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Sigmoid Colon Vaginoplasty in a Mayer-Rokitansky-Kuster-Hauser (Mrkh) Syndrome Patient with Situs Inversus Totalis and Review

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Abstract

A 20 year-old-lady from suburban area presented to us with primary amenorrhea. On examination and investigation she was found to have MRKH syndrome with Situs inversus totalis. We did sigmoid colon vaginoplasty. This is one of the uncommon anatomical variants we encountered in our series of cases. Our literature search did not reveal such patient undergoing sigmoid vaginoplasty and so is this case presented for publication.

Keywords

MRKH; Sigmoid vaginoplasty; Situs inversus totalis; Primary amenorrhea

Introduction

MRKH is congenital malformation occurs due to failure of Mullarian duct to develop resulting in absent uterus and variable degree of vaginal hypoplasia, it occurs at a frequency of 1 in 4500 female births. Situs inversus totalis is a rare autosomal recessive condition with complete transposition of abdominal and thoracic organs, it occurs at a frequency of 1 in 10000 births. However, situs inverses totalis with MRKH is a rare entity. There are a few case reports published in the literature about the mere occurrence of these two conditions together. We discuss the sigmoid vaginoplasty done on this patient and the follow up. This case is presented here to bring to light of this unique combination of genital and gastrointestinal anomalies and its management. And to bring reassurance that bowel vaginoplasty is also an option in these patients.

Case Capsule

A 20 year-old-lady from suburban area presented to us with primary amenorrhea. She did not have history of auditory or skeletal abnormalities. She had her first consultation for the same problem at the age of 16 years. An ultrasound was done showed absent uterus and normal ovaries. She was advised a Magnetic Resonance (MR) imaging for further clarification. At that point, she could not get this done due to financial and social circumstances. On examination, she had fully developed secondary sexual characteristics. Perineal examination revealed a shallow pit of one centimeter in the place of vagina. Karyotyping was 46XX; FSH and LH levels were normal. A MR imaging done now showed transposition of intra-abdominal viscera and dextrocardia-situs inversus totalis; horseshoe kidney and absent left ovary and the uterus (Figure 1).

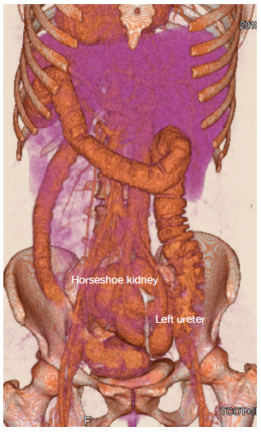

Figure 1: MRI of the abdomen and thorax showing situs inversus totalis, horseshoe pelvic kidney and the left ureter.

The absence of left ovary was not detected on Ultrasonography probably due the presence of Horseshoe Kidney. As the patient has situs inversus totalis, we decided to do an arteriogram to look for the inferior mesenteric arterial arcade. The vascular pattern was an exact mirror image of the normal pattern (Figure 2).

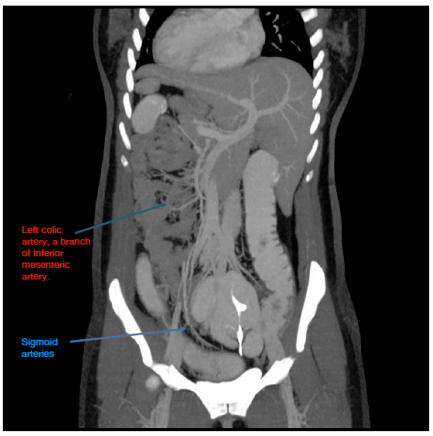

Figure 2: CT aortogram showing the mirror image of the vascular architecture. The left colic artery and sigmoid arteries, the branch of Inferior mesenteric artery are on the right side.

