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Case Report

Accessory and cavitated uterine mass: a rare case

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ABSTRACT

Accessory and cavitated uterine mass (ACUM) are a rare congenital Müllerian duct anomaly characterized by a non-communicating uterine mass containing functional endometrial tissue, typically located near the insertion of the round ligament. It predominantly affects adolescents and young women presenting with severe dysmenorrhea or cyclic pelvic pain unresponsive to medical therapy. Due to its similarity in presentation to adenomyosis, fibroids, and rudimentary uterine horn, ACUM is frequently misdiagnosed. We report a case of a reproductive-age woman with intractable dysmenorrhea since adolescence that did not improve with hormonal treatment. Preoperative magnetic resonance imaging (MRI) suggested ACUM, and the patient underwent laparoscopic ACUM removal. Intraoperative findings revealed an isolated cavitated mass at the left round ligament. Histopathological examination confirmed the presence of functional endometrial lining surrounded by hypertrophic myometrium, consistent with ACUM. Postoperatively, symptoms resolved completely. Early recognition of ACUM is essential in young women with unexplained dysmenorrhea, and surgery remains the definitive treatment.

Keywords: Accessory and cavitated uterine mass, Müllerian anomaly, Severe dysmenorrhea, Non-communicating uterine mass

INTRODUCTION

Congenital Müllerian anomalies arise from abnormal development, fusion, or resorption of the Müllerian ducts and affect approximately 5-7% of women in reproductive age group, often presenting with infertility, pelvic pain, or dysmenorrhea.¹ Among these anomalies, ACUM has been recognized as a distinct entity in recent years.

ACUM is defined as a solitary, non-communicating cavitated mass lined by functional endometrium and surrounded by smooth muscle fibers in an otherwise normal uterus, fallopian tubes, and ovaries.² The lesion is most frequently located near the insertion of the round ligament, suggesting a possible developmental defect related to dysfunction of the female gubernaculum or segmental Müllerian duct abnormality.³ Unlike uterine anomalies such as bicornuate uterus or unicornuate uterus with a rudimentary horn, ACUM is not associated with duplication or distortion of the main uterine cavity.⁴

Patients commonly present during adolescence or early adulthood with severe primary dysmenorrhea, chronic pelvic pain, or cyclical pain that is resistant to analgesics and hormonal therapy.⁵ Due to its rarity and nonspecific imaging findings, ACUM is often misdiagnosed as adenomyosis, endometriosis, leiomyoma with degeneration, or obstructive Müllerian anomalies.⁶ MRI is the preferred diagnostic modality and typically shows a well-demarcated cavitary mass with hemorrhagic contents and a myometrial rim.⁷

Surgical excision of the accessory mass via laparoscopy is considered the treatment of choice, especially in women desiring fertility.⁸ However, hysterectomy may be chosen in older or multiparous patients, or in cases where conservative surgery fails to relieve symptoms.⁹ Due to the limited number of documented hysterectomy cases for ACUM, further reports are essential to improve clinical understanding and management strategies. The present study describes a rare case of ACUM treated successfully

with laparoscopic ACUM removal in a symptomatic reproductive-age woman.

CASE REPORT

Patient information

A 19-year-old unmarried female presented to the gynecology outpatient department with complaints of severe dysmenorrhea and secondary amenorrhea for the past three months. She had no history of previous surgeries or chronic medical illness.

Menstrual and clinical history

The patient attained menarche at the age of 14 years and had regular menstrual cycles thereafter. Since the onset of menstruation, she experienced progressively worsening dysmenorrhea, which was initially cyclical but later became continuous, dull aching pelvic pain occurring on and off. The pain was associated with nausea, headache and dizziness. She had received hormonal therapy and analgesics at a local clinic but did not respond to the treatment.

Physical examination and investigations

General physical examination was unremarkable. Per abdominal examination was normal. As the patient was unmarried, per vaginal examination was deferred.

Routine blood investigations, including complete blood count, liver and renal function tests were within normal limits (Figure 1).

Ultrasonography

Round cystic left uterine lesion with thin myometrial mantle, low level internal echoes and non-communicating with endometrial cavity, Possible differential diagnosis- Unicornuate uterus with rudimentary horn and accessory cavitated uterine mass.

