

# A giant cystic adenomyoma mimicking the degeneration of uterine myoma

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## Summary

**Purpose:** To report a rare giant isolated cystic adenomyoma mimicking the degeneration of uterine myoma. **Materials and Methods:** The authors present a 39-year-old woman referred to this hospital complaining of lower abdominal pain. Transvaginal ultrasonography (US) showed a 6.0×3.9 cm cystic region within an 8.0×6.1 cm solid mass on the posterior of the uterine wall mimicking the degeneration of uterine myoma. Laparotomy was performed and the mass was excised completely. **Results:** Pathology of the specimen demonstrated the cystic cavity was lined by endometrial glands and stroma, hemosiderin deposition was also seen, confirming the diagnosis of cystic adenomyoma. The patient made a good recovery and she was relieved from symptoms and remained asymptomatic over her follow-up interval. **Conclusion:** Clinicians need to pay attention to its differential diagnosis of uterine cystic lesions preoperatively.

**Key words:** Cystic adenomyosis; Adenomyotic cyst; Cystic adenomyoma; Degeneration of myoma; Chocolate-like liquid.

## Introduction

Adenomyosis is a gynecologic benign disease characterized by the presence of endometrial tissue within the uterine myometrium. Adenomyosis is usually observed in perimenopausal or multiparous women. Clinical symptoms are often non-specific, generally including menorrhagia, dysmenorrhoea, and somewhat pelvic pain. Diffuse adenomyosis with adjacent smooth muscle hypertrophy is the most common form, while focal or nodular adenomyosis, especially the cystic variant named cystic adenomyosis, adenomyotic cyst or cystic adenomyoma, is quite rare. The preoperative diagnosis becomes more challenging. It is often misdiagnosed as the degeneration of uterine myoma, uterine malformation, and congenital mesonephric and paramesonephric cysts. Transvaginal/transabdominal ultrasonographic tomography, CT, MRI, hysteroscopy, and hysterosalpinography (HSG) contribute to its diagnosis and differential diagnosis, ultimately histological examination is the golden standard confirming the disease. Conservative treatment including gonadotrophin releasing hormone agonists (GnRHa), oral contraceptive, non-steroid anti-inflammatory drugs or even progestogen-loaded intrauterine device may alleviate symptoms temporarily[1]. Eventually patients may require surgical treatment for excision of the lesion or hysterectomy. In this article, the authors report a giant isolated cystic adenomyoma preoperatively diagnosed as the degeneration of uterine myoma in a 39-year-old woman.

## Case Report

A 39-year old woman (gravida 5, para 1) presented with the complaint of lower abdominal pain for two weeks. The patient was clinically diagnosed by a clinician as uterine myoma for five years. In her regular follow-up, the lesion of the uterus became gradually larger. She lacked any associated symptoms including urogenital and gastrointestinal systems. Her menstrual cycle was regular (30-day cycle, normal menstrual capacity, without dysmenorrhoea). On gynecologic bimanual examination, an 8×6 cm hard mass within the posterior of the uterine wall was palpated. Transvaginal ultrasonography (US) showed an enlarged uterus, a 6.0×3.9 cm cystic region within an 8.0×6.1 cm solid mass on the posterior of the uterine wall. The patient was initially diagnosed with degeneration of uterine myoma (cystic degeneration or red degeneration). The serum concentration of tumor marker cancer antigen 125 (CA125) was elevated (85.62 U/ml, normal range within 0-35 U/ml). Under laparotomy observation, a palpable hard mass within the posterior wall of the uterine corpus was seen. Bilateral ovaries and fallopian tubes were normal in appearance, and there were no other abdominal abnormalities or ascites. Then the excision of the cyst was performed which appeared difficult since the cleavage plane is unclear unlike uterine myoma. When the incision reached the cystic cavity, chocolate-like content about 300 ml flowed from the cyst and the mass was then excised completely (Figure 1a). Histological examination showed endometrial epithelium and stroma lining the inner cyst wall, and extensive hemosiderin-laden macrophages were seen, confirming the diagnosis of cystic adenomyoma (Figure 1b). One month after the surgery, transabdominal ultrasonographic tomography of the patient was within normal limits, the serum CA125 became normal, her symptoms vanished.