The patient was informed about non-operative vaginal dilatation and surgical management in the form of sigmoid colon vaginoplasty. She was started on less morbid vaginal dilatation first; however, she was not compliant with this procedure due to persistent pain on

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dilatation; so, the decision was made for sigmoid colon vaginoplasty. After thorough counseling with the patient and the parents, we prepared her bowel and took her for bowel vaginoplasty. There were a few modifications done to perform surgery on this patient. The operating surgeon was standing on the right side of the patient at the beginning of the surgery. A vertical midline below umbilical incision was made. The pelvis was inspected and there was normal looking right ovary above the pelvic brim and the left ovary was absent. The uterus was rudimentary. There was transposition of rest of the abdominal organs. A tunnel was created in the recto-vesical channel. The tunnel was made between an incision made just below the uterus from inside and dissection in the center of the vaginal pit outside. The track was dilated for its entire length using the 16 size Hegar dilaters. At this juncture, the operating surgeon changed his place to left side of the Patient. This was for the ease of operating on the descending and sigmoid colon, which were on the right side of the abdomen and for better visualization of the inferior mesenteric vascular arcade from the opposite side. The descending colon was mobilized from the splenic flexure. The sigmoid colon was severed beneath the left colic artery and at the level of the pelvic brim (Figure 3).

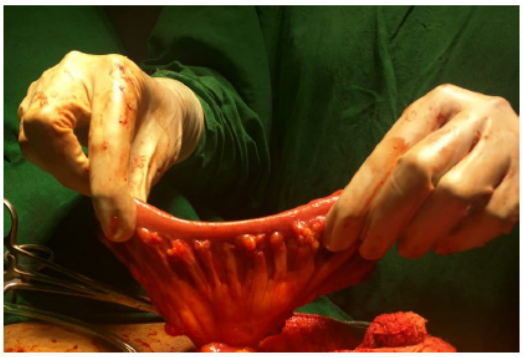

Figure 3: Sigmoid colon graft based on the sigmoid vessels is ready for pull through as neo-vagina.

The length of the graft was 13 cms. Few proximal sigmoid vessels supplying graft were tied to gain more length. The distal end of the sigmoid colon was closed using the absorbable sutures. The proximal end now rotated at 180 degrees on the sigmoid vascular pedical (inverted pedicle graft). The open proximal end is brought out of the vaginal pit in the Introits and anastomosed circumferentially using absorbable sutures. The descending colon was suture anastomosed to the rectum after mobilization of splenic flexure and the abdomen was closed. The operating surgeon again moved back to the right of the patient for abdominal closure. The post-operative period was uneventful. She was started on feeds from the 4th day of surgery and was discharged on the 6th day of surgery. She was started on dilatation on the second postoperative week and was advised to do it once a day. Vaginal irrigation was advised once a day-only on the day of mucus discharges. She is on regular follow-up with no other complaints; her vagina admits two fingers and is about 10 cms long. The patient is not sexually active yet.

Discussion

Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is a rare congenital anomaly with a genotype of 46XX. Here we see aplasia of the uterus and vagina with normal ovaries and normal development of secondary sexual characteristics. MRKH presents in two types. Type 1 (Rokitansky sequence) is Mullarian duct aplasia in isolation. Type 2 or MURCS

(Mullarian renal cervical somite) association. The MURCS association has the following manifestations - genital, renal (unilateral absent kidneys, ectopic kidneys, horseshoe kidneys), and skeletal defects (kypho-scoliosis, fused cervical vertebrae, stapedial ankylosis) [1]. MRKH type 2 is thought to be the more severe form of the MRKH spectrum [2]. This disease when occurs in familial settings, is inherited as Autosomal dominant pattern. The disease presents with incomplete penetrance and variable expressivity. Incomplete or reduced penetrance means the genetic trait is expressed in only part of the population. Variable expressivity refers to the range of signs and symptoms that can occur in different people with same genetic population. The Situs Inversus Totalis (SIT) is seen in MRKH type 2 or MURCS association [2]. These defects suggest the involvement of the homeobox (HOX) genes. HOX genes play a key role during the embryogenesis of axial skeleton, hind brain and urogenital system [3].