3D ultrasonography

Well defined round to oval thin-walled lesion filled with low level internal echoes is seen arising from left antero lateral part of uterine fundus and body causing mild mass effect over adjacent uterine contour.

The lesion is at a distance of around 8-10 mm from endometrial cavity and does not communicate with it.

The layer of myometrium (2 cm) appears to surround the lesion.

MRI

A well-defined focal rounded intramural lesion of size ~2.7×3.5×5.7 cm (Antero posterior × transverse cranio caudal) is seen in left antero lateral wall. The lesion is hypointense on T2 and hyperintense on T1 with a subtle fluid fluid level and its contents are not communicating with endometrial cavity (Figure 2).

Diagnosis of an ACUM was made based on clinical presentation and imaging findings. The patient underwent laparoscopic excision of the ACUM under general anesthesia. During the procedure, a well-circumscribed mass measuring 3×3×2 cm was identified adjacent to the left round ligament, confirmed by MRI as non-communicating with the uterine cavity. Bilateral tubes and adnexa were clear, and the pouch of Douglas was free. A small incision was made on the mass to remove it along with its wall, and the myometrium was approximated with barb sutures. (Figure 3) The surgical duration was 1 hour and 30 min, with an estimated blood loss of approximately 100 ml. HPE revealed patchy areas of endometrial tissue and associated fibrocollagenous and fibromuscular tissue with hemorrhage and hemosiderin-laden macrophages, confirming ACUM with Adenomyosis. Postoperatively, the patient experienced complete relief from pelvic pain and dysmenorrhea, resumed regular menstruation during follow-up, and remains symptom-free at her follow-up appointment.

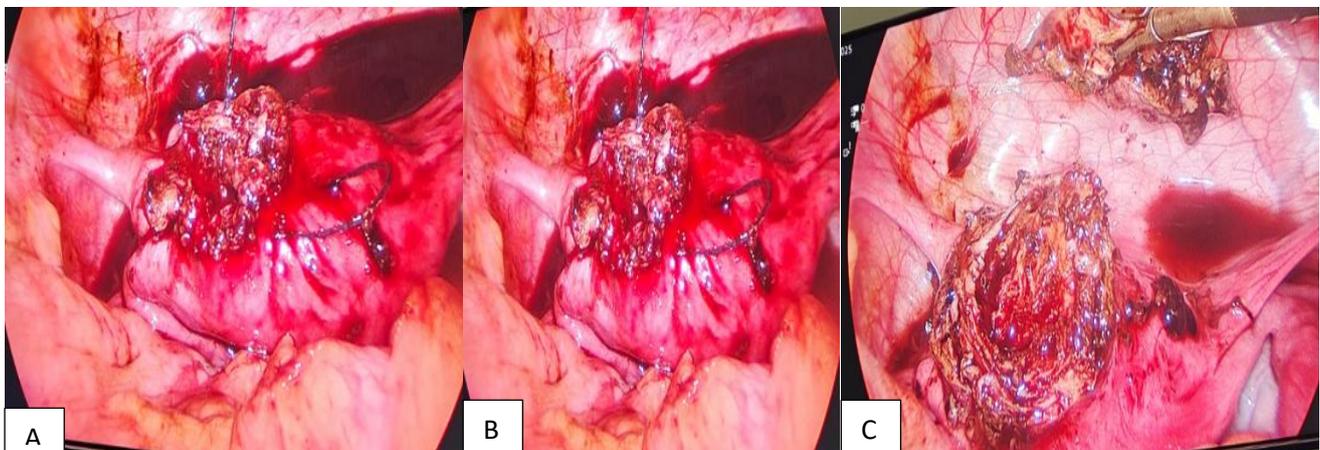


Figure 1 (A-C): ACUM visualised intraoperatively.

