Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Urology

Vaginoplasty on Androgen Insesibility Syndrome: A Props of A Case

H. Lachhab*, Y. Staouni Benabdallah, Y. Rachid, A. Benkerroum, A. Moataz, M. Dakir, A. Debbagh, R. Abouateib

Service d'Urologie, CHU Ibn Rochd-Casablanca Maroc

DOI: 10.36347/sjmcr.2022.v10i05.003 | **Received:** 21.03.2022 | **Accepted:** 25.04.2022 | **Published:** 09.05.2022

***Corresponding author:** H. Lachhab
Service d'Urologie, CHU Ibn Rochd-Casablanca Maroc

Abstract **Case Report**

Feminizing Testicular Syndrome is a very rare genetic disease of recessive transmission linked to the X chromosome, currently called Androgen Insensitivity Syndrome (AIS). The affected subjects are apparently normal women, but with a 46 XY male karyotype with ectopic testicles. Through a case of "AIS revealed" by a germ tumor (GT) on an ectopic testicle in a 33-year-old patient who underwent a bilateral orchidectomy. The patient received 10 sessions of chemotherapy declared in remission. Following a sexual desire, the patient received a sigmoid vaginoplasty 5 years later according to Schmid's technique. The surgical follow-up was favorable and the patient quickly resumed a satisfactory sexual activity.

Keywords: genetic disease, Feminizing Testicular Syndrome, Androgen Insensitivity Syndrome (AIS). germ tumor.

INTRODUCTION

Lack of vagina is a malformative syndrome that can be isolated or associated with other genital malformations.

Thus vaginal aplasia can be associated with a uterus reduced to two rudimentary uterine horns. This malformative aspect corresponds to Rokitansky-Kuster-Hauser syndrome. The absence of vagina can be isolated with a normal uterus.

Finally, vaginal aplasia may be associated with male pseudohermaphrodism falling within the framework of a sexual ambiguity. The absence of vagina is usually discovered in adolescence, by amenorrhea. This discovery may only occur in adulthood.

Vaginal reconstruction involves several therapeutic methods, from simple dilation or Frank's technique, to vaginal implantation by a free cutaneous flap or McIndoe technique or by a sigmoid or ileal intestinal graft. The goal is to create an inter-vesico-rectal cavity for satisfactory sexual intercourse.

We report our experience of vaginoplasty using the sigmoid colon of a patient with androgen insensitivity syndrome on testicular germ tumor.

OBSERVATION

She is a 33-year-old patient, declared a girl at birth, underwent surgery at the age of 7 for a right inguinal hernia, with the concept of primary amenorrhea. No similar cases in the family and no concept of inbreeding. The patient consulted the Gynaecology and Obstetrics department of the Ibn Rochd Hospital in Casablanca for an abdomino-pelvic mass that had been evolving for 7 months and was referred to the P40 after an exeresis of the mass for the management of a germ tumor on the ectopic testicle. Clinical examination before surgery found a patient in good general condition, sub umbilical laparotomy wound is well healed, the external genitals are typically female without pubic or axillary hair with vaginal aplasia (Figure 1) and TANNER stage 2 breasts. Before the tumor mass surgery, the radiological assessment showed a voluminous hypervascularized solid-cystic centro-pelvic mass measuring 126x180x100mm associated with left lateral-aortic metastatic lymphadenopathy without individualization of the uterus or ovaries.

No remote metastases. Tumour markers were requested: BHCG at 125.4mIU/ml, AFP at 3.6ng/l, LDH at 229IU/l. Blood hormonal balance was disturbed with a high level of total testosterone. Genetic examination revealed a male chromosome formula of type 46, XY. The patient benefited from a tumor mass

removal with anatomical-pathological and immuno-histochemical study, a TGNS (seminoma and yalk sac tumor) of 18.5cm of major axis with images of vascular embolus on the left ectopic testicle. Postoperative tumour markers have normalized.

Postoperative thoraco-abdomino-pelvic CT showed deep abdominal lymphadenopathies. At the end of this assessment, the non-seminomatous germ tumour is classified as stage IIB with good prognosis. The patient received three courses of chemotherapy (BEP protocol). Thoraco-abdomino-pelvic scanner performed at the end of chemotherapy showed the disappearance of the abdominal lymphadenopathy. The patient was referred to her gynecologist for AIS management.

