SAR Journal of Medical Case Reports

Abbreviated Key Title: *SAR J Med Case Rep*
Home page: https://sarpublication.com/journal/sarjmcr/home
DOI: 10.36346/sarjmcr.2022.v03i04.003

ISSN 2707-7780 (P)
ISSN 2709-6947 (O)

Case Report

Vaginal Aplasia with Functional Uterus, A Rare Entity: A Case Report

Lounas Benghanem[1*], Lydia Faïd[1], Mounir Bisker[1], Kamel Haïl[2], Radia Benyahia[3]
[1]Gynecology and Obstetrics Department, Mustapha University Hospital Center, Faculty of Algiers I. Algeria
[2]General Surgery Department, Mustapha University Hospital Center Algiers Algeria
[3]Radiology Department, Marie Curie Centre Algiers Algeria

***Corresponding Author:** Lounas Benghanem
Gynecology and Obstetrics Department, Mustapha University Hospital Center, Faculty of Algiers I. Algeria

Article History: | Received: 08.10.2022 | Accepted: 17.11.2022 | Published: 24.11.2022 |

Abstract: Isolated vaginal agenesis is a rare malformation of the genital tract. It is characterized by the absence of the vagina, or the presence of a vaginal dimple measuring less than 5 cm. We report the case of a 21-year-old married patient who presents with primary amenorrhea, an inability to have sexual intercourse due to isolated vaginal aplasia.
Keywords: Isolated vaginal atresia, Functional uterus, Mixed approach, A case report.

INTRODUCTION

Vaginal agenesis is a rare congenital malformation [1], it is often part of Mayer-Rokitansky-Kuster-Hauser syndrome which associates uterovaginal aplasia. On the other hand, isolated vaginal aplasia is exceptional due to an abnormality in the development of the terminal part of the Müller ducts; it represents 9% of vaginal malformations [2].

PATIENT AND OBSERVATION

Patient Information

This is a 21-year-old married patient, with no particular pathological history, who consulted for associated primary amenorrhea and the inability to have intercourse with her partner. The clinical examination reveals a normal morphotype and well-developed secondary sexual characteristics. The vulvoperineal inspection finds the labia majora and minora and a perfectly formed clitoris and a vaginal cup of one centimeter (figure 1).

Ultrasound revealed hydrometry with cervical atresia and left hematosalpinx.

Objective magnetic resonance imaging (figure 2) absence of vaginal canal, malformation of the uterine cervix such as atresia with teardrop appearance, presence of a uterine cavity with hematic content, dilation of the left tube to 17 mm without other anomalies.

The diagnosis of total vaginal agenesis on a functional uterus with cervical atresia and retained.

Therapeutic Intervention

The patient was operated on under general anesthesia; we performed a vaginal plasty using a mixed abdominoperineal approach [3].

The technique used consisted of an incision in the mucosa of the vaginal cup with careful separation of the interuretero and vesico-rectal space, starting laterally and ending in the center of the virtual uretero and vesico-rectal space, guided by the urinary catheter. Via the upper route we made a longitudinal fundal hysterotomy and we introduced a Heggar bougie to help guide the dissection to the atretic neck (figure 3). The walls of the isthmus were thus installed on the vestibule. A Foley catheter was kept for 7 days to reduce the risk of cervical stenosis.

The postoperative course was simple, the Foley catheter was removed after the 7th day, the patient was seen again on the 14th day (figure 4) to check the patency of the cervix, the patient had her period and began to have sexual activity.

