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Vaginal Reconstruction Using Sigmoid Colon in Type II Mayer-Rokitansky-Kuster-Hauser Syndrome-: A Rare Case with Real Challenge

Dr. Vishwajeet Singh [1]*, Dr. Mohit Pandey [2], Dr. Mohd Rehan Akhtar [3], Dr. Krishna Kumar Bhandari [4], Mr. Mukul Kumar Singh [5], Mr. Anil Kumar [6]

1. Professor, Department of Urology, King George's Medical University, Chowk, Lucknow, Uttar Pradesh, India.

2. Assistant Professor, Department of Radio-Diagnosis, T.S Misra Medical College & Hospital, Amausi, Lucknow.

3,4. Senior Resident, Department of Urology, King George's Medical University, Chowk, Lucknow, Uttar Pradesh, India.

5,6. PhD Scholar, Department of Urology, King George's Medical University, Chowk, Lucknow, Uttar Pradesh, India.

Corresponding Author: Dr. Vishwajeet Singh, Professor, Department of Urology, King George's Medical University, Chowk, Lucknow, Uttar Pradesh, India, 226003.

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Abstract

An 18 years young female presented with primary amenorrhea. She was investigated and found to have type II Mayer-Rokitansky-Kuster-Hauser syndrome. The patient had an atrophic uterus with vaginal agenesis, left upper limb polydactyl, vertebral anomalies, solitary ectopic pelvic malrotated left kidney, and aortic regurgitation. Her vaginal reconstruction was done by using a sigmoid colon. The procedure was uneventful. The patient is sexually active and doing well in 2 years of follow-up.

Keywords: Mayer-Rokitansky-Kuster-Hauser syndrome, polydactyl, uterus, reconstruction

Introduction

Vaginal agenesis is an integral part of Mayer-Rokitanski-Kuster-Hauser Syndrome. The other components are uterine aplasia with varying degrees of renal and skeletal abnormalities [1]. There are two types of MRKH syndrome. Type I MRKH syndrome is characterized by uterine and vaginal atresia but the ovaries are normal. The lower one-third of the vagina may be in form of a shallow pit or there may be complete atresia. Type II MRKHS is characterized by uterine and vaginal atresia with renal, vertebral, skeletal, auditory, and cardiac anomalies [2]. The most common presentation in MRKH syndrome is primary Amenorrhea due to agenesis/atretic uterus. Vaginal reconstruction in a such patients is needed at puberty when they become emotionally mature and sexually active [3]. We present a case of type II MRKH syndrome in which vaginal reconstruction was carried out by using a sigmoid colon. The patient is sexually active and doing well in 2 years of follow-up.

Case History

An 18 years young female presented with primary amenorrhea. Her general physical examination revealed a height of 142 cm, a body weight of 45 Kg with BMI of 31.69. The secondary sexual characters were normal. The examination of the perineum showed normal labia with a 1cm shallow dimple at the site of the vagina with non-canalization of the vagina. She had a short neck with polydactyly of the left upper limb. Her routine biochemical examination was normal. Her karyotype was 46XX. The ultrasound of the abdomen and pelvis showed an atrophic uterus with normal bilateral ovaries. There was a solitary ectopic pelvic malrotated left kidney. The Echocardiography revealed

moderate aortic regurgitation. The plain radiographic of the left upper limb showed polydactyly (Figure. 1). The X-ray spine showed fused L2, L3, and L4 vertebrae. The Contrast-enhanced CT scan abdomen & pelvis showed an atrophic uterus with an ectopic malrotated solitary kidney lying in the pelvis on the left side (Figure. 2). There was a complete absence of a vagina. She was subjected to vaginal reconstruction using a sigmoid colon. Lower abdomen transverse laparotomy through Pfannenstiel incision was performed. The findings were an atrophic uterus with normal ovaries. The cervix was also hypoplastic and blind ending with complete vaginal agenesis. This atrophic uterus was removed. A 12 cm mid-sigmoid colon was isolated based on sigmoid mesenteric vessels (Figure 3a). End-to-end colo-colostomy was performed. The isolated sigmoid segment was used as a vaginal conduit. The upper end of the sigmoid conduit was closed by 3-0 polyglactin in two layers and fixed to the sacral promontory. The distal end of the conduit was brought out in the vestibule through a plane created between the bladder and rectum. The margins of the vaginal conduit were sutured to mucocutaneous margins at the vestibule by 3-0 polyglactin and neovagina reconstruction was completed (Figure. 3b). A soft vaginal mold was kept in the vaginal lumen for 24 hours. The patient is doing well within 2 years of follow-up and is sexually active.