5 years later and following a sexual desire, a sigmoid vaginoplasty according to Schmid's technique was performed (Figure 2), with vaginal dilation by candles per op (Figure 3)

The operating sequences were favourable with a resumption of transit at d+3 and feeding at d+5.

Anatomical results were good with a neovaginal length of nine centimeters. Functionally, the patient quickly returned to satisfactory sexual activity. She didn't have to perform any autodilation of the neovagina. Only regular sexual activity had maintained a good anatomical and functional result of the vaginal implantation. No stenosis of the vulvar orifice was noted.

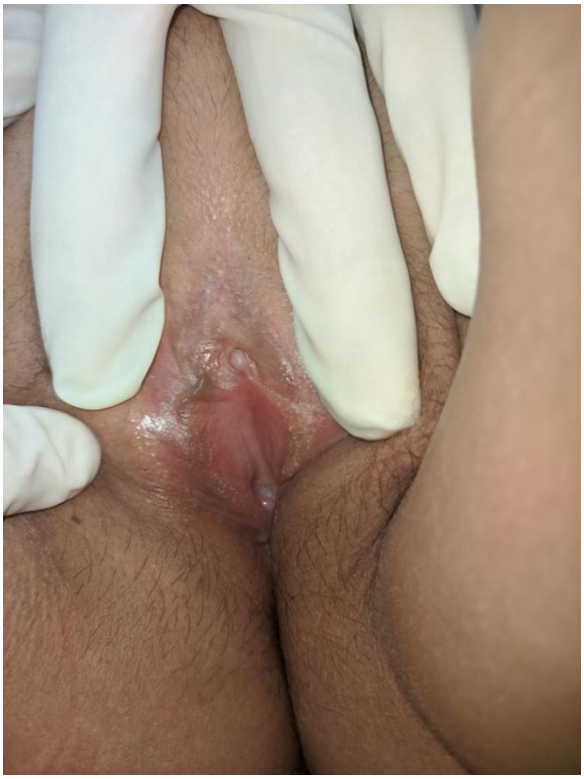

Figure 1: Initial state of the patient

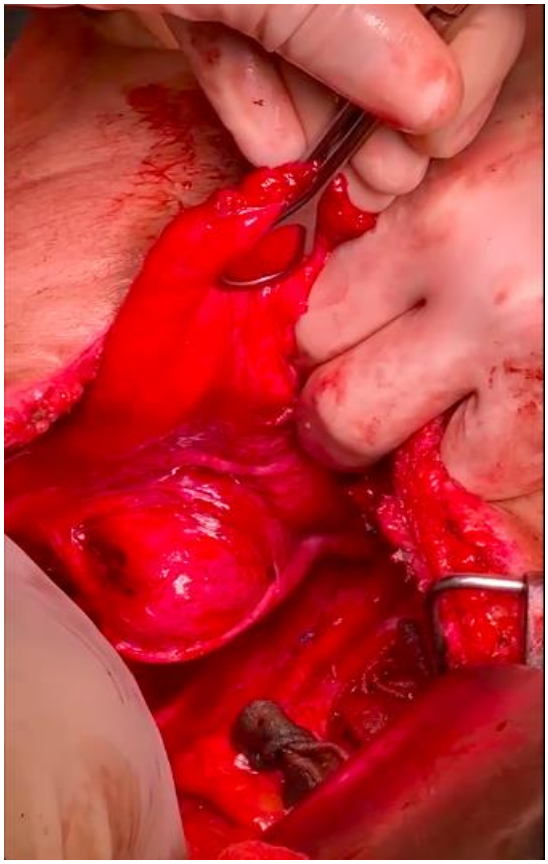

Figure 2: Vaginoplasty

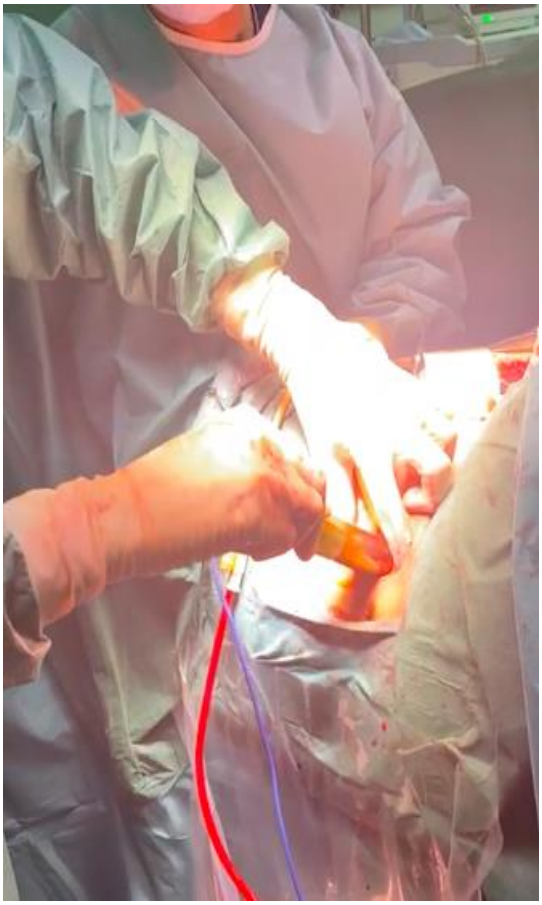

Figure 3: Vaginal dilatation

DISCUSSION

Vaginal sigmoid implantation was first described in 1911 by Albrecht. Already in 1904, Balwin had described the first vaginoplasty using hail [2]. In 1977, Schmid took up this technique and modified it and is now known as his name [1]. This type of vaginoplasty is widely adopted by Oriental and Germanic authors [3]. Vaginal implantation is indicated for the treatment of vaginal aplasia. These aplasias are mainly represented by the Müllerian aplasia known as Rokitansky-Küster-Hauser syndrome characterized by the congenital absence of the upper two thirds of the vagina associated with a normal vulva. The uterus is absent or rudimentary with normal ovaries.

The sigmoid vaginal implantation is a simple, reproducible and not morbid technique. No complication was noted in our case, as was the case for most authors [4-6]. This plastic surgery could be performed under celiac surgery as described by Delga and Ohashi [7, 8]. Anatomical results are satisfactory for the majority of authors with a mean length of more than 9 cm [3, 6, 9]. Vulvar stenosis is rarely noted with a rate ranging from 0 to 5% [3, 6]. Martinez noted only one post-operative vulvar stenosis on 19 plasties [6]. She gave in with simple erased incisions. This complication did not occur in our patient.

