Distal vaginal atresia presented with apareunia with regular menses and not primary amenorrhea diagnosed by 3D ultrasound and managed by vaginal pull-through aided by 3D ultrasound: A case report and review of literature

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Abstract

Introduction: We report a rare case of distal vaginal atresia not presented by primary amenorrhea but with the inability to have the first sexual intercourse after marriage.

Case: The patient had normal menarche and regular cycles. Thus, the challenge was in diagnosing such cases with vaginal atresia with normal menses. The menstrual flow was small in amount. The complaint came after her marriage when her husband felt strong resistance. On examination, no vaginal opening was observed. When examined again during menses, a very small opening was observed, from which the menses flow was discharged: it admitted only a pediatric Foley catheter (denoting a small fistulous tract). 3D ultrasound revealed complete uterus, cervix, and vaginal pouch development. Vaginal atresia was identified: an operation was done under a 3D rectal ultrasound guide to identifying the dissection plane. Perineal dissection was done, and pull-through of the vaginal pouch, then suturing the lower end of the vagina to the perineum.

Conclusion: not all cases of vaginal atresia cause primary amenorrhea but may present later with sexual problems. 3D rectal ultrasound is an excellent aid in diagnosis and treatment aid.

Keywords: Apareunia; Ultrasound; Distal vaginal atresia

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INTRODUCTION

Vaginal atresia is a rare congenital defect of the female genital tract due to canalization failure in the urogenital sinus [1]. During embryogenesis, the uterus, tubes, and upper 2/3 of the vagina arise from the Müllerian ducts, and the urogenital sinus gives the lower 1/3 of the vagina. Complete vaginal atresia is considered a part of Mullerian agenesis, as described by the American Society for Fertility Medicine [2]. Most reported cases of the imperforate hymen or vaginal atresia presented in the literature were presented by primary amenorrhea at the time of puberty by cryptomenorrhea, hematocolpus, or hematometra, unlike

this case which presented after marriage with a history of regular menses and no Vagina.

CASE PRESENTATION

The patient's primary concern was to have her first intercourse after her husband complained that he felt that there is no vagina, they thought that she might have a thick hymen, and she came asking for a hymenotomy. There was no relevant family history of a similar condition, no past surgical interventions or circumcision, and the patient before marriage has never complained. Physical examination (PE) during the first visit revealed no hymen, and per rectal examination, the uterus and cervix were felt with no hematocolpus. When the patient insisted that she had regular menses, we asked her to come during menses; on the second visit, the patient was menstruating, and a small dimple was seen, admitting a small pediatric foley catheter to a distance of 5 cm denoting a small fistulous tract.

Three Dimensional US was done through an endorectal probe revealing a normal uterus, cervix, and vagina was 6 cm (upper part 3.5cm x 1 cm dilatation). The lower part is 2.5 cm long and constricted, suggesting atresia rather than stenosis **(Fig. 1.)**

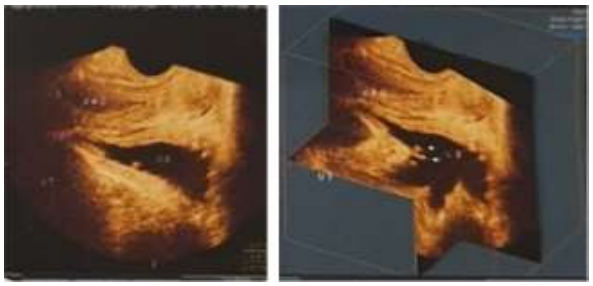

Fig. 1. a,b showing upper and lower part of vagina.

The patient was counseled for the operation about her future fertility. After preparation of the patient and routine preoperative assessment, surgery was performed; No vagina was seen even the dimple seen before was not identified; 3D endorectal probe was placed, identifying the uterus, then cervix, then the upper vaginal, perineal dissection began under 3D guidance for about 3 cm till we reached the upper vaginal pouch, then freeing of the vagina from all sides and pull through of the vaginal pouch then suturing lower end of the vagina to the perineum **(Fig. 2.)**. The patient was discharged on regular analgesics and was instructed that no sexual intercourse for three weeks.

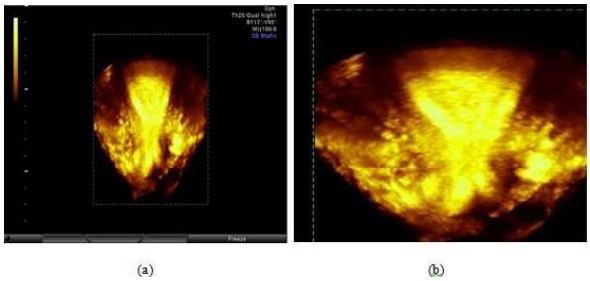

Fig. 2. a & b showed uterine cav- ity triangular which is completely normal which encouraged us to proceed for the surgery.

The patient was assessed in the outpatient clinic after the operation by one week, and a pelvic examination revealed good vaginal length; after 3 weeks, the patient had her first intercourse, and after one week of regular intercourse, she came for a second follow-up and she was happy by the procedure.