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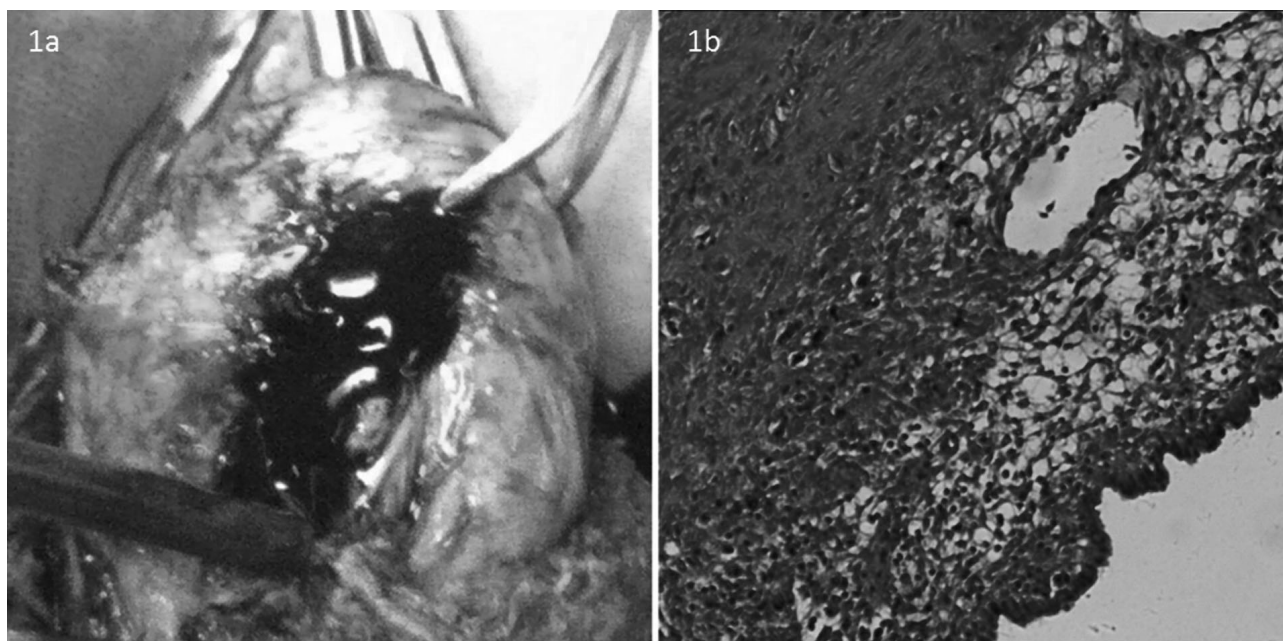


Figure 1a. — Dark-brown content flows from the cyst when the incision reaches the cystic cavity during laparotomy.

Figure 1b. — Microscopic appearance of the cystic wall, endometrial epithelium, and stromal lining of the inner cyst wall and extensive hemosiderin-laden macrophages are also seen. (Hematoxylin-Eosin stain; magnification  $\times 100$ ).

## Discussion

Adenomyosis is a benign invasion of endometrium into the uterine myometrium, which is histologically defined by the presence of endometrial glands and stroma in the myometrium. Adjacent smooth muscle hyperplasia is commonly seen in its pathology. Adenomyosis usually affects parous women, especially those who are perimenopausal. It often appears as a diffuse lesion which has no clear boundary with the myometrium. Cystic adenomyoma is an extremely rare variation of adenomyosis and in most cases, contains hemorrhagic material. Presumably repeated hemorrhage during menstruation into the ectopic endometrium surrounded by myometrium is a cause of the cyst formation [2]. Considering the common embryological origin from the Müllerian ducts of the endometrium and the subjacent myometrium, another etiology of adenomyosis may be estrogen stimulation of Müllerian rests (a direct proliferation of metaplastic myometrial cells of endometrial tissue after long periods of quiescence). Iatrogenic implantation is also a potential mechanism in women who have undergone any uterine surgical intervention. The present patient reported underwent artificial abortion four times, which added the risk of adenomyosis. In the reported cases, most were small cystic lesions, that rarely had a diameter larger than 5 mm [3]. The present case was a giant solid mass measuring  $8.0 \times 6.1$  cm with a  $6.0 \times 3.9$  cm cystic centralis. The cystic adenomyoma shows similarly clinical characteristics mainly with complains of progressively in-

creasing dysmenorrhea, menorrhagia, and chronic pelvic pain, while 35% patients remain asymptomatic. Intracystic bleeding accompanied by the menstrual cycle, then progressive increase in the size of the mass and the stretching of the endometrial cavity, may be causes of severe dysmenorrhea and pelvic pain.