Sexual activity is satisfactory for the majority of authors with a rapid resumption of sexual intercourse from the first month [3, 4,6, 9, 10]. Dyspareunia is rarely reported as it is found in less than 10% of cases [3, 6]. The major problem during sexual resumption is psychological with the apprehension of a "vaginism" requiring psychological support since Guillet noted that a third of his patients needed psychological advice with their partners [9]. The guarantor of a good result of plastic surgery is the rapid resumption of sexual activity with regular intercourse allowing an excellent sexual rehabilitation as shown by Mobus in a study on the long-term evolution of 44 patients [11].

Freundt reports two cases of prolapse occurring at 4 years and at 3 years requiring suspension of the Cooper ligament with a good progression [12].

A case of sigmoid graft cancer was reported by Hiroi 30 years after vaginoplasty requiring a complete resection of the neo-vagina and additional radiotherapy [13].

In addition to sigmoid vaginoplasty, other techniques of vaginoplasty exist, namely the techniques of peritoneal cleavage, the method of Vecchietti which is a technique of cleavage by the upper route and consists of a progressive traction by an olive of the vulvar mucosa to obtain a neovagina, the plasties using cutaneous grafts called Mac Indoe technique or bladder mucosa. These techniques are grafted with a fairly high morbidity and require the immediate postoperative wearing of a mandrel [6]. The reports require lubrication with a high rate of dyspareunia and a risk of plastic stenosis forcing the patient to perform regular autodilations [6].

Vaginoplasty can only be indicated if the young patient explicitly expresses the desire for an active sex life. A certain maturity of the patient is indeed desirable in order to obtain a favourable result for her. The neovagina should be used regularly to maintain the result after the procedure.