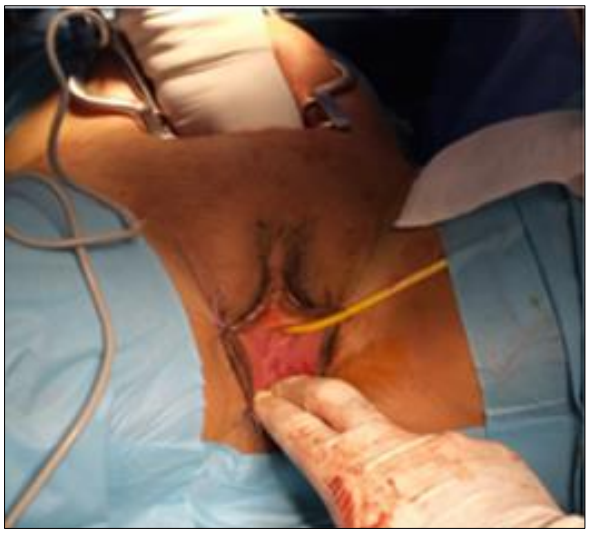

Figure 1: Vaginal cup

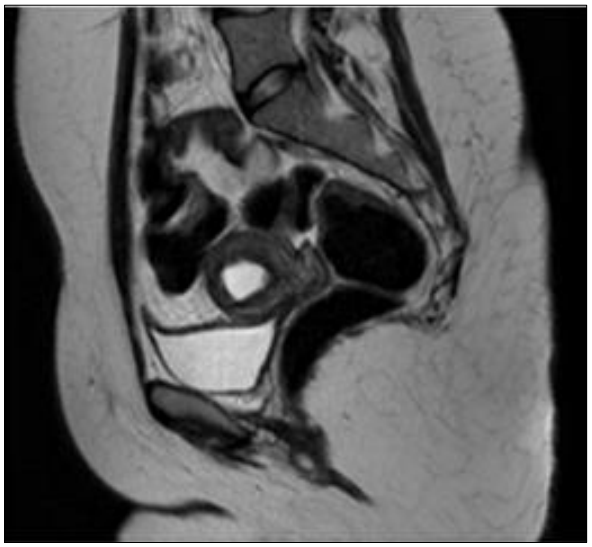

Figure 2: Absence of vagina and functional uterus

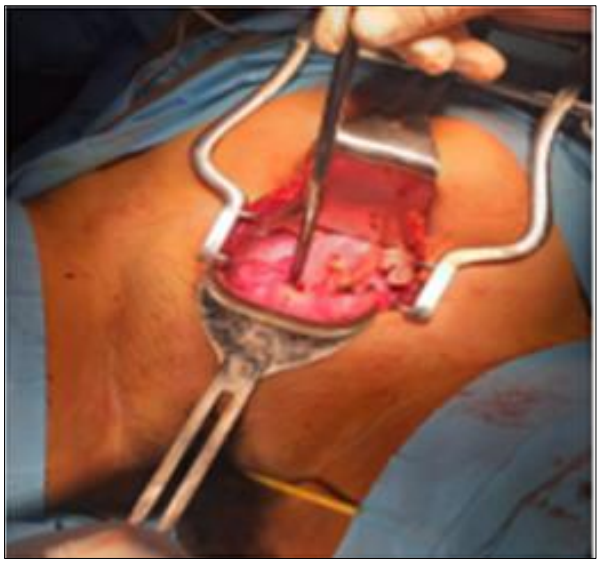

Figure 3: Longitudinal fundal hysterotomy and we introduced a Heggar bougie

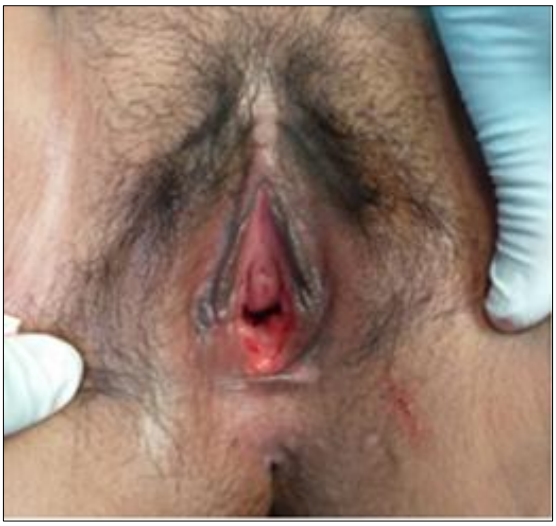

Figure 4: Appearance of the vaginal entrance postoperatively on day 14

DISCUSSION

Most vaginal congenital malformations are rare; these malformations may correspond to hymenal imperforation, duplication, fusion and agenesis or aplasia. Vaginal aplasia is a congenital pathology which, in 90% of cases, results in Mayer-Rokitanski-Kuster-Hauser syndrome [4].

