Vaginal Agenesis in Mayer Rokitansky Kuster Hauser Syndrome

Lt Col A Menon*, Col M Biswas⁺, Col SPS Kochar[#], Col H Bal^{**}, Maj R Abhichandani⁺⁺, Maj S Singh^{##}

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Introduction

The Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is characterized by congenital aplasia of the uterus and the upper part (2/3) of the vagina. The first sign of MRKH syndrome is primary amenorrhea in young women presenting otherwise with normal development of secondary sexual characteristics and normal external genitalia, with normal and functional ovaries and karyotype 46, XX [1].

The vaginal agenesis in this syndrome can be treated by various methods, both surgical and nonsurgical. We present a case treated by creating a neovagina using amnion as a graft for the vaginoplasty.

Case report

A 25 year old girl reported to the OPD with history of primary amenorrhea. On clinical examination she was found to have well developed secondary sexual characteristics and a small blind vaginal dimple. Ultrasonography and magnetic resonance imaging (MRI) revealed the absence of an uterus but ovaries were reported to be present. An intravenous urogram (IVU) did not reveal any renal abnormalities, but radiological examination of the spine revealed skeletal deformities in the form of fused vertebra (C7) and a cervical rib.

Based on these clinical and radiological findings a diagnosis of MRKH syndrome was made. As the patient was psychologically mature and contemplating marriage, she was offered treatment in the form of creating a neovagina. She was taken up for the modified Abbe McIndoe procedure under spinal anaesthesia. The procedure consisted of prosthesis assembly, creating a new space between the rectum and the vagina and placing the graft prosthesis assembly in the neovagina. The prosthesis was made with a 10x10cm sponge which was sterilized and tightly rolled around a silicon Malecot catheter and the ends sutured (Fig.1). Two condoms were placed over this mould and tied at the lower end. Amnion was washed thoroughly in saline mixed with antibiotic several

times and secured around the mould using 3-0 catgut.

The patient was placed in dorsal lithotomy position and catheterised to define the anterior urethra bladder boundary. The rectum was tamponaded with No.11 metal Hawkin Ambler's dilator. Dissection was initiated in the fibro connective tissue on either side of the midline creating a space between the bladder and rectum by inserting progressively thicker Hawkins Ambler's dilators. This was done for a depth of around 4 cms which was the thickness of the intervening tissue between the perineum and pelvic peritoneum as determined by MRI. The dissection was aided by placing a laparoscope in the abdominal cavity to ensure maximum length of area dissected and to prevent any inadvertent breaching of the peritoneum. Once the space created was deemed to be adequate, the intervening median raphe was excised using cautery.

After ensuring hemostasis, the amnion covered mould was placed in the neovagina. The labia minora were now sutured to keep the mould in position. Postoperatively, the patient was kept on intravenous and oral fluids, antibiotics and a low residue diet for a period of 72 hours. After six days, the labial sutures were removed and the mould gently withdrawn. The neovagina was irrigated with saline and povidone-iodine and inspected which revealed that the graft had taken well. Two similar soft moulds were prepared without the catheter and amnion and were changed at 48 hour intervals thrice.

The patient was trained to use an acrylic mould after two weeks and further recovery was uneventful. She was discharged after three weeks and is on follow up. The accompanying photograph (Fig.2) shows the neovagina with a vaginal speculum placed inside it showing the depth to be around 4 inches which is likely to improve with use of the mould and later by regular intercourse. The patient and her family was further counselled regarding benefits of an early marriage after the surgery. It was also made clear to them that she would never be able to bear a child, but a genetic child was possible through surrogacy.

*Reader, ⁺ Reader, [#] Professor & Head, ^{**}Associate Professor, ⁺⁺Clinical Tutor, ^{##}Resident (Department of Obstetric & Gynaecology), AFMC, Pune.

Fig. 1 : Pre-op, before inserting mould

Discussion

Isolated uterovaginal aplasia is referred to as Type I MRKH which is less common than Type II MRKH, also known as MURCS (Mullerian, Renal Cervical Somite). The latter is associated with (a) Renal (unilateral agenesis, ectopia of kidneys or horseshoe kidney) (b) skeletal and vertebral (Klippel-Feil anomaly; fused vertebrae, scoliosis) or (c) hearing defects [1]. The incidence of MRKH syndrome has been estimated as 1 in 4500 female births [2]. Our case fits in Type II MRKH syndrome.

The clinical picture is typically that of a girl with well developed secondary sexual characteristics with no androgen excess, normal karyotype and primary amenorrhea with the vagina reduced more or less to a dimple [3]. Diagnostic imaging includes USG and MRI, the latter being a gold standard [1].

Treatment consisting of creating a neovagina must be offered to patients only when they are ready to start sexual activity. Of the two main types of procedures, the first one consists of the creation of a new cavity and can be nonsurgical or surgical. The second is vaginal replacement with a pre-existing canal lined with a mucous membrane like bowel [4].

Our patient, on the threshold of marriage was an ideal candidate for this surgery. The Abbe McIndoe procedure, adopted by us to create the neovagina, first described in 1888, is still the most effective and preferred one although numerous methods have been described since then. The original surgery used split skin or full thickness skin grafts to line the newly created vagina. Various alternatives like amnion [5], peritoneum, minora labia grafting, or synthetic materials [6] have since been used.

Laparoscopic Vecchietti procedure aims to create a neovagina by invagination by using an acrylic olive mould that is placed against the vaginal dimple. The olive is

Fig. 2 : 10th post-op day

attached to a traction device mounted on the abdomen with laparoscopically placed sub peritoneal sutures. Then, traction is applied to the olive to produce 1.0-1.5 cm of invagination per day, creating a neovagina in approximately 7-9 days.

No consensus has been reached regarding the ideal method for creating a functional vagina. At present, the most common operation is McIndoe vaginoplasty. Though noninvasive laparoscopic surgery seems promising, additional data is required for it to gain general acceptance [7].

Conflicts of Interest

None identified

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