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Case Report

Presence of fibroids in the absence of uterus- Mayer-Rokitansky-Küster-Hauser syndrome with fibroids: a case report

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ABSTRACT

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is rare congenital anomaly with complete absence of uterus and upper part of vagina. Fibroids are one of the most common benign tumours arising in women. Fibroids arising from Mullerian remnants are very rare. These fibroids often pose difficulty in diagnosis and management. Here, we report a rare case of fibroids arising from Mullerian remnants in MRKH syndrome and the treatment provided for the patient.

Keywords: MRKH syndrome, Fibroids, Laparoscopic myomectomy, Davydov's vaginoplasty

INTRODUCTION

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome refers to congenital aplasia of uterus and upper part of vagina in women with normal secondary sexual characteristics and 46, XX karyotype. Often associated with defects in other organ systems. MRKH syndrome has been estimated to affect 1 in 4500 female births.[1] Uterine fibroids are the most common gynecological tumours, often requiring surgery when symptomatic. They may arise from any of the smooth muscles in the body, most commonly arising from smooth muscles in myometrium. Surgical removal constitutes the best treatment option when they are symptomatic.[2] In such cases, proper diagnosis and clinical management is a challenge. This case report discusses the clinical and surgical interventions taken in such a rare presentation.

CASE REPORT

A 32-year-old patient, presented with primary amenorrhoea, occasional abdominal cramps and dyspareunia. On examination, all secondary sexual characters present. Per speculum examination revealed a short vagina (~3 cm), with absence of cervix. Karyotype was consistent with female phenotype, 46XX. Hormonal assay was within normal limits. On magnetic resonance imaging (MRI), absence of uterus with multiple uterine fibroids arising from uterine remnants bilaterally, with normal ovaries bilaterally was noted (Figure 1).

With above findings, patient was informed regarding the diagnosis of MRKH syndrome with fibroids arising from the uterine remnants. After taking informed consent, patient was taken for laparoscopic myomectomy with Davydov's vaginoplasty.

On laparoscopy, complete absence of uterus with fibroids arising from the Mullerian remnants bilaterally was noted. Bilateral ovaries were present and appeared normal (Figure 2a and b). The fibroids arising from both the remnants were excised at the base with bipolar and laparoscopic scissors and specimen retrieved by contained morcellation. For vaginoplasty, firstly, space between the fused ends of Mullerian remnant bands and rectum was dissected with harmonic. Cervical colpotomy CCL (CCL) retractor introduced vaginally to delineate the short vagina and an incision was made over the apex of short vagina laparoscopically with harmonic (Figure 3a). Pelvic peritoneum over the pararectal space mobilized with

harmonic to be fashioned as walls for neovagina. (Figure 3b).

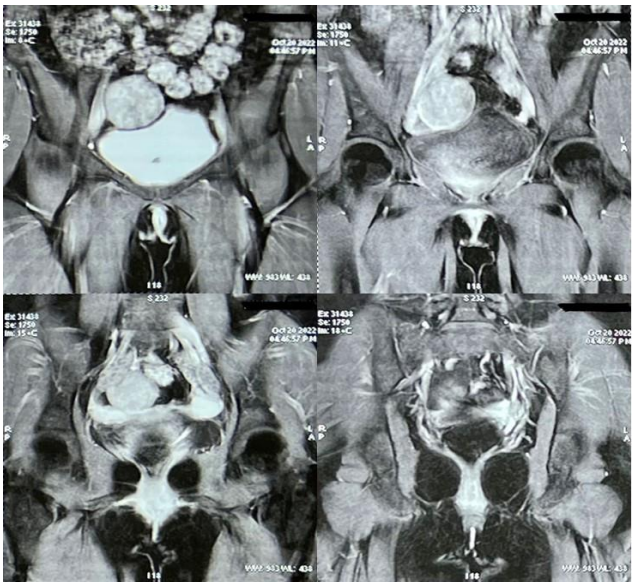

Figure 1: MRI showing uterine remnant bands with fibroids arising from them bilaterally. Ovaries present bilaterally.