There are two types of aplasia: partial vaginal aplasia, particularly of the upper 2/3, and total vaginal aplasia. In terms of embryogenesis, the Müller ducts form the upper two thirds of the vagina, the uterus and the tubes, the lower part of the vagina responds to the urogenital sinus. Isolated vaginal aplasia is therefore the consequence of a lack of development of the terminal part of the paramesonephrotic ducts [5]. Often the external part of the vagina is present since it has a different embryological origin.

Clinically, it is often a pubescent girl who presents primary amenorrhea with the presence of secondary sexual characteristics (this is the case of our patient). With the notion of chronic and cyclical pelvic pain. Inspection of a vaginal cup. Rarely, the diagnosis may be late in a woman who consults for difficult sexual relations (the case of our patient) or who consults for primary infertility.

Radiologically, ultrasound and magnetic resonance imaging [6], make the diagnosis by revealing partial or total vaginal agenesis which is replaced by fibrous tissue, the uterus is usually increased in size due to the hematometrocolpos. Imaging will look for an associated urinary malformation (renal agenesis, ectopy, malrotation or renal dystrophy.

The treatment of vaginal agenesis is surgical, the objective is to create a neovagina to allow the menstrual flow and sexual intercourse. The management of vaginal aplasia is divided into two groups: when the

atresia is limited it benefits from an anastomosis of the vaginal mucosa with the isthmus. When the atresia is complete, surgery is more delicate: uterovestibuloplasty as in our patient, skin or intestinal grafts [7].

CONCLUSION

Isolated vaginal aplasia is a very rare pathology, the diagnosis is clinical and radiological, given amenorrhea and cyclical pain, additional examinations in particular magnetic resonance imaging are established. The treatment is surgical to allow the flow of periods, sexual activity with hope of pregnancy.

Conflicts of Interest: The authors declare no conflicts of interest.

Authors' Contributions

Lounas BENGHANEM: data collection, bibliographic research and writing of the article.
Lydia FAÏD: proofreading and supervision of the writing of the article.
Mounir BISKER: proofreading and supervision of the writing of the article.
Kamel HAÏL: proofreading and supervision of the writing of the article.
Radia BENYAHIA: proofreading and supervision of the writing of the article.

REFERENCES

1. Practice Committee of American Society for Reproductive Medicine. (2008). Current evaluation of amenorrhea. *Fertil Steril*, *90*(5 Suppl), S219-25. doi: 10.1016/j.fertnstert.2008.08.038. PMID: 19007635.
2. ACOG Committee on Adolescent Health Care. (2006). ACOG Committee Opinion No. 355: Vaginal agenesis: diagnosis, management, and routine care. *Obstetrics and gynecology*, *108*(6), 1605-1609.
3. Randriamananjara, N., & Rabarioelina, L. (1995). Un cas d'agénésie vaginale isolée traitée par plastie péritonéale en T renversé. Journal de chirurgie (Paris. 1908), 132(1), 49-50.
4. Paniel, B. J., Haddad, B., & Meneux, E. (1996). Chirurgie des malformations vaginales (I). *Encycl Méd Chir*, 41-855.
5. Roberts, C. P., Haber, M. J., & Rock, J. A. (2001). Vaginal creation for müllerian agenesis. *American journal of Obstetrics and gynecology*, *185*(6), 1349-1353.
6. Laroche, E., Bricaire, L., & Christin-Maitre, S. (2013). Diagnosis and management of amenorrhea in adolescent girls. *Archives de pediatrie: organe officiel de la Societe francaise de pediatrie*, *20*(7), 817-822.
7. Ugur, M. G., Balat, O., Ozturk, E., Bekerecioglu, M., & Dikensoy, E. (2012). Pitfalls in diagnosis and management of distal vaginal agenesis: 10-year experience at a single centre. *European Journal of Obstetrics & Gynecology and Reproductive Biology*, *163*(1), 85-90.