In the present case, the solid mass with a cystic lesion of the uterus had been initially incorrectly diagnosed as cystic or red degeneration of myoma. Other differential diagnoses include congenital uterine cysts such as Müllerian or Wolffian duct cysts, uterine malformation such as unicornuate uterus with an obstructive horn, and ovarian tumors. Since the introduction of non-invasive imaging techniques such as transabdominal/transvaginal US/CT, and especially MRI, allow accurate evaluation of the content surrounded by myometrial tissue. In the present case, transvaginal US showed an enlarged uterus, an  $8.0 \times 6.1$  cm solid lesion on the posterior of the uterine corpus with a  $6.0 \times 3.9$  cm cystic centralis. The major role of US lies in initial screening for and exclusion of gynecologic conditions such as ovarian torsion and pregnancy [4]. It is difficult to distinguish these cystic lesions through US. The radiologic appearance of cystic adenomyosis is highly characteristic. CT demonstrates thick-walled cysts with fluid contents and contrast wall enhancement. MRI is especially valuable, due to its superior contrast resolution and utilization of multiple pulse sequences for tissue characterization. Pelvic MRI identifies complex cysts with internal fluid-fluid levels, myometrial

wall hypertrophy, and hemosiderin deposition. In the case of hemorrhagic leiomyoma, methemoglobin accumulates in obstructed veins at the periphery of the mass, producing a rim that is T1-hyperintense and T2-hypointense. This is distinct from the T1- and T2-hypointense rim of hemosiderin in cystic adenomyosis [5]. However, the present authors did not use this imaging method for their patient due to the patient's economic reasons. Despite use of MRI, more complex cases of cystic adenomyosis can be difficult to discern from malignancy.

To directly visualize the uterine cavity, a minimally invasively hysteroscopic approach offers the possibility of obtaining endometrial/myometrial biopsies under visual control and/or ultrasound guidance. Some of the lesions are directly recognizable at hysteroscopy because they can directly bulge into the endometrial cavity. Treatment by resection or ablation can be performed. HSG is very useful for the differential diagnosis from uterine anomaly. In cases with cystic adenomyosis uterine cavity and where both fallopian tubes are patent, while in cases with uterine anomaly such as a rudimentary uterine horn, the fallopian tube on the affected side is not visualized through HSG. Distinguishing congenital Müllerian from Wolffian cysts remains difficult and it depends on pathology. On pathology, cystic adenomyoma is characterized by ectopic endometrial epithelium, glands or stroma invading the myometrium. A region of myometrial hyperplasia surrounding the uterine muscle and hemorrhage and hemosiderin-laden macrophages associated with endometrial sloughing may also be seen.

Treatment of these cases pose great difficulties to clinicians that will be difficult to overcome until well-designed studies are launched to guide management. It depends on the size and location of the cyst, severity of the patients' symptoms, as well as on the age of patients. Non-steroid anti-inflammatory drugs are initially used for alleviation of painful symptoms. Hormonal suppressive therapy with oral contraceptives or GnRH-a include mostly a temporary effect. According to a recent study, the levonorgestrel-releasing intrauterine system is an effective method in alleviating dysmenorrhea in women with adenomyosis [6], but there are no reports of its use in women with myometrial cysts. Refractory and severe cases may require surgery. Surgical treatment options are either excision of the lesion or hysterectomy. Unlike diffuse adenomyosis, cystic adenomyoma can generally be excised along the tissue planes created by their thick walls. In young patients, a minimally invasive procedure, such as laparoscopic excision is considered preferable. This spares the remainder of the uterus, ultimately enabling preservation of fertility. However, many cases have been treated by laparotomy mainly due to the difficulty in determining the exact location of the mass

and the high expertise needed to safely remove the entire lesion by laparoscopy. Radical hysterectomy, although curative for diffuse adenomyosis, is not indicated for cystic adenomyosis unless the disease is multifocal, symptoms are intolerable, and the patient no longer desires fertility. Furthermore, due to rare malignant potential of the cystic adenomyosis, the present authors emphasize the importance of intraoperative frozen section work-up, which is a useful guide in the surgical and therapeutic plan. Previous studies have reported rare cases of endometrial carcinoma and six cases of clear cell carcinoma from cystic adenomyosis in the last 20 years [7]. In the present case, the authors performed laparotomy and excised the lesion without intraoperative frozen section work-up for their preoperative misdiagnosis and limited medical conditions. The patient made a good recovery and she was relieved from symptoms and remained asymptomatic over her follow-up interval.

## Conclusion

Cystic adenomyoma, a rare variant, may be easily misdiagnosed as degeneration of myoma. This present case emphasizes that in the differential diagnosis of cystic uterine mass, cystic adenomyoma must be kept in mind.

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