

Adenomyosis in a Rudimentary Horn of a Mayer-Rokitansky-Küster-Hauser Syndrome

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ABSTRACT

The prevalence of congenital uterine malformations is about 0.5%. In Mayer-Rokitansky-Küster-Hauser syndrome (MRKH), patients usually present with primary amenorrhea. Here, we report a case of adenomyosis in a rudimentary horn in a 37-year-old woman with MRKH syndrome who had cyclical abdominal pain with increasing frequencies since her 24th year. This case is an evidence for adenomyotic lesions in the myometrium of Müllerian remnant with no functioning endometrium.

Keywords: Mayer-Rokitansky-Küster-Hauser syndrome, Adenomyosis, Endometrium.

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INTRODUCTION

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome causes primary amenorrhea in one in 4,500 to 10,000 women. They exhibit normal secondary sexual characters and uterovaginal aplasia/hypoplasia. In approximately 10% of MRKH, functional islands of endometrium may result in a hematometra and symptoms of cyclic pain.¹ Uterine adenomyosis is a benign disorder characterized by the extension of endometrial glands and stroma into the myometrium. Here, we report a case of adenomyosis in a rudimentary horn in a woman with MRKH syndrome.

CASE REPORT

A 37-year-old woman with primary amenorrhea diagnosed as a case of Müllerian agenesis, presented with severe lower abdominal pain for 15 days. The lady had cyclical abdominal pain with increasing frequencies

since her 24th year. She was symptomatically treated with analgesics. Laparoscopy was done as a part of her evaluation of primary amenorrhea and was diagnosed to have MRKH syndrome with two unfused rudimentary horns with normal bilateral ovaries and a blind vagina. She was phenotypically female and her karyotyping was 46 XX. She was advised vaginal dilators. She is married, has an active sexual life and has adopted a child.

In January 2013, she had an episode of acute abdominal pain. Ultrasonogram and computed tomography abdomen showed bulky uterus 9.0 × 6.8 × 5.3 cm with features suggestive of multiple seedling fibroids (Fig. 1). She was symptomatically treated for her cyclic abdominal pain.

In the month of April 2013, she presented to our hospital with continuous pain in the lower abdomen for 15 days. Ultrasound showed an enlarged uterus with features suggestive of fibroids. Clinically, a differential diagnosis of adenomyosis was made, and GnRH analogues were administered after counseling. She became symptom free when GnRH analogues were given at an interval of 4 weeks for 2 months.

Two months later, she presented with similar complaints. The possibility of adenomyosis was discussed with the patient. She opted for a surgical removal of the uterus. Total abdominal hysterectomy was planned. Intraoperatively, there was an enlarged left horn of uterus measuring size 8 × 8 cm. A small right rudimentary horn measured 0.5 × 0.5 cm (Figs 2 and 3). Both ovaries were normal. Both horns of the uterus were surgically removed (Fig. 4). Histopathological examination was reported as adenomyosis in the enlarged uterine horn (Fig. 5). Patient came for follow-up and is asymptomatic at present.

DISCUSSION

The etiology of adenomyosis has been postulated as the infiltration of eutopic endometrium into the myometrium. Though confirmation of this hypothesis is lacking, the frequent surgical finding of adenomyomas² that are contiguous with the endometrium lend credence to the hypothesis. The absence of a functional endometrial lining in the present case does not support the above theory. Another hypothesis promotes the possibility that metaplasia of stromal cells under the influence of autocrine factors or paracrine factors which are the intermediaries of

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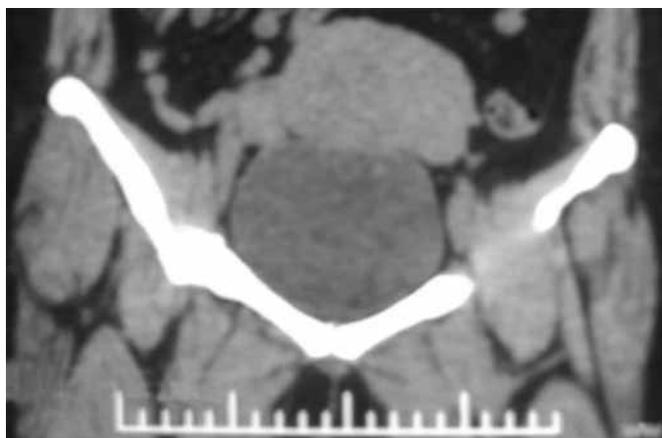


Fig 1: Computed tomography of abdomen showing enlarged left rudimentary horn



Fig. 2: (Left) small right rudimentary horn with right ovary, (right) enlarged left rudimentary horn

