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CASE REPORT

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

Soma Singh, Baidyanath Chakravarty, Manju Chakravarty, Astha Chakravarty

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.[1] 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.[2-16] 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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CASE REPORT

A 39-year-old unmarried, nulliparous female who was a known case of MRKH syndrome with vaginal agenesis with recent diagnoses of type II DM, presented with complaints of loss of weight and appetite since 3 months and pain in lower abdomen of 5 days duration. There were no associated urinary and bowel complaints. At 19 years of age, she was diagnosed with MRKH syndrome, and diagnostic laparoscopy had been performed. At laparoscopy, two mullerian knobs on either side of pelvis, two rudimentary fallopian tubes and normal ovaries were seen. Her karyotype was normal (XX). She had been advised for vaginoplasty which she refused and was lost to follow-up.

On physical examination, her BMI was 21.08, mild pallor was present and secondary sexual characters were adult female type. Thyroid was not enlarged. On abdominal examination, a lump was visible in the hypogastrium which on palpation, was nontender, firm to hard in consistency with slightly irregular surface and lower border of the lump was not reachable. It measured approximately 10 × 8 cm, with restricted sideways mobility. There was no free fluid in abdomen. Her external genitalia were normal female type. Vagina was absent and, on rectal examination, lower border of the pelvic mass could be felt. Laboratory investigations showed mild anemia (hemoglobin—10 gm%), CA-125 was 14.5 U/ml (normal). USG abdomen revealed a huge solid hypoechoic, heterogeneous SOL (11.31 × 9.97 cm) with irregular margin in pelvis, suspicious of ovarian neoplastic mass and mild hepatomegaly. CT scan of abdomen showed a multilobulated heterogeneous enhancing mass (11.7 × 10.6 cm) in pelvis (left ovarian tumor). Uterus could not been seen and right ovary was normal (Fig. 1). It also showed mild hydroureter, hydronephrosis on left side and mild hepatomegaly. Our provisional diagnosis was of left-sided ovarian tumor and decision was taken for exploratory laparotomy. Abdomen was opened by low transverse incision and a huge tumor was seen occupying the entire pelvis, encroaching into left broad ligament and sacral hollow. Right-sided mullerian knob and ovary appeared normal but left-sided ovary could not be visualized. The tumor was solid in consistency, free of adhesions to neighboring structures and seemed to arise from left mullerian knob. There was no free fluid in the abdomen and no metastatic lesions seen. Both uterine knobs and tumor were removed uneventfully (Fig. 2). After removal of the

whole mullerian structure, left ovary could be seen which appeared normal. On gross examination, tumor was approximately 12 × 9 × 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.

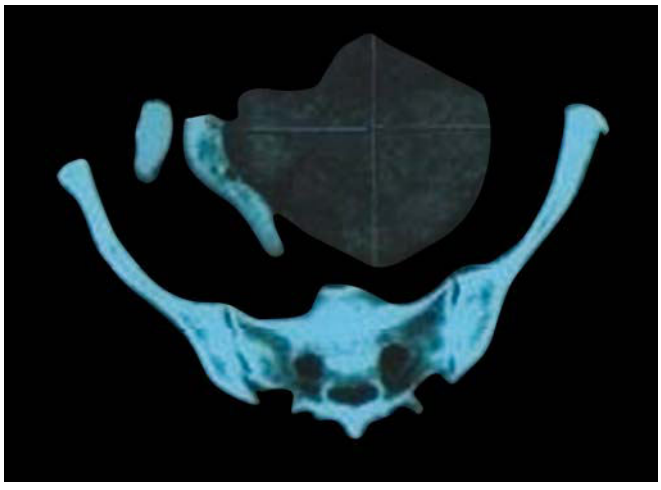

Fig. 1: Contrast enhanced CT scan picture of the pelvic tumor

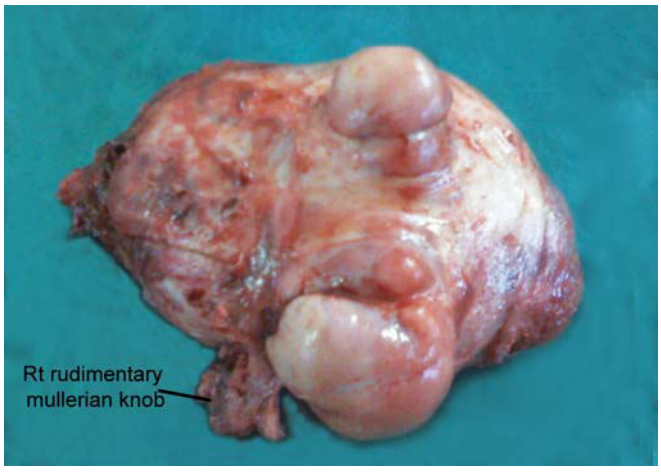

Fig. 2: Multiple leiomyomas arising from left mullerian knob alongwith the rudimentary right knob

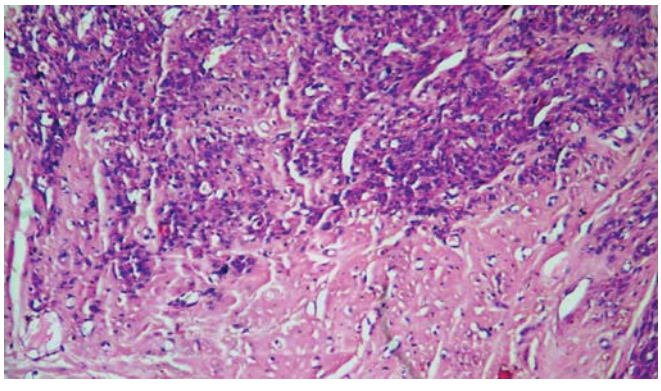

Fig. 3: Histophotomicrograph showing leiomyoma with hyaline degeneration

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.

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