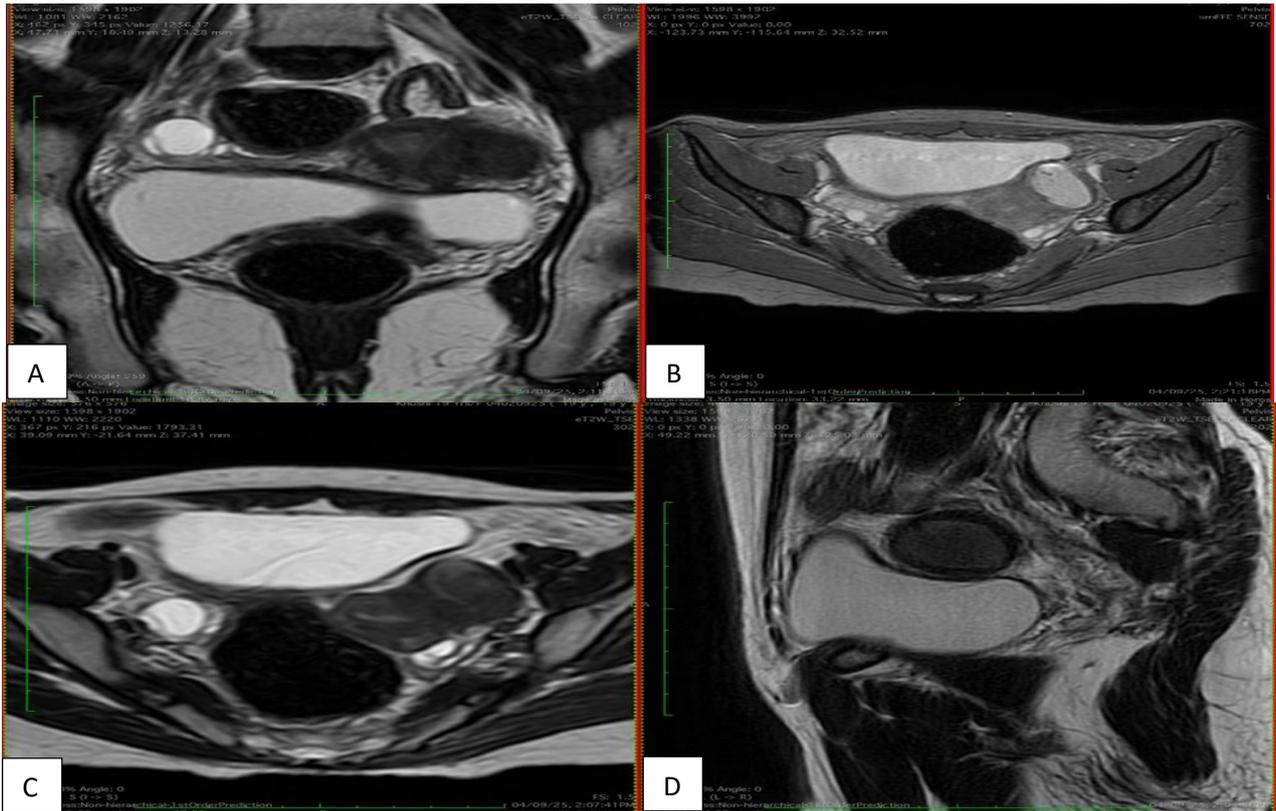


Figure 2 (A-D): MRI image showing ACUM. A separate well defined focal rounded intramural lesion in left anterolateral wall. The lesion is hypointense on T2 and hyperintense on T1 with a subtle fluid filled level and its content is not communicating with endometrial cavity. Right cornua is well visualised. However, left cornua is obscured.