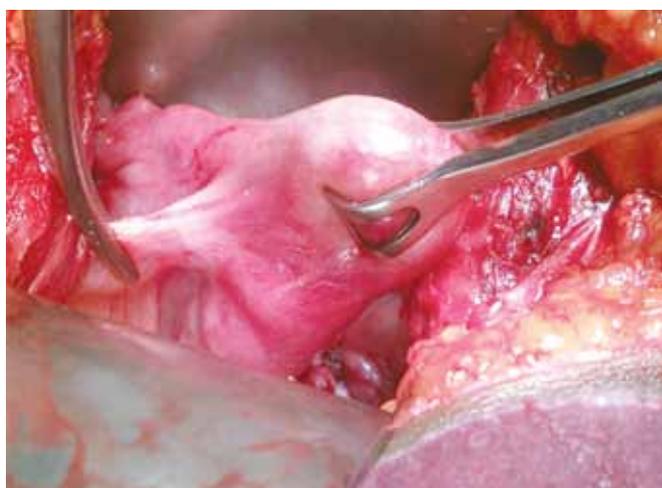


Fig. 3: Intraoperative picture showing small right rudimentary horn with right round ligament



Fig. 4: (Left) enlarged left rudimentary horn, (right) small right rudimentary horn

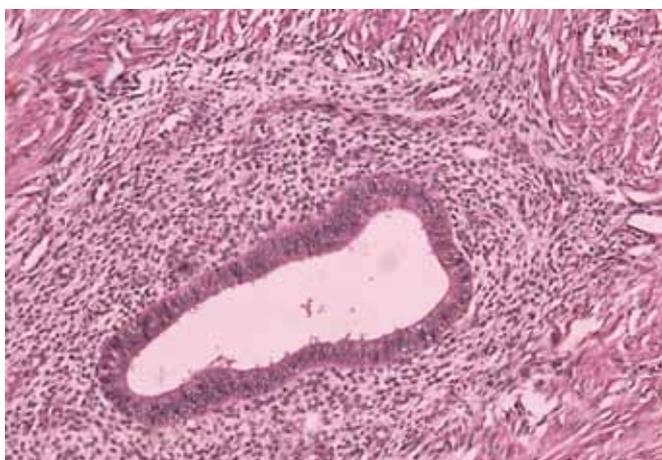


Fig. 5: High power field view of endometrial glands and stroma

genetic, immunologic, and endocrine influences can lead to the development of adenomyosis *in situ*.^{3,4} In the present case, histopathological diagnosis of adenomyosis, in the absence of an endometrial lining supports the metaplasia theory. Parikh in his review of MRKH syndrome states that fibroids and adenomyosis rarely develop in the rudimentary nonfunctioning uterus.^{5,6}

Enatsu et al⁶ reported the first case of adenomyosis in MRKH syndrome. Chun et al⁷ reported a case of simple endometrial hyperplasia of ectopic endometrial tissue in myometrium with normal endometrial cavity in patient with adenomyosis, and the authors proposed the possibility of spontaneous hyperplasia of ectopic endometrium independent of eutopic endometrium, which partially supports the hypothesis of metaplasia in the development of adenomyosis. The present case is an evidence for adenomyotic lesions in the myometrium of Müllerian remnant which had no functioning endometrium. The histogenesis of adenomyosis in this patient may be metaplasia, rather than direct invasion of eutopic endometrium.

CONCLUSION

The present case is an evidence for adenomyotic lesions in the myometrium of Müllerian remnant which had no functioning endometrium. The histogenesis of adenomyosis in this patient may be metaplasia, rather than direct invasion of eutopic endometrium.

REFERENCES

1. Fedele L, Bianchi S, Frontino G, Ciappina N, Fontana E, Borruto F. Laparoscopic findings and pelvic anatomy in Mayer-Rokitansky-Küster-Hauser syndrome. *Obstet Gynecol* 2007 May;109(5):1111-1115.
2. Yan CM, Mok KM. Uterine fibroids and adenomyosis in a woman with Rokitansky-Küster-Hauser syndrome. *J Obstet Gynaecol* 2002;22:561-562.
3. Parrot E, Butterworth M, Green A, White IN, Graves P. Adenomyosis—a result of disordered stromal differentiation. *Am J Pathol* 2001;159(2):623-630.
4. Mai KT, Yazdi HM, Perkins DG, Parks W. Pathogenetic role of the stromal cells in endometriosis and adenomyosis. *Histopathology* 1997;30(5):430-442.
5. Parikh MN. Congenital absence of vagina: MRKH syndrome. *J Obstet Gynecol* 2000;50(5):128-138.
6. Enatsu A, Hasada T, Yoshida S, et al. Adenomyosis in a patient with the Rokitansky-Küster-Hauser syndrome. *Fertil Steril* 2000;73(4):862.
7. Chun S, Jeon GH, Cho HJ, Kim YM, Ji YI. Endometrial hyperplasia in myometrium of woman with uterine adenomyosis: a case report. *J Reprod Endocrinol* 2012 Apr;4(1):56-60.