The choice of the sigmoid colon as a graft for the neo-vagina is wise since it provides sufficient length with a good vascularization of the neovagina. The walls of the sigmoid are more tolerant of sexual trauma than the small intestine or skin graft, thus limiting dyspareunia. Furthermore, it does not require postoperative modelling or self-dilation with rapid resumption of sexual activity. It is naturally lubricated by the secretions although sometimes they are abundant enough to require the permanent wearing of trims. This type of vaginoplasty may be performed in early childhood together with correction of other caudal abnormalities [4, 14].

The creation of a neo-vagina by the sigmoid appears to be the method of choice for the treatment of vaginal aplasia.

CONCLUSION

One of the main goals of therapeutic management is to treat vaginal aplasia and to optimize the onset of sexuality. The management of vaginal aplasia can be either non-surgical with Frank's method, which consists of progressive vaginal dilation using candles, or surgical with different techniques described.

BIBLIOGRAPHY

1. Schmid, Tamnwald, I., & Hauser, G. A. (1977). Formes atypiques du syndrome de RKH. *Geburtshilfe Frauenheilkd*, 37, 386-392.
2. JFXIV, B. (1904). The formation of an artificial vagina by intestinal trransplantation. *Ann Surg*, *40*(3), 398-403.
3. Bennani, O., & Aderbour, M. (1996). Néo-vagin par anse sigmoïdienne: A propos de 15 cas. *Revue française de gynécologie et d'obstétrique*, *91*(5), 209-216.
4. Wesley, J. R., & Coran, A. G. (1992). Intestinal vaginoplasty for congenital absence of the vagina. *Journal of pediatric surgery*, *27*(7), 885-889.
5. Del Rossi, C. A. R. M. I. N. E., Attanasio, A., Domenichelli, V., & De Castro, R. O. B. E. R. T. O. (1999). Treatment of the Mayer-Rokitansky-Kuster-Hauser syndrome in Bangladesh: results of 10 total vaginal replacements with sigmoid colon at

a missionary hospital. *The Journal of urology*, *162*(3), 1138-1139.

6. [6] Martinez-Mora J, Isnard R, Castellvi A, Lopez Ortiz P. Neovagina in vaginal agenesis: surgical methods and long-term results. J Pediatr Surg 1992 Jan;27(1):10–4.
7. Delga, P., & Potiron, L. (1997). Sigmoid colpoplasty by laparoscopic and perineal surgery: a first case relative to Rokitansky-Kuster-Hauser syndrome. *Journal of Laparoendoscopic & Advanced Surgical Techniques*, *7*(3), 195-199.
8. Ohashi, S., Ikuma, K., Koyasu, Y., Tei, K., Kanno, H., Akashi, A., & Haque, S. M. M. (1996). Laparoscopic reconstruction of vagina using sigmoid autograft. *Surgical endoscopy*, *10*(10), 1019-1021.
9. Guillet, J. L., Letessier, A., Clot, J. P., Madelenat, P., & Ravina, J. H. (1984). Néo-vagin par plastie sigmoïdienne. A propos de 12 observations d'aplasie vaginale. *Journal de chirurgie (Paris. 1908)*, *121*(2), 81-84.
10. Freundt, I., Toolenaar, T. A., Huikeshoven, F. J., Jeekel, H., & Drogendijk, A. C. (1993). Long-term psychosexual and psychosocial performance of patients with a sigmoid neovagina. *American journal of obstetrics and gynecology*, *169*(5), 1210-1214.
11. Mobus, V. J., Kortenhorn, K., Kreienberg, R., & Friedberg, V. (1996). Long-term results after operative correction of vaginal aplasia. *American journal of obstetrics and gynecology*, *175*(3), 617-624.
12. Freundt, I., Toolenaar, T. A., Jeekel, H., Drogendijk, A. C., & Huikeshoven, F. J. (1994). Prolapse of the sigmoid neovagina: report of three cases. *Obstetrics and gynecology*, *83*(5 Pt 2), 876-879.
13. Hiroi, H., Yasugi, T., Matsumoto, K., Fujii, T., Watanabe, T., Yoshikawa, H., & Taketani, Y. (2001). Mucinous adenocarcinoma arising in a neovagina using the sigmoid colon thirty years after operation: a case report. *Journal of surgical oncology*, *77*(1), 61-64.
14. Hitchcock, R. J., & Malone, P. S. (1994). Colovaginoplasty in infants and children. *British journal of urology*, *73*(2), 196-199.