Discussion

In most cases described in the literature of distal vaginal atresia, patients were presented before the age of 16 and with primary amenorrhea [3-6]; their diagnosis is confirmed by haematocolpus and hematometra as in cases of imperforate hymen and distal vaginal atresia. In cases described distal vaginal atresia in the literature as in the case report of Awad and El-agwan [7], where the patient was a 13 years old girl who presented with abdominal swelling with a radiological finding of hematocolpus and haematometra; this case reported lower vaginal agenesis which was managed by dissecting the lower part of vagina and pull-through vaginoplasty.

The rare presentation of this case is that it describes atresia of the lower part only of the vagina with a normal upper part presenting with normal menstruation; the presence of a small fistulous opening allowed the menstrual flow. We recommend using 3D ultrasound intraoperatively in cases of vaginal atresia to identify the dissection plane.

Review of Literature

Embryology

Vaginal atresia is defined as an anomaly with failure of the natural development of the lower part of the vagina. This congenital anomaly reflects the incomplete canalization of the Mullerien ducts. Beginning from the 6th week, the embryo develops the Mullerian ducts. The ducts give origin to the Fallopian tubes and the urogenital sinus and then give the Muller tuberculum, from which epithelium grows up to give the Mullerian ducts. This contributes to the obliteration and forming of the vaginal canal and a rigid vaginal epithelial plate. In the 17th week, vaginal canalization takes place. (8)

The vagina is completely canalized by the 20th week. Vaginal anomalies result as a failure of canalization or fusion in the vertical plane and are presented clinically with a vaginal septum, atresia, or agenesis. Vaginal atresia is a Mullerian duct anomaly where fibrous tissue replaces the missing part of the vagina. (9)

Epidemiology

Female genital tract anomalies occur in 2–3% of women; The most common abnormality is the imperforate hymen (10). Isolated vaginal atresia occurs in 1:5000 women, while that uterine didelphys in 16:1000. (11). Women with impaired fertility have higher Müllerian duct anomalies (8% of women having uterus didelphys). (12)

Classifications

Mullerian anomalies are rare congenital anomalies of the female genital tract. Many classification systems for Mullerian anomalies have been proposed. The American Fertility Society Classification 1988 has been the most utilized and recognized (13).

However, the American Fertility Society classification has been criticized for focusing on uterine anomalies and excluding vaginal and cervical anomalies. This classification lacks clear diagnostic criteria and is unable to classify complex anomalies. (14)

THE European Society for Gynecological Endoscopy classification focused primarily on the uterine anatomy and classified the cervical and vaginal anomalies as independent subclasses to identify each anomaly precisely. (15)

The European Society for Gynecological Endoscopy classifies the vaginal anomalies inti 5 subclasses (V0-V4):

Sub-class V0; which includes all cases of normal vagina and normal vaginal development.

Sub-class V1; non-obstructing longitudinal vaginal septum. The included anomaly here is clear and allows classifying different variants of bicorporal or septate uterus and double or septate cervices.

Sub-class V2; non-obstructing longitudinal vaginal septum. The included anomaly here is clear and allows classifying different variants of obstructing vaginal defects.

Sub-class V3; imperforate hymen and/or transverse vaginal septum. They usually present with the same clinical presentation.

Sub-class V4; all cases of partial or complete vaginal aplasia. (16).

ESHRE/ESGE classification Female genital tract anomalies

	Uterine anomaly		Cervical/vaginal anomaly	
	Main class	**Sub-class**	**Co-existent class**	
U0	Normal uterus		C0	Normal cervix
U1	Dysmorphic uterus	a. T-shaped b. Infantilis c. Others	C1	Septate cervix
			C2	Double "normal" cervix
U2	Septate uterus	a. Partial b. Complete	C3	Unilateral cervical aplasia
			C4	Cervical aplasia
U3	Bicorporeal uterus	a. Partial b. Complete c. Bicorporeal septate		
			V0	Normal vagina
U4	Hemi-uterus	a. With rudimentary cavity (communicating or not horn) b. Without rudimentary cavity (horn without cavity/no horn)	V1	Longitudunal non-obstructing vaginal septum
			V2	Longitudunal obstructing vaginal septum
U5	Aplastic	a. With rudimentary cavity (bi-or unilateral horn) Without rudimentary cavity (bi-or unilateral uterine remnants/aplasia)	V3	Transverse vaginal septum and/or imperforate hymen
			V4	Vaginal aplasia
U6	Unclassified malformations			
U			**C**	**V**

Table 1 classification of uterine and vaginal anomalies (ESHRE/ESGE) classification system (16)

The demand for a new classification was needed so the ASRM Task Force made a new classification (17), where the categories were modified, including three additional groups; transverse vaginal septum, longitudinal vaginal septum, and finally, the complex anomalies.

