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Colovaginoplasty for a Patient with Mayer-Rokitansky-Kuster-Hauser's Syndrome (Mrkh) our Experience in Federal Medical Center Katsina

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Key Clinical Message: Colovaginoplasty is a rewarding surgical procedure in returning the social life of the patient with Mayer-Rokitansky-Kuster-Hauster's syndrome, as it happens to our patient with the rare congenital absent vagina now happily married, after we did the successful surgery in our center.

1. Keywords: Colovaginoplasty; Mayer-Rokitansky-Kuster-Hauster; Syndrome

2. Introduction

The Mayer-Rokitansky-Kuster-Hauster (MRKH) Syndrome is a rare congenital mullerian duct agenesis with various degrees of clinical manifestations ranging from primary amenorrhea to complete absence of vagina, even though they may have secondary female sexual characteristics [1-3]. the incidence varies from region to region with different researchers reporting various figures from 1 in 4,000 to 1 in 10,000 female life births [4-6] some of the patients may present with a normal uterus, rudimentary bi-cornuate, with or without lumen or it may totally be absent as it occurs in our patient and most of the patients being 46XX have ovaries, they may have blind end or completely absent vagina [7]. They may present with associated renal anomalies such as malposition, horse shoe kidney or agenesis of the kidney and skeletal abnormalities are noted in 12% of patients such as primarily spine defects and limb and rib defects and auditory defects [7-9]our patient presented with a renal agenesis, treatment of these patients varies depending on the severity of the pathology, the expertise available and the patient choice. The treatment can be from serial vaginal dilatation, myocuteneous flaps, split thickness and full thickness skin grafts, using peritoneum or bladder mucosa and oxidized cellulose fabric to colovaginoplasty [10] .

Different segments of intestine have been used in vaginal reconstruction notably Baldwin was the first to describe the techniques of using sigmoid colon for colovaginoplasty, others described the use ofileum and cecum even though sigmoid colon is more preferred because of its anatomical proximity and easy mobile vascular pedicle [11-13].

3. Methods

The case note file of the patient was review.

4. Case Report

Our patient was 24 years old who presented with history of primary amenorrhea, there was no history of trauma or surgery in the perineum, no family history of similar problem, there was no history of lower urinary tract or gastrointestinal symptoms.

On examination she was found to have well developed breast bilaterally with female hair distribution, abdominal examination revealed normal gynecoid pelvis, normal female pubic hair distribution with well-developed labia and absent vagina with a vagina dimple about 2cm, Abdominal USS revealed hypoplastic uterus with left renal agenesis and the right kidney is normal in shape, size and location, her packed cell volume was 35% and her U,E&Cr were essentially normal, Urine MCS cultured no growth.

She was prepared and has surgery Intra operative findings were normal female urethra opening accommodating size 16 F Foley's catheter, normal bladder, absent vaginal canal with a blind enddimple (**Figure 1**). There was absent uterus and

fallopian tubes however the ovaries were present bilaterally (**Figure 2**).We mobilized about 15cm of the sigmoid colon and resected it (**Figure 3**) and colo-colic anastomosis was done to restore colonic continuity, the proximal end of the resected sigmoid colon was sutured (**Figure 4**). The distal segment of the sigmoid colon was full through the space created between the bladder anteriorly and the rectum posteriorly and sutured to the interoitus (**Figure 5**), the neovagina accommodated a vaginal dilator (**Figure 6**), and the neovagina was cleaned (**Figure 7**), she did well Post operatively, we started neovaginal daily dilatation and later the patient was doing it, she was discharge 2 weeks after the surgery and was on follow up 4 months post operatively the neovagina was completely healed (**Figure 8**) and she is presently happily married.

Figure 1: Absent vagina with a 2cm dimple

Figure 2: Absent uterus with bilateral ovaries present

Figure 3: 20cm of sigmoid colon resected

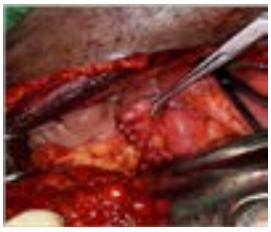

Figure 4: showing the closed proximal segment of the sigmoid colon

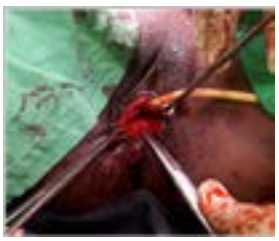

Figure 5: showing the distal sigmoid segment full through the space created and sutured.

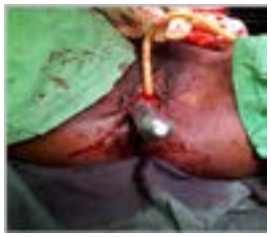

Figure 6: showing the neovaginal accommodating the vaginal dilator

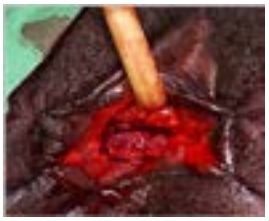

Figure 7: showing the neovaginal immediate post operatively

Figure 8: The Healed Neovagina 4 months post op

5. Discussion

The MRKH syndrome is a mullerian duct agenesis presenting with an absent vagina as it is the case of our patient who presented with an absent vagina with about 2cm dimple, as reported by Muhammad Saleem et al and Mungadi et al, our patient presented with normal female external secondary sexual characteristic as reported by Muhammad Saleem et al however Mungadi et al reported delay in secondary sexual characteristics, The MRKH Syndrome is classified into Type 1&2, Type 1 is when the patient presents with purely genital malformation, this type is also called isolated MRKH syndrome or Rokitansky sequence and Type 2 when the patient have associated Mullerian renal, cervicothoracic, somite abnormalities (MURCS) involving the Genital, Renal and Ear anomalies as described byet al [14,15] Our patient presented with Type 2 MRKH syndrome with associated left Renal agenesis, as reported by Patricia G. Oppelt et al were they found 18.7% of their patients with renal anomalies,

Peter Oppeltet al in another study found 32% of patients with associated renal anomalies while Lacey S. Williams 7 patients out of 52 cases studied with renal agenesis,[16-17]. However Muhammad Saleem et al reported a case of Type 1 MRKH with no associated renal, musculoskeletal or ear anomalies, as well as [18]et al who reported 4 patients with Type 1 MRKH syndrome and [19]et al found 4 out of 19 patients with Type 1 MRKH syndrome[18-19].

In a study by Wenqing Ma et al among the 182 unrelated Chinese patients 155 of the cases were found to have Type 1 MRKH syndrome. Our patient has a successful Sigmoid Colovaginoplasty and now happily married as reported by et al[21-22].

6. Conclusions

Sigmoid colovaginoplasty is one of the promising procedures in the long time management of patients with MRKH syndrome as was recorded in this 24 years old lady with Type 2 MRKH syndrome.

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