Case Report

Leiomyoma Arising From The Rudimentary Uterus

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ABSTRACT

In this article, leiomyoma arising from the uterus remnant in a patient with Mayer–Rokitansky–Kuster–Hauser (MRKH) syndrome is reported.

The patient was a 41-year–old woman with pelvic mass and a history of Mullerian agenesis and serum level of CA125 = 45 U/ml who had exploratory laparotomy. A 6×8×4 cm³ solid and firm mass with smooth surface arising from the uterine remnant as myoma was confirmed.

Uterine myoma can even occur in rudimentary uterus and it is one of the differential diagnosis for pelvic mass in cases with Mayer-Rokitansky-Kuster-Hauser syndrome.

Key words: Leiomyoma, Mullerian Ducts, Uterine, Development

Introduction

Congenital anomalies of the Müllerian system are common defects, reported in up to 3.2% of all women (1,2). Many of cases are asymptomatic and therefore unrecognized. Diagnosis is usually made with clinical presentations of infertility, miscarriage, premature birth, abnormal fetal presentation, cyclic pain, or as incidental findings during pelvic or monographic examinations. They occur due to partial development of the Müllerian ducts. Any portion or segment of a duct may experience agenesis. Various classifications have been proposed for congenital uterine anomalies based on the understood embryology of the Müllerian system (2-4). Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is the lack of Mullerian development. Leiomyomas are a rather common occurrence in the normal uterus that can arise from remnant uterus (1-7). However, our review of the literature has failed to found a case report of leiomyoma arising from a Müllerian remnant, as is the case reported here.

Case Report

A 41-year-old nulligravida was presented with pelvic mass for 1 year. According to her previous gynecologist’s history, she had MRKH syndrome for 20 years. The patient got married at 20 and had vaginal reconstruction at the age 21. She divorced at 30. She had a height of 158 cm with normal breast development, axillary and pubic hair. Her karyotype was XX. Vaginal examination showed a firm and mobile mass in pelvis. MRI was performed. A bulky mass lesion in pelvic cavity at anterolateral was shown which caused deformation and distention of the bladder to left side. Urinary tract was normal. There was no free fluid and pelvic adenopathy (Fig 1&2). Meanwhile, CA125 was 45 U/ml. Because of the progressive and persistent pelvic mass, exploratory laparotomy was conducted. It revealed a firm mass 6×8×4 cm³ in size in remnant uterus (Fig 3&4). Pathologic result showed a fresh, red and elastic to firm lobulated and encapsulated mass. The section showed a benign...
neoplasm composed of spindle shaped smooth muscle cells. There was no evidence of high mitotic or atypical activity and necrosis. Subserosal leiomyoma was finally confirmed. The patient was discharged 3 days after the operation with a stable condition.

**Discussion**

A thorough review of the literature shows that this is a rare case report (leiomyoma in remnant uterus). Uterine remnants in patients with MRKH syndrome consist of fibromuscular tissue. The tumors resembling leiomyomas can follow the same pathogenic mechanisms as normal uteri. These tumors can be asymptomatic (can be found during regular follow-up) or can cause acute or chronic pelvic pain.

When a pelvic mass is found in MRKH cases, a laparotomy is indicated. Appropriate management depends on underlying pathology. Extirpation of tumor with adjacent uterine remnant has been described (5-7), which can usually be performed using laparoscope (8). Surgical considerations should be done by identification of the blood supply, separation of the mass from the broad ligament and care in identifying and keeping the entire ureter. Removal of the opposite side of uterine remnant can be done at the same time with patient's preoperative consent (8-10).

Although the development of leiomyomas on uterine remnants is rare, but it is still possible to happen in patients with MRKH syndrome. Due to the extreme rarity of leiomyoma in Müllerian remnants, it should be considered low on the list of differential diagnoses of pelvic masses in women with Müllerian dysgenesis.

**Figure 1.** MRI shows a bulky mass lesion within pelvic cavity at anterolateral and left side of the bladder

**Figure 2.** Leiomyoma arising from the uterine remnant in a patient with MRKH syndrome.

**Figure 3.** Normal ovaries were anterior to external iliac vessels

**Figure 4.** The tumor is comprised of bundles of spindle-shaped smooth muscle cells with normal nuclei and mitotic activity
References


