CASE REPORT

Large Fibroid arising from Mullerian Remnant Mimicking as Ovarian Neoplasm in a Woman with MRKH Syndrome

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ABSTRACT

In this article, a large leiomyoma with degenerative changes arising from the rudimentary uterine knob in a patient with Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome and mimicking an ovarian neoplasm is reported.

The patient was a 39-year-old woman, known case of MRKH with vaginal aplasia who presented with pain abdomen, loss of appetite and weight. On physical examination, a large pelvic mass was detected. A provisional diagnosis of left ovarian neoplasm was made on USG and CT scan for which an exploratory laparotomy was performed. Finally, it was diagnosed as a case of multiple leiomyomas with hyaline degeneration on histopathological examination.

Myoma arising from a rudimentary uterine knob/anlage is a rare finding but should be considered in the differential diagnosis of pelvic mass in patients with MRKH syndrome.

Keywords: Leiomyoma, Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome, Mullerian remnant.

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INTRODUCTION

Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is a congenital abnormality of the genital tract and affects one in 4000 to 5000 liveborn females. The etiology of the syndrome remains unclear, and a multifactorial mode of inheritance has been proposed, including genetic and environmental factors. It usually presents with primary amenorrhea during adolescence. Concurrent association of pelvic mass with mullerian agenesis can be a diagnostic dilemma. Occurrence of myoma arising from a mullerian remnant is an extremely rare finding and only few cases have been reported so far in the literature. To the best of our knowledge, only one such case of mullerian duct remnant leiomyoma with hyaline degeneration mimicking ovarian neoplasm by USG scan has been reported in the past.7

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whole mullerian structure, left ovary could be seen which appeared normal. On gross examination, tumor was approximately \(12 \times 9 \times 8\) cm. Cut section showed whorled-appearance, suggestive of leiomyoma. Histopathology confirmed the diagnosis of fibroid with hyaline degeneration (Fig. 3). Her postoperative period was uneventful. She was well at 3 months follow-up.

**DISCUSSION**

The MRKH syndrome is characterized by failure of fusion of the two mullerian ducts in the seventh week of embryological development which results in congenital absence of vagina and either the absence of uterine tissue or presence of two laterally situated, solid, muscular rudimentary uterine anlage/knob connected by midline, fibrous bands. Occasionally, a small palpable cord (3rd knob) may lie in the midline, but it is again nonfunctional.\(^{12,14}\) Rarely, endometrium can exist which becomes active under the influence of estrogen. Reports have described patients with functioning endometrial tissue or even a hematometra in one or both of the rudimentary uterine anlage.\(^{17}\) As ovarian function is normal, estrogen dependent pathological conditions can develop, including myomas, neoplasms and endometriosis. Theoretically fibroids can undergo degenerative changes as seen in normal uteri but only one such case has been reported in literature (fibroid with degenerative changes in a MRKH case).\(^{7}\)

When a pelvic mass is found in MRKH cases, a laparotomy/laparoscopy is indicated and appropriate management depends on underlying pathology. The removal of the symptomatic tumor with the adjacent uterine remnant is indicated,\(^{5}\) which can be done laparoscopically.\(^{10-12}\) 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. The removal of opposite uterine remnant can be performed at the same time, with the patient’s preoperative consent. In our case, removal of opposite side of uterine remnant along with leiomyomas was done to prevent the risk of recurrence.

Although the development of leiomyomas from uterine remnant is a rare finding but it is still possible in patients with MRKH and it should be considered in the list of differential diagnosis of pelvic masses with mullerian dysgenesis.

**REFERENCES**


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