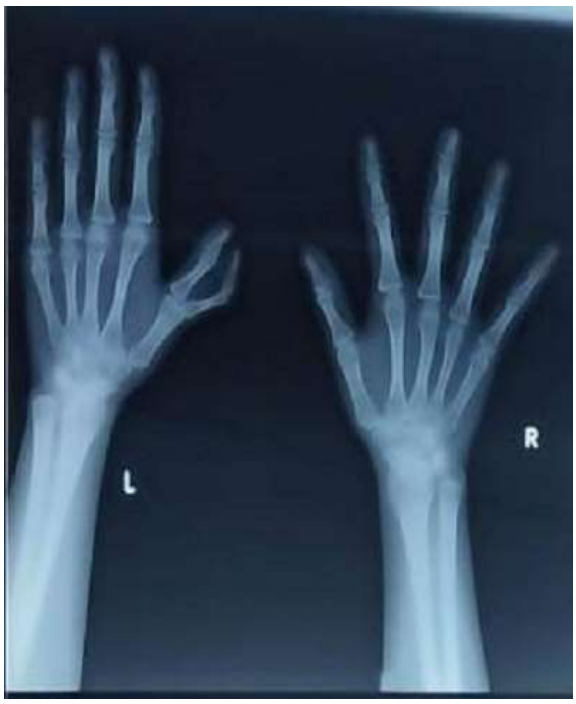

Figure 1: Plain radiographic of bilateral upper limb showing polydactyly

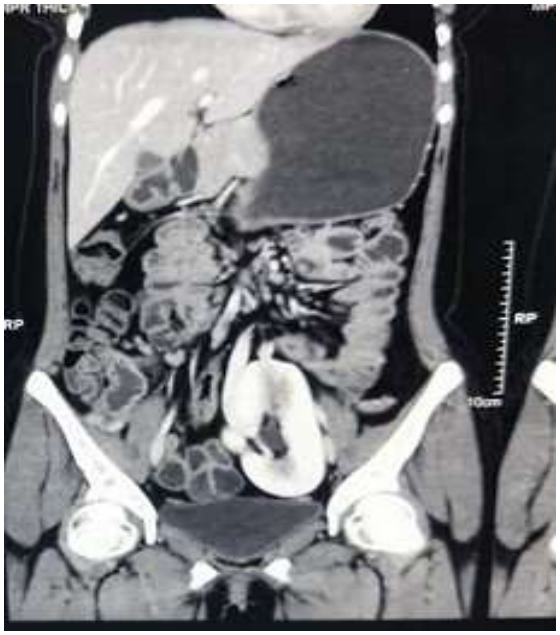

Figure 2: CT scan showing atrophic uterus with ectopic malrotated solitary kidney lying in pelvis on the left side.

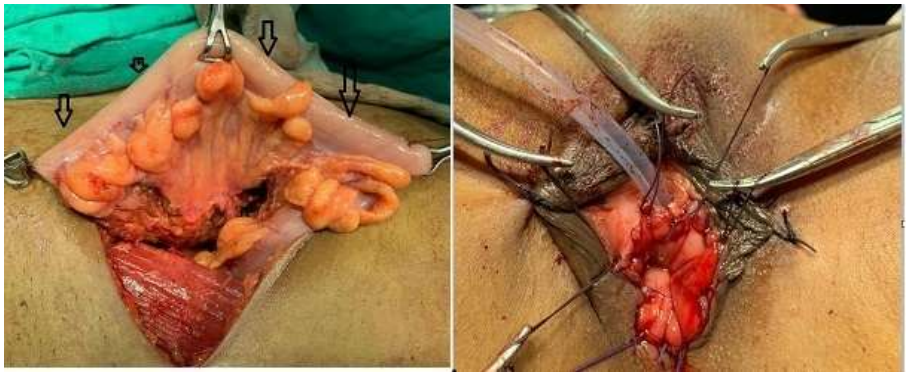

Figure 3: **(a)** 12 cm mid segment of sigmoid colon **(b)** Vaginal opening at vestibule

Discussion

Mayer-Rokitanski-Kuster-Hauser syndrome is characterized by primary amenorrhea with normal secondary sexual characteristics having 46XX karyotype with noncanalization of the vagina. The ovaries are normal but the uterus is hypoplastic or atretic [1]. There are two types of MRKHS. Type I is more common and usually has a small atretic uterus with vaginal agenesis. Type II MRKHS is relatively rare and has more associated anomalies such as short stature, short neck, auditory, cardiac, skeletal, vertebral, and renal malformations [2]. Our patient had short stature of 142 cm with a short neck and skeletal and vertebral anomalies. The patient was expecting to get married and worried about

her sexual function because of the absence of a vagina. She had moderate cardiac risk due to aortic regurgitation and laparotomy for sigmoid vaginoplasty was a real challenge for this patient. It is also challenging in the presence of a solitary ectopic malrotated left pelvic kidney. The dissection of the sigmoid colon and left ectopic pelvic kidney need a meticulous dissection to prevent any ureteric injury. The management of such patients needs a multidisciplinary approach involving a radiologist, anesthetist, gynecologist, urologist, psychologist, and cardiologist. The aim of creating a neovagina either by non-operative or operative methods must have a vaginal length of more than 7 cm with girth (more than 3 cm) admitting 2 fingers [3].

Vaginal dilatation is a nonoperative method of creating neovagina and has a success rate of 90-96%. It can be offered to patients who have vaginal dimples of at least 2 cm in the vestibule. A specially designed dilator is given to these patients who put in vaginal dimple for varying times (10-30 minutes) for 4-6 months. In our patient, she did not agree to accept this method of non-operative vaginoplasty. On counseling for various surgical vaginoplaties, she was willing for sigmoid neovagina for its advantages of being capacious with very few complications [3,4,5].

Sigmoid vaginoplasty has shown long-term results. It has the advantages of an adequate lumen with a thick wall and is less prone to trauma. The mucus secretion acts like natural lubrication. In sexually active patients it does not require any kind of dilatation. Our patient had type II MRKH syndrome and sigmoid vaginoplasty was a real challenge due to associated cardiac and renal anomalies [5,6].

Conclusion

The vaginal reconstruction using a sigmoid colon in type II MRKH syndrome is challenging due to associated anomalies. The patient is sexually active in follow-up without any complications.

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