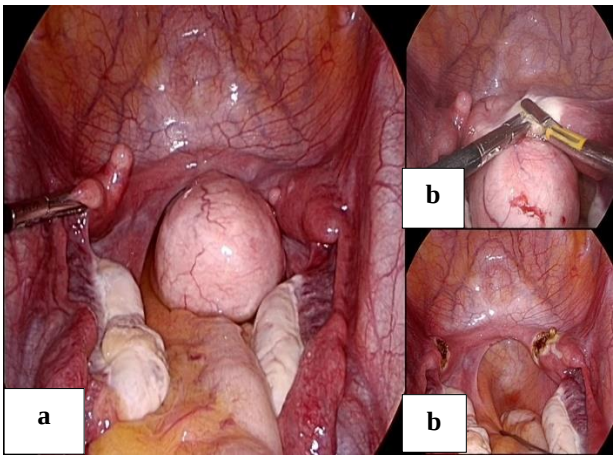

Figure 2: Laparoscopic intra operative findings (a) uterine remnants with fibroids bilaterally, tubes and ovaries appear normal; (b) laparoscopic myomectomy with bilpolar; and (c) post myomectomy picture of pelvis.

The free ends of mobilized peritoneum fixed to vulva with polyglactin sutures (Figure 3c). Neovagina was created by closing the mobilized peritoneum over a mould placed vaginally with polyglactin sutures (Figure 3d). Postoperative recovery was uneventful.

Patient was explained regarding mould care and advised to use it regularly for next 6 months, to maintain patency and adequate length of neovagina. She was counselled regarding surrogacy for rearing biological off springs.

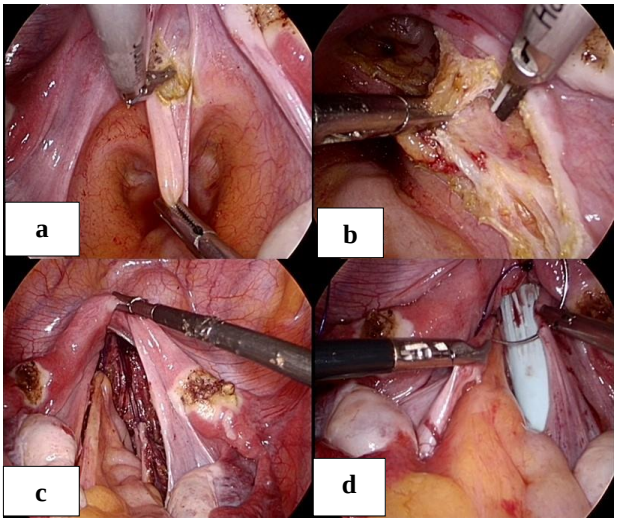

Figure 3: Steps of Davydov's vaginoplasty.

DISCUSSION

MRKH syndrome refers to congenital developmental defect of female genital tract which is characterized by absence of uterus with blind vagina (absence of upper 2/3rd of vagina). Secondary sexual characters are normal with a normal female karyotype. When seen in isolation it is MRKH syndrome type I. It is often associated with other malformations in the body like, renal (unilateral agenesis, ectopia of kidneys or horseshoe kidney), skeletal- in particular, vertebral (Klippel-Feil anomaly; fused vertebrae, mainly cervical; scoliosis), hearing defects, more rarely, cardiac and digital anomalies (syndactyly, polydactyly), which is termed MRKH syndrome type II. It is diagnosed by clinical features and radiological absence of uterus and cervix.[1] Although ovaries are present, pregnancy is a challenge for such cases, they may be offered surrogacy or uterine transplant.[3,4]

Fibroids are the most common gynaecological tumours, they commonly arise from smooth muscle cells in the uterus but also known to arise from any smooth muscle cell in the body.[2,5] Various medical managements have been proposed for their treatment, but surgical excision remains the most effective mode of treatment for fibroids.[2]

Fibroids arising in MRKH syndrome is a rare association. Presence of fibroids in the absence of uterus may be explained by the presence of smooth muscle fibres in proximal ends of Mullerian ducts. These smooth muscle cells under the influence of hormones from normally functioning ovaries may grow into fibroids as seen in women with uterus. Additionally, there may be somatic genetic mutations or clonal chromosomal abnormalities in patients with MRKH syndrome contributing to different levels of regulators of estrogen signaling.[6] These fibroids may present with features of mass abdomen, pain in lower abdomen, sometimes pressure effect over the ureter/bladder/rectum may cause bowel and bladder disturbances.[7] Any mass in pelvis requires adequate

evaluation. Association of other systems as seen in MRKH syndrome type II must be ruled out.[1] Surgical management often involves myomectomy and offering vaginoplasty as per the sexual needs of the patient.

CONCLUSION

Fibroids can arise from any smooth muscle tissue in the body. Fibroids arising from Mullerian remnants in the absence of uterus is rare. During evaluation of pelvic mass in MRKH syndrome, a differential of fibroids should not be ruled out due to the absence of uterus. Complete excision of fibroids and performing the best reconstructive procedure according to the needs of the patient constitutes proper management. Surrogacy is an option for such cases desiring fertility.

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