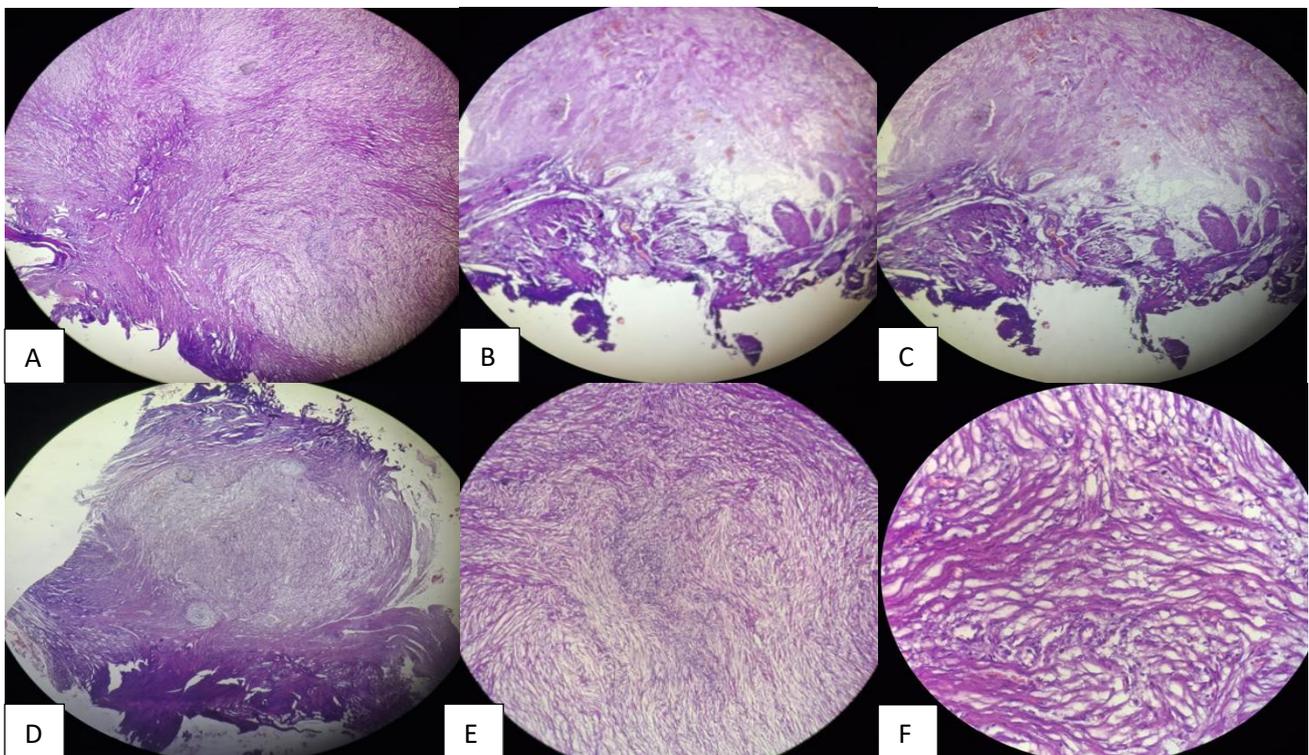


Figure 3 (A-F): Histopathological examination revealed patchy areas of endometrial tissue and associated fibrocollagenous and fibromuscular tissue.

DISCUSSION

ACUM are a rare Müllerian duct anomaly, typically found in adolescents and young women presenting with severe dysmenorrhea and pelvic pain. The anomaly is characterized by an isolated, non-communicating uterine mass containing a functional endometrial cavity, usually located near the insertion of the round ligament.

The exact pathogenesis remains unclear. Acién et al proposed that ACUM originates from dysfunction of the female gubernaculum during Müllerian duct fusion, resulting in ectopic endometrial tissue surrounded by smooth muscle.^{2,3}

Patients with Müllerian anomalies or obstructive reproductive tract disorders typically present in adolescence, often after the onset of menstruation. The primary symptom is severe dysmenorrhea, which is progressive and resistant to standard treatments like non-steroidal anti-inflammatory drugs (NSAIDs) or hormonal therapy. This pain begins soon after menarche and intensifies due to the accumulation of menstrual blood behind the obstruction, causing increased pelvic pressure and inflammation. Chronic pelvic pain is also common, stemming from ongoing distension, endometriosis, or recurrent hematometra or hematosalpinx. This pain can be cyclic or chronic and disrupts daily life, education, and quality of life. Patients may experience amenorrhea, either primary or secondary, due to obstruction hindering the normal menstrual flow. Prolonged unrecognized conditions can lead to complications such as retrograde menstruation, endometriosis, pelvic adhesions, and infertility. In the described case, the patient displayed typical signs of obstructive Müllerian conditions, including severe dysmenorrhea since menarche, indicative of an early obstructive process, and secondary amenorrhea suggestive of menstrual entrapment or hormonal alterations over time. Her symptoms were resistant to medical treatment, reinforcing the likelihood of a structural anomaly rather than a functional issue. The chronic nature and worsening of her symptoms highlighted the necessity for timely assessment and targeted treatment to avert long-term reproductive and pelvic issues.

