenderness, breast examination and lymph node examination were abnormal (Figure 1).

Ultrasound showed hydrometry with cervical atresia without visualization of the vagina, two small cystic images of the ovaries, homogeneous, with a finely echoic content measuring 27 and 30 mm, without peritoneal effusion (Figure 2).

The diagnosis of vaginal agenesis on functional uterus with cervical atresia was retained. The patient underwent vaginoplasty by a double abdomino-perineal route. The surgical procedure began with a laparoscopy that was performed of which at intrabdominal exploration revealed a normal size of the uterus, appendages without particularity. Given the
difficulty of identifying the atretic neck and in order to minimize the risk of a wrong course, a body hysterotomy was performed with the discovery of cervico-isthmic agenesis and the creation of a tunnel between the uterine cavity and the corresponding fibrous region. cervico-isthmic region and vaginoplasty with a skin graft and placing a condom filled with compresses to prevent stenosis for 7 days (figure 4) and (figure 5).

During the follow-up, the patient benefited from a control hysteroscopy; The visualization of the cervico-isthmic canal which was gaping, the visualization of the orifice communicating the cervico-isthmic canal. After 2 months, the patient presented the same symptomatology with speculum gynecological examination: cervix not seen, vaginal touch uterus increased in size by two fingers above pubic symphysis. A pelvic ultrasound revealed an enlarged uterus, an image of hematometry with bilateral haematosalpinx. Subtotal hysterectomy + right oophorectomy + bilateral salpingectomy was performed. The evolution was favorable with disappearance of pelvic pain.

Case two
22-year-old patient, married with no particular pathological history, who consulted for primary amenorrhea plus chronic, cyclic pelvic pain for three years. The clinical examination revealed a normal morphotype, well-developed secondary sexual characteristics, without abdominal-pelvic mass. At the vulvo-perineal inspection, a normal appearance of the external genitalia with large, perfectly defined labia minora and clitoris. On examination with speculum and vaginal examination, found a vaginal cup of 5 cm, without lateral mass uterine. It was a one-eyed vagina. Ultrasound showed hydrometry with cervical atresia without visualization of the upper part of the vagina. Magnetic resonance imaging showed an anteverted uterus of 70x48 mm, with fluid retention at the level of the emptiness line with 13 mm endometrial thickening and homogeneous myometrial wall. The cervix was atretic with no visualization of the upper end of the vaginal area.

The diagnosis of partial vaginal agenesis with functional uterus and cervical atresia was retained. The patient underwent vaginoplasty by the abdominoperineal approach. The technique used by the perineal approach was to make a transverse incision of the mucosa of the vaginal cup with detachment of the intervalesco-rectal space, with a laparoscopy which was carried out which at intra-abdominal exploration revealed a size normal of the uterus, appendages without particularity. Given the difficulty of locating the atretic neck and in order to minimize the risk of a wrong course, a corporal hysterotomy was performed with the introduction of a probe to guide the dissection to the atretic neck, which allowed a permeabilization of the passage of the probe to the vagina. The vaginal walls have been implanted on the uterine isthmus. The probe was kept for ten days to reduce the risk of cervicovaginal stenosis and to maintain continuity between the uterine cavity and the vagina. The postoperative course was simple. The evolution was marked by the appearance of regular cycles with improvement of pelvic pain.

4. DISCUSSION
Isolated vaginal aplasia is a rare pathological entity due to abnormal development of the terminal part of Müller’s canals. The vagina is formed during the third
month of embryonic life. The most common etiology of these agenesis is Mayer-Rokitansky-Kuster-Hauser syndrome associated with uterine aplasia\(^1\). Maldevelopment is usually combined with aplasia / hypoplasia of the uterus, resulting in a functional uterus in less than 10% of cases\(^2\). The incidence of this anomaly is between 1: 4000 and 1: 20,000\(^3\). Usually, vaginal aplasia is partial and the distal, medial or cranial third is affected. Vaginal aplasia may also be associated with cervical atresia. The cervix may be normal or atretic. This malformation is secondary to a lack of development of para-mesonephrotic ducts during embryogenesis\(^1,8\). Atresia of the cervix is a very rare Müllerian malformation associated with 50% of cases with vaginal aplasia\(^9\). In our case both patients had cervical atresia and vaginal aplasia. Any abdominal or pelvic pain, acute or chronic, in a girl should be suggestive of an obstructive genital syndrome. (Both cases had chronic abdominopelvic pain) The presence of a mass in the vagina, discovered during a rectal examination, suggests a blood retention over an obstacle\(^10\).

The common clinical picture is that of a girl of pubertal age who consults for primary amenorrhea whose normo-hormonal character is evident, from the outset to the inspection before the developed secondary sexual characteristics. Interrogation raises the notion of cyclic chronic pelvic pain. The clinical examination finds a normal morphotype, a correct traille, a good hairiness supplied, perfectly constituted external genitals. This is the case of our two patients. The speculum examination associated with the vaginal touch or the digital single-digit vaginal touch combined with the rectal examination perceives only a vaginal cup that measures 3-4cm, this vaginal cup is one-eyed without any slits\(^11\). (This is the case of the second patient). Clinical examination easily eliminates hymen imperfection or blind hemivagin, but can not differentiate cervical atresia from high vaginal diaphragm. Trans-abdominal or trans-perineal ultrasound may specify the level of the obstruction\(^10\) but does not seem very reliable for the diagnosis of cervical atresia\(^12\). Pelvic ultrasound confirms the diagnosis. It objectifies the image of a hematology with total or partial vaginal agenesis, functional ovaries containing follicles. Renal ultrasound or intravenous urography will look for an associated urinary malformation. Indeed, early recognition of a particular pelvic kidney will avoid possible trauma during surgical reconstruction. In contrast, transrectal ultrasound can help to analyze the cervix because it provides a precise view of the pelvic organs\(^12\) and three-dimensional ultrasound\(^13\) can help to analyze the external shape of the cervix, uterus. The interest of magnetic resonance imaging is to better specify the height and extent of vaginal aplasia thus making it possible to guide the choice of the most appropriate surgical technique\(^14\).

The management of these patients is surgical. It aims not only to create a neo vagina that would allow satisfactory sexual intercourse but to reconstitute a normal utero-vaginal chain to allow the evacuation of menstrual blood and cerebral secretions. Laparoscopic exploration has the ability to assess the type of uterine malformation and reveals other complications of the upper genital tract that require appropriate surgery. This is the case of our two patients. Although some cases have been successful, others have been reported\(^15\), with a restoration of continuity between the functioning of the uterus and vagina in cases of cervical or vaginal atresia that is intimidating. As a result, hysterectomy has been recommended as the most direct treatment option\(^16\). Our first patient had a hysterectomy. Deffarges et al. reported a weak secondary vaginal stenosis in two patients (11%) away from the uterine anastomosis and cervical stenosis, resulting in dysmenorrhea and requiring multiple channeling procedures\(^17\). In the case report of Hampton et al. The vagina was reconstructed using standard thickness McIndoe vaginoplasty skin graft\(^17\). Our first patient had vaginoplasty with a graft of the skin.

5. CONCLUSION

Vaginal aplasia is a rare congenital malformation whose classic clinical picture is that of primary amenorrhea with chronic cyclic pelvic pain. Diagnosis is based on clinical examination and imaging. Magnetic resonance imaging aims to assess the importance of atresia and thus guide surgical management. Whatever the surgical technique practiced, it will aim to restore the integrity of the utero-vaginal sector and allow these patients, whose psychological experience is particularly difficult, to have a satisfactory sex life and a possibility of pregnancy.
REFERENCES