Neovagina Using Omental Flap in Patient with Mullerian Aplasia

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Abstract: Background: Vaginal agenesis is characterized by the absence or hypoplasia of the uterus, proximal vagina and the fallopian tubes. This anomaly has been recently termed müllerian aplasia by the American College of Obstetricians and Gynecologist[3]. Müllerian aplasia is an uncommon, but not rare, anomaly. The estimated incidence is 1 in 5000 newborn females[1,3]. Case: A 25 years old divorced female presents with history of primary amenorrhea. She was diagnosed prior to getting married with absence of the vagina along with the right kidney as case of Mayer–Rokitansky-Kuster-Hauser syndrome. The patient had a neovagina by omental flap. After six months of follow up she did well with an excellent anatomical outcome.
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1. Introduction

Vaginal agenesis is characterized by the absence or hypoplasia of the uterus, proximal vagina and the fallopian tubes. Müllerian aplasia is an uncommon, but not rare, anomaly. It occurs in approximately 1 in 5000 newborn female [1,3].

The general obstetrician and gynecologist can expect to encounter this condition once or twice during their career. Multiple variants; some with complicated associated anomalies, have been reported. In approximately 7-10% of women with müllerian aplasia; a normal but obstructed uterus, or a rudimentary uterus with functional endometrium is present [2].

Müllerian aplasia can be partial or complete. Partial müllerian aplasia is rarely encountered and is characterized by a normal uterus and small vaginal pouch distal to the cervix. Complete müllerian aplasia (MRKH syndrome) is the most common variant encountered and it is characterized by congenital absence of the vagina and the uterus in 90-95% of cases. The fallopian tubes maybe present but the ovaries have normal endocrine and oocyte function.

Müllerian aplasia can be an isolated finding although associated anomalies often coexist. The incidence of associated urologic abnormalities ranges between 15-40%, and the skeletal anomalies such as congenital fusion or absence of vertebra, occur in approximately 12-50% of cases [4]. An association between MRKH syndrome and Klippel-Feil syndrome has been reported. This syndrome is characterized by congenital fusion of the cervical spine, a short neck, a low posterior hairline, and limited range of motion in the cervical spine [5]. Infrequently, auditory deficits, cardiac defects, and other abnormalities can be found.

2. The case

A 25 years old divorced female presented with a history of primary amenorrhea. She was diagnosed prior of getting married with absence of the vagina along with the right kidney. She complained of cyclical abdominal pain and mastodynia and she did not complain of any gastrointestinal or urinary symptoms . In physical examination she had normal secondary sexual characteristics, examination of the external genitalia was unremarkable. However she had 2 cm blind vaginal pouch and the uterus was not felt.

MRI abdomen and pelvis showed absence of the upper two-third of the vagina with very small pouch with absence of the uterus and right kidney. Chromosomal analysis and the hormonal profile were normal. The condition was explained to the patient and the surgical procedure (Neovagina with omental flap) was discussed in details including the all possible complications, post-operative care and the usage of dilator was reinforced. An informed consent was signed.

Operative procedure:

In the lithotomy position a mid-line incision was used, omental J flap was created. The original vagina (2-3cm) was identified and incised transversely.

Dermatome was used and a split-thickness skin graft was taken from the lateral aspect of the thigh. The omental flap was carpeted by the skin graft using interrupted 3-0 vicryl sutures. The distal portion of the flap was rolled into a cylinder. The lateral wall of the cylinder was sutured with interrupted 3-0 vicryl. The wall of the omental cylinder was sutured to the vaginal introitus with interrupted 3-0 Vicryl sutures. A vaginal form had been shaped to an appropriate size, length, and diameter. It was introduced and laid on the new vagina (into a latex condom), The labia majora is approximated loosely by several 2-0 nylon sutures. These remain in place for 10 days. On the tenth postoperative day the patient was returned to the operating room for an examination under anesthesia. The vulvar sutures and vaginal form were removed, and the graft covering the neovagina was inspected. The form was returned into the neovagina.

Post Operative Care And Fellow Up:

The patient was fitted with a soft vaginal form that was worn for approximately 6 months. She would wash it and change the condom once or twice a day.

A dynamic pelvic MRI was done to assess the graft and the perfusion at 3 months post operative and it was normal with excellent perfusion.