ACUM is a rare Müllerian anomaly characterized by an ectopic, noncommunicating uterine cavity located within the myometrium, typically near the round ligament insertion. It presents most often in adolescents and young women with severe dysmenorrhea and chronic pelvic pain that is frequently unresponsive to hormonal therapy. ACUM is considered a congenital defect arising from abnormal Müllerian duct resorption or duplication, distinct from juvenile cystic adenomyoma or noncommunicating uterine horn anomalies.² Imaging-especially MRI-plays a crucial role in diagnosis by demonstrating a well-circumscribed myometrial mass containing a functional endometrial lining.¹⁰ Surgical excision is the treatment of choice and is associated with significant symptomatic relief and preservation of reproductive function.³ In this

case, the patient's longstanding dysmenorrhea and refractory symptoms were strongly indicative of ACUM, and surgical management provided definitive treatment.

Diagnostic criteria for ACUM (Acién's criteria) specify the following essentials: an isolated, accessory, cavitated uterine mass; a normal uterus, fallopian tubes, and adnexa; surgical excision paired with histopathological confirmation; the presence of a cavity lined by functional endometrium; the presence of chocolate-brown fluid inside the mass; and the absence of adenomyosis. The current case has been determined to meet all these outlined criteria. In terms of imaging, ultrasound may reveal a myometrial cystic lesion, which is commonly misdiagnosed as either a degenerating fibroid or an adenomyoma. However, MRI is recognized as the superior imaging modality for this condition, as it effectively displays a clearly defined myometrial lesion with either a hemorrhagic or fluid-filled cavity. The MRI findings pertinent to this case will be included once they are available.

The differential diagnosis of ACUM encompasses various gynecological conditions that share similar clinical and imaging characteristics. One notable condition is juvenile cystic adenomyoma, which may closely resemble ACUM but is generally accompanied by adenomyotic alterations in the adjacent myometrium. Another condition to consider is a non-communicating rudimentary uterine horn, which features a functional endometrial cavity and tends to be asymmetrically attached to the uterus, potentially maintaining vascular continuity. Degenerating fibroids can also present with cystic appearances on imaging yet are devoid of an endometrial lining, distinguishing them from ACUM. Additionally, endometriomas situated in the round ligament may imitate ACUM, though they are marked by ectopic endometrial tissue associated with the ligament rather than indicating a congenital uterine anomaly. The management of these conditions primarily hinges on the severity of symptoms and the reproductive objectives of the patient. Medical treatments, such as NSAIDs or hormonal suppression, frequently yield only temporary or insufficient relief of symptoms. As such, surgical excision is advocated as the definitive treatment modality, which serves both to confirm the diagnosis and provide a cure. For women wishing to preserve their fertility, laparoscopic excision is particularly beneficial, while hysterectomy is considered for those who have completed their childbearing. In the case described, laparoscopic removal was successfully performed with minimal blood loss and an uneventful postoperative recovery.

The prognosis for patients with ACUM is predominantly positive, mainly when correctly diagnosed and treated surgically. Postoperative outcomes are outstanding, with many patients experiencing significant relief from severe dysmenorrhea and chronic pelvic pain shortly after excision. Research indicates that laparoscopic removal of ACUM provides lasting symptom relief by addressing the

functional ectopic cavity that collects menstrual blood.^{2,3} This minimally invasive method diminishes postoperative complications, shortens hospital stays, and facilitates quicker resumption of normal activities. Fertility remains mostly unaffected since surgical excision targets the accessory cavitated mass while safeguarding uterine structure. Documented case series support successful conception and term pregnancies post-conservative management, thus confirming the reproductive safety of laparoscopic excision.² The likelihood of recurrence is extremely low, as ACUM is a congenital anomaly that does not regenerate once entirely excised. Long-term follow-up reveals continued improvement in symptoms and a minimal risk of complications when complete removal is performed.¹⁰ Consequently, with prompt diagnosis and suitable surgical treatment, patients with ACUM can anticipate remarkable symptom resolution, very low rates of recurrence, and the preservation of reproductive capabilities.

CONCLUSION

ACUM is a rare Müllerian anomaly that should be considered in young women presenting with severe dysmenorrhea and pelvic pain. Accurate radiological evaluation aids early diagnosis. Surgical excision provides definitive treatment with excellent symptomatic relief, prevents misdiagnosis, and improves quality of life.

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