Case Report

A GIANT LEIOMYOMA ORIGINATING FROM THE RUDIMENTARY UTERINE BUDS IN ROKITANSKY-KUSTER-HAUSER SYNDROME

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ABSTRACT

The development of leiomyoma from the rudimentary buds in Rokitansky-Kuster-Hauser syndrome is extremely rare. Reviewing the literature, we detected only five cases that had been previously reported. We present a case of Rokitansky-Kuster-Hauser syndrome whom we diagnosed and treated a giant leiomyoma with a diameter of 25 cm.

Key Words: Rokitansky-Kuster-Hauser syndrome, Leiomyoma

INTRODUCTION

The defective fusion of the Müllerian system is a commonly encountered congenital gynecological abnormality. Its prevalence is 2-3% in the entire population (1). The fusion abnormalities of the uterus are fundamentally divided into three groups: agenesis of the uterus, vertical fusion defects and lateral fusion defects. Rokitansky-Kuster-Hauser syndrome is also classified in the fusion abnormalities of the uterus and features vaginal agenesis with a set of normal fallopian tubes and ovaries. However, the uterus is generally absent or replaced with a centrally located Y-shaped rudimentary fibrotic buds or a streak (1,2).

CASE REPORT

A 42 year old woman applied to our clinic with a complaint of abdominal enlargement and tenderness. These symptoms had started six months before.

In retrospective evaluation of her medical history, it was found that the patient had applied to our clinic in 1981 with primary amenorrhea. Her pelvic and ultrasonographic examinations had revealed vaginal agenesis and the uterus had not been detected. Her secondary sex characteristics and hormonal profile had been normal. A definitive genetic counselling had not been performed but Barr Body had been detected in her buccal smear. She had been diagnosed as suffering from the Rokitansky-Kuster-Hauser syndrome by one of the authors (MFA) and artificial vaginal reconstruction had been performed with McIndoe procedure. A year after the operation the patient had got married.
and since then a follow-up had not been available.

The present genital examination revealed that the artificially reconstructed vagina was narrowed and ended as a blind pouch with a length of 6 cm. The pelvis was filled with a mass with an upper border reaching the level of the umbilicus. On ultrasonographic examination, the diagnosis of a solid pelvic mass with a long dimension of 25 cm was confirmed. Other physical examinations and laboratory evaluation including whole blood count, blood chemistry, hormonal profile and tumour markes were within normal range. On magnetic resonance imaging, the dimensions of the mass were measured to be 25x17x15 cm and it was centrally located in the pelvis. In addition, another mass lying posterior to the first one was detected with the dimensions of 5x4x4 cm. Both masses compressed the adjacent structures. The masses showed heterogeneous signal intensity in both T1W and T2W, which resembled uterine leiomyomas. On IVP, the anatomy of the urinary tract was normal but pelvi-caliceal dilatation was observed.

An explorative laparatomy was performed. The abdomen was entered with a midline incision. On exploration, two pelvic masses with approximate diameters of 25 and 5 cm respectively were detected. The masses resembled leiomyomas on gross examination. The masses were attached to one another with a fibrotic stalk at their lower poles. The fallopian tubes originated from the upper lateral aspects of each mass. (Fig 1). The gross appearance of the fallopian tubes and the ovaries was normal. Total fibroidectomy (the masses and the fibratic stalk were excised) and bilateral salphingo oferectomy were performed. The preservation of the ovaries was technically impossible since there were firm adhesions between the masses and the ovaries.

In the operating room, an immediate section of the excised material was performed and neither endometrial cavity, nor any structure resembling uterine cervix was observed. The frozen section of the material identified benign leiomyoma.

The postoperative course was normal and the patient discharged on the postoperative 8th day. The definitive histopathological evaluation of the excised material confirmed the initial diagnosis of leiomyoma. No endometrial and cervical tissues were detected on thorough examination of the material. The fallopian tubes and the ovaries were reported to be normal.

**DISCUSSION**

Abnormalities in the formation or fusion of the Müllerian ducts can result in a number of

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**Fig. 1.**
The appearance of the giant leiomyoma rising from rudimentary uterine buds during the operation.
1. The right ovary and the fallopian tube
2. The left ovary and the fallopian tube
3. The mass with 25 cm diameter
4. The mass with 5 cm diameter
anomalies of the uterus and vagina. In 1988, the American Fertility Society classification of Müllerian anomalies was introduced. According to this classification, Rokitansky-Kuster-Hauser syndrome is classified as Class I "Dysgenesis of the Müllerian ducts" with no reproductive potential. The syndrome includes congenital absence of the vagina and uterus. Despite the absence of the uterus, rudimentary uterine buds are found that are comparable in size (2).

Rare as it may be, the development of leiomyoma from the rudimentary buds in Rokitansky-Kuster-Hauser syndrome is a possibility since the rudimentary uterine buds are the embryologic remnants of the Müllerian duct which are formed as a result of insufficient mesenchymal differentiation. The etiology of the myomatous differentiation and the growth of the leiomyomas arising from the rudimentary buds is not clear but rarely reported cases indicate that these rudimentary buds have a potential of neoplastic growth. The neoplastic growth can be exaggerated, causing confusion in the differential diagnosis.

In 1962, Crosby and Hill collected 3 cases of leiomyoma from the rudimentary buds in Rokitansky-Kuster-Hauser syndrome (3). Farber et al reported a leiomyoma with a 15 cm diameter arising from the rudimentary uterine buds in 1978 (4). In 1988, Powell et al reported another case leiomyoma of 4 cm diameter associated with Rokitansky-Kuster-Hauser syndrome (5).

Our patient is the 6th reported case of leiomyoma in Rokitansky-Kuster-Hauser syndrome. However, a leiomyoma of this dimensions (25 cm in diameter) has never been reported in association with the syndrome. It is quite intriguing that Müllerian fusion deficiency can potentialize the growth of a leiomyoma in the absence of a uterus. Therefore, a careful and long term follow-up of these patients is essential, not only concerning the ovaries for possible neoplastic transformation but also the entire genital tract.

REFERENCES