3. Discussion

The etiology of müllerian aplasia is unclear. The wide spectrum of anomalies encountered in this anomaly suggests a field defect involving closely related structures derived from intermediate mesoderm. Most cases occur sporadically, although the rising number of reported familial cases indicates a genetic etiology. The karyotype of females having müllerian aplasia is 46, XX. Approximately 4% of reported cases are familial, with affected siblings, and in some cases it is transmitted as an autosomal dominant trait[6]. Müllerian aplasia is the second most common cause of primary amenorrhea in adolescents and usually diagnosed at puberty when adolescents present to the gynecologist with primary amenorrhea[7]. Physical examination reveals normal growth and development with age-appropriate secondary sexual characteristics. External genitalia are normal. Pelvic examination often reveals a patulous urethra. Ultrasonographic findings can add support to the clinical findings suggesting the absence of uterus and fallopian tubes in the presence of normal ovaries. MRI is extremely useful; absence of the vagina and uterus on a technically adequate image confirms the diagnosis of agenesis hypoplasia[8]. This modality can also detect a rudimentary uterus and any coexisting renal abnormalities. The hormonal profile is that of a normal female with age-appropriate luteinizing hormone, follicle-stimulating hormone, estradiol, and testosterone levels. This profile helps to distinguish the MRKH syndrome from androgen insensitivity syndrome in which post pubertal testosterone is elevated. Surgical and nonsurgical methods of treatment have been used. The nonsurgical approach relies on graduated dilators that progressively create a neovagina. This method may take several months or a few years before a functional vagina is formed. Surgery remains the most effective method of treatment for müllerian aplasia. Choosing the proper time to perform a vaginoplasty is of paramount importance. Surgical treatment should be considered only when the patient can participate in the decision making wishes to become sexually active and is highly motivated to use a vaginal prosthesis for several months after surgery[9,10]. Psychological support and counseling are essential components of the preoperative evaluation and care for many reasons. Since the ovaries are normal, oocyte harvesting can be conducted so that these women can have children with a surrogate, and these young women should be counseled about various reproductive options. Routine preoperative evaluation should include intravenous pyelography (IVP) and renal sonography to exclude urinary tract anomalies. Associated skeletal defects can be detected by reviewing images from IVP radiographic studies for vertebral abnormalities. The aim of surgical treatment is to create a neovagina. While several vaginoplasty methods have been developed, refined, and modified; no definitive surgical approach has been established. This is due to a number of factors including regional differences, the surgeon's experience and preference for a method, and the patient's choice[11]. Often, the procedure must be individualized. The strategy for vaginoplasty is to develop a space between the bladder and the rectum. In some approaches, a stent or form is placed in the newly created space to ensure patency while healing occurs. Skin grafting remains the most popular material used in vaginoplasties; however, scar formation at the graft site has been a concern[12].

Transposition flaps were described in 2 reports. In 1 method, a de-epithelialized vulvar transposition flap was used as the graft, and in the other, a pudendal thigh fasciocutaneous flap was described. Both authors reported good cosmetic and functional results[13,14]. Autologous buccal mucosa has also been used as a graft source. The use of artificial dermis and absorbable adhesion barriers show promise as exogenous graft sources in vaginal reconstruction. In a case study by Noguchi and associates, a modified Warton method was used to create an artificial space between the bladder and rectum The authors reported good results with continued use of the form at nighttime[15].

Motoyama and colleagues described a similar technique using absorbable adhesion barrier (Interceed; Ethicon, Somerville, NJ). They reported neovaginal epithelization in 1-4 months. All 10 patients were

satisfied with the results[16]. Some pediatric surgeons prefer using a bowel segment in place of a skin graft. Indeed, bowel vaginoplasty involving the distal sigmoid colon to line the neovaginal canal has gained popularity in some centers. This approach usually requires concomitant laparotomy and bowel anastomosis, although recent reports describe a laparoscopic approach[18]. The modified McIndoe procedure remains the most common surgical approach to vaginoplasty. The Williams vulvovaginoplasty is an alternative to the McIndoe procedure. This procedure is particularly useful for patients with previously failed vaginoplasty or patients who have undergone radical pelvic surgery[19]. Alternative vaginoplasty methods have been developed and refined and are achieving increasing success. The laparoscopic approach has augmented surgical approaches for neovaginal creation[20]. In 1969, **Davydov** described a laparotomy procedure in which peritoneum from the uterorectal space (Pouch of Douglas) was advanced in such manner that a vaginal canal was created. Laparoscopic modifications were later developed and continue to be refined. The modifications use laparoscopy to mobilize the bladder peritoneum, lateral pelvic side walls and the sigmoid colon. Authors report several benefits of this procedure, including minimal scarring and functional vaginas associated with comfortable intercourse[21]. In the Department of Pediatric Surgery, University of Bologna[22] over 34 years, they evaluated 47 patients. The mean observation period was 12 years. Forty-six patients were treated with vaginal reconstruction by interposition of sigmoid colon. Only in 1 case they performed a vaginal construction with an ileal loop. The outcome for 47 patients was excellent: 18 were sexually active and 4 married. Complications occurred in 17 cases: in 1 patient a necrosis of the replaced vagina occurred, another patient had an abdominal abscess. In 12 cases a second procedure was required: 6 had stenotic new-vaginal introitus, 4 had new-vaginal prolapse, and 2 had intestinal obstruction. In the Department of Gynecology and Obstetrics, Faculty of Medicine, Yeditepe University, Istanbul, Turkey[23] they investigated the long-term effects of intestinal vaginoplasty in cases with Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome. 29 patients with MRKH syndrome underwent intestinal vaginoplasty. Two of the patients were treated with ileal and 27 with sigmoid vaginoplasty. One of the patients for whom ileal vaginoplasty was performed had 40 cm ileal necrosis requiring bilateral ileostomy for 2 months. Introital stenosis was detected in 15 cases who were unmarried, while none of the married cases had introital stenosis. However, all patients responded to finger-dilatation. All married patients were sexually satisfied after operation. In one patient an intraluminal abscess developed in the proximal segment of the neovagina, in another patient who had a rudimentary uterine horn, hematometra developed 3 years after operation[23].

In department of Pediatric Surgery, Necker-Enfants Malades Hospital, Paris, France[24] they followed 26 patients with the Rokitansky syndrome, vaginoplasty was performed in 23 patients. It was performed at a mean age of 16 years (range 10.3 to 18.8). Median postoperative follow up was 3.4 years. Postoperative complications included lower extremity compartment syndrome (1), pelvic hematoma (1), mucosal prolapse (2), cystitis (2) and introital stenosis (1). Of the 23 patients undergoing surgery 9 (39%) had an active sex life postoperatively. They believe that the preferable technique for vaginal replacement is the use of intestinal conduit. The sigmoid colon is the best intestinal tract to be used owing to its size, location and preserved blood supply. They consider ileal loop as a good alternative when the sigmoid colon is not available[22-24].

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Page 30 of 40
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