In the ASRM classification, the categories are described by different terminologies:

- 1-Mullerian agenesis
- 2-Cervical agenesis
- 3-Unicornuate uterus
- 4-Uterus didelphys
- 5-Bicornuate uterus
- 6-Septate uterus
- 7-Longitudinal vaginal septum
- 8-Transverse vaginal septum
- 9-Complex anomalies (17)

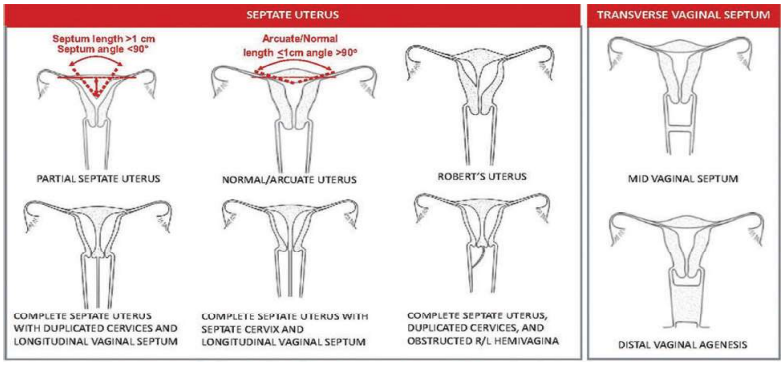

Fig (3) showing some uterine and vaginal anomalies including distal vaginal atresia according to ASRM classification

Clinical Presentations

It is reported that 39 patients with vaginal atresia are at an age average of 16 years. Primary amenorrhea was the most common presentation (71%), periodic abdominal pain (41%), lower abdominal pain (36%), dyspareunia (10%), menstrual irregularities (5%), and pelvic swellings (5%) (18). In another study on seven neonates, six had vaginal malformations combined with anorectal anomalies. The majority of infants show fewer abdominal masses and intestinal or urinary obstruction. These are caused by hydrometrocolpos (19).

Other malformations may accompany congenital vaginal atresia. In a report, two 14 aged adolescent girls with congenital cervical agenesis combined vaginal atresia, representing a rare type of obstructive mullerian anomaly (20). In a study of Troiano et al. of 24 children with imperforate hymen and vaginal atresia, show urinary tract anomalies (21).

In a report of 39 cases, 10 patients had vaginal atresia in addition to cervical agenesis. One patient had a bicornuate uterus and double cervix, and three had imperforate hymen. However, the patients were spinal malformations free (22).

Distal vaginal atresia is characterized by the presence of an atretic lower segment of vagina and presence of the upper vagina, cervix and uterus. Patients usually present by lower abdominal pain and a pelviabdominal mass, and no vaginal opening. Endorectal and abdominal ultrasound, abdominal and endorectal can confirm the diagnosis (23).

Pelvic MRI can ascertain the presence of a vagina, cervix, and uterus. MRI can diagnose the urinary tract anomalies associated with atresia or other Mullerian anomalies. MRI can also help in planning surgery by measuring the distance from the perineum to the vaginal bulge. (24).

Management of Distal Vaginal Atresia

A Pull-through vaginoplasty is usually performed for distal vaginal atresia. There is no agreement on the optimal surgical method in the literature. This is explained by different patient presentations, the relative rarity of the condition, and associations of different anomalies. Postoperatively, patients may experience infection, graft failure, vaginal restenosis, fistula formation, and injury to surrounding structures. (24)

Several techniques were described of pull-through vaginoplasty that ensure the anastomosing the vaginal mucosa to the perineum. The commonly used method is the perineal approach; where a crescentic incision at the hymen or on the introitus, with pediculus dissection of planes between the bladder and the rectum, creating a space

in the vagina till reaching to the cervix. This is followed by simple anastomosis to the perineum. An additional graft may be needed using the bowel or skin for higher atresia. (25,26) Kresowik et al. reported using ultrasound assistance in the dissection and using a posterior. (27)

Nikolaev and Bizhanova reported dissection in high atresias with an incision on the perineum (H-shaped incision) with a posterior and anterior U-shaped flap. (28)

The combined abdominoperineal approach can be done either by laparotomy or laparoscopy. These approaches help dissect the proximal vagina or push an instrument through the vagina from the abdomen to help in vaginal dissection. To prevent recurrence, postoperative therapies, vaginal stents, or dilators are used, with no agreement on their duration of use. (29,30)

In a retrospective Study of Sixteen patients with pull-through vaginoplasty done in one center with the same operative technique; The average distance from the perineum to the highest point of the atretic vagina was 1.84 ± 1.2 cm. In this study, 2 patients with atretic length greater than 3 cm, had postoperative vaginal restenosis, 4 patients had had postoperative vaginitis. Only 1 patient experienced UTI. (25).

ETHICS APPROVAL

Not required.

CONSENT FOR PUBLICATION

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

COMPETING INTERESTS

The authors report there are no competing interests to declare.

AVAILABILITY OF DATA AND MATERI- ALS

All Data are available from the corresponding author on reasonable request.

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