However, we could not find skeletal association in our patient. The SIT is also called the "mirror man", where the internal organs of body are rotated by 180 degrees. It is called "totalis" when even the heart is on the right side. Surgical diagnosis and surgical procedures in these patients is challenging due to the mirror image anatomy. This degree of difficulty is more pronounced while doing laparoscopic surgeries, especially colon resections that have been seen sparsely. In patients with SIT, performing laparoscopic surgeries may not be easy even for an experienced surgeon. During dissection, clipping and use of laparoscopic instruments including the stapler by the left hand of the surgeon may be a limiting factor for right-handed surgeons. That means to say surgeon's dominant hand is passive, and non-dominant hand is active during dissection [4]. This problem can be averted in open surgery by changing the side of operating by the surgeon. Other important aspect of situs inversus is the associated vascular anomalies [5]. This made us to do the angiography in our patient to have prior vascular anatomy knowledge. Intra operatively, the vascular arcades looked better from the left side of the patient. We did not encounter any problems during the procedure.

Very important aspect of treatment of MRKH is age of the patient. Treatment of MRKH is a multi-departmental approach. The non-operative Frank and Ingram technique of progressive pressure onto the vaginal pit using dilators has been less popular due to its unsatisfactory results and poor compliance [6].

Many operative (McIndoe's, William's, Vecchieti's,) methods of vaginal replacement have been explained, McIndoe's technique, where the skin grafts are placed between the urethra and the rectum by blunt dissection in this area. William's vaginoplasty involves a vulvar flap to make the vaginal tube. The Vecchieti technique is based on stretching of the vaginal dimple *via* acrylic mould passed in the neo-vaginal space and pulled by sub-peritoneal threads. The vagina will lengthen over a period of days to weeks [6]. Bowel vaginoplasty is preferred due to reasons like–minimal likelihood of "poor take" or later contraction because of vascularity and epithelial lining, easy post-operative management with minimal or no dilatation, early coitus, spontaneous mucous production mimicking normal vagina, greater resistance of sigmoid colon epithelium and importantly less long term care [7]. Nonetheless, it is involves risks of major intestinal surgery and anastomosis. However, a team approach, pre-op bowel

preparation and meticulous attention into minor details can avoid this problem. The sigmoid vaginoplasty has stood the test of time. It is popular because of its proximity and easily mobilized vascular pedicle [8]. The disadvantage with this technique is its pre-operative preparation, invasiveness and bowel complications like anastomotic leak, mechanical bowel obstruction, and diversion colitis6. Definitely, there is nothing to match characters of the normal vagina; however sigmoid colon has many similarities compared to normal vagina. The sigmoid colon can be harvested either in isoperistaltic or anti peristaltic manner. Few studies have also shown the advantages of ileum over sigmoid colon - no feculent smell and diversion colitis6. The diversion colitis is a problem in colonic diversions due to lack of short chain fatty acids and colonic bacteria of feces in the diverted segment [6].

Selecting the part of the intestine (Ilium *vs* Sigmoid) is more to do with surgeon's comfort and familiarity with the procedure. We have been doing Sigmoid vaginal replacements in all our series of patients. To avoid injury to the rectum and the urinary bladder - we start the abdominal dissection first by following the utero-sacral ligaments to the rudimentary uterus. Then the vaginal dissection started. Laparoscopic sigmoid vaginoplasty has its own advantages like better cosmetic results, less invasiveness and early recovery. However, the disadvantage of Laparoscopy being long learning curve and long operating time [4,6]. Laparoscopy was not attempted in our patient, as there was anticipated technical difficulty in completing the procedure due to SIT. Various methods have been explained on harvesting the sigmoid graft based on the vascular pedicle; what we did was the inverted sigmoid colon graft based on sigmoid arteries. The other disadvantage of sigmoid vaginoplasty is prolonged periods of fasting due to bowel preparation before surgery and bowel anastomosis. Recently many centers are using Enhanced Recovery After Surgery (ERAS) protocols - fast track bowel regimes have been used for early recuperation from this major surgery [9]. We counseled the patient regarding this; however patient was apprehensive about her condition and she did not give consent for ERAS protocol to be followed.

In rare instances, MRKH mothers can have children with their oocytes being harvested, fertilized and implanted in a surrogate mother. This procedure is expensive and has practical difficulties and has an average success rate of 30%. It is also important that these mothers discuss with geneticist before they decide to conceive.

Conclusion

MRKH with SIT is a rare congenital anomaly. Operating on this unique combination of anomalies needs prior vascular anatomy knowledge for mental orientation. Sigmoid vaginoplasty can still be done safely in these patients.

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