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Accessory and Cavitated Uterine Mass - An Underdiagnosed Cause of Dysmenorrhea

Patricia Gomes Ferreira[1*], Sara Sereno[1], Susana Leitao[1], Horacio Scigliano[2], Soledade Ferreira[1]

[1]Obstetrics and Gynecology Department, Local Health Unit of Entre Douro e Vouga, Santa Maria da Feira, Portugal

[2]Anatomical Pathology Department, Local Health Unit of Entre Douro e Vouga, Santa Maria da Feira, Portugal

[*]Corresponding author:

Patricia Gomes Ferreira,
Obstetrics and Gynecology Department, Local Health Unit of Entre Douro e Vouga Santa Maria da Feira, Portugal

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1. Abstract

This report describes a rare case of severe dysmenorrhea caused by an accessory cavitated uterine malformation (ACUM). The diagnosis was based on the patient's symptoms and magnetic resonance imaging (MRI) findings. Laparoscopic surgery was performed successfully, with no complications during the procedure or postoperative period. ACUM is a rare but significant differential diagnosis in patients with severe dysmenorrhea unresponsive to medical management. This case highlights the importance of considering ACUM in such scenarios, with surgery remaining the most effective treatment for this condition.

2. Introduction

Accessory cavitated uterine malformation (ACUM) is an isolated, non-communicating uterine lesion with functional endometrium that lies within the normally shaped and functioning uterus and is most often located close to the uterine insertion of the round ligament [1]. The exact pathogenesis of this rare entity is still unknown, but it is believed that the origin of this uterine anomaly could be caused by a gubernaculum dysfunction during embryogenesis, expressed through a duplication and persistence of the ductal Müllerian tissue at the attachment level of the round ligament [2]. An ACUM was first described in the medical literature by J. Oliver in 1912, who wrote about a patient who presented with dysmenorrhea and had a tumor removed from the broad ligament. He described the tumor as "a globular-shaped closed sac, containing chocolate-colored fluid, thick-walled and lined by cuboidal epithelium, which resembled very closely the stroma of normal endometrium" [3]. The lesion has been described in the literature with different terminologies: juvenile cystic adenomyoma, isolated cystic adenomyoma, myometrial cyst, cavitated adenomyoma, accessory cavitated masses, and uterine-like mass, until Acién et al. described these lesions in a case series as ACUM in 2010 [4]. Currently, ACUM remains unclassified in the uterine anomaly classifications of the European Society of Human Reproduction and Embryology (ESHRE), the European Society of Gynecological Endoscopy (ESGE), and the revised American Society of Reproductive Medicine (rASRM) [5,6].ACUM is an increasingly recognized but still possibly underdiagnosed clinical entity with sporadic case reports in the literature. ACUM is usually diagnosed in menstruating girls and young women with severe and recurrent dysmenorrhea, which ranges from mild to severe. It typically starts soon after menarche and rapidly increases in severity thereafter. Chronic pelvic pain (CPP) and dysfunctional uterine bleeding are also frequent [7]. Dysmenorrhea and CPP are often primarily or secondarily resistant to common analgesics and classical hormonal treatments such as progestogen-only pills (POP), combined oral contraceptive pills (COC), or gonadotropin-releasing hormone agonists (GnRH agonists) [8].The diagnostic criteria proposed by Acién et al. [1] to diagnose ACUM are as follows: (a) an isolated, accessory, cavitated mass; (b) a normal uterus (with endometrial lumen), fallopian tubes, and ovaries; (c) surgical evidence with an excised mass and pathological findings; (d) an accessory cavity lined by endometrial epithelium, with

glands and stroma; (e) chocolate-brown-colored fluid content; and (f) no adenomyosis. Rudimentary uterine horns and adenomyotic cysts are pathological conditions to consider in the differential diagnosis [9].Non-invasive imaging modalities that help make an accurate preoperative diagnosis are ultrasound (US) and magnetic resonance imaging (MRI) [8]. According to Gupta et al. [10], the pelvic US criteria to establish the correct diagnosis of ACUM are: (a) site of the lesion along the presumed location of the round ligament, (b) characteristic appearance of the lesion: echotexture similar to uterine myometrium with central contents showing endometrioma-like ground-glass appearance and lining of the cavity similar to uterine endometrium, (c) documentation of vascularity in the lesion, (d) appearance of bilateral normal uterine cornua (on 3D US) and normal ovaries, (e) normal appearance of the rest of the uterus with no features suggestive of adenomyosis. The MRI findings in ACUM are a well-defined, cavitated uterine wall mass located immediately below the usual round ligament insertion site. Normal central T2 hyperintense endometrial lining shows hemorrhagic contents within, which appear hyperintense on T1WI with no signal suppression on fat-suppressed images and reflect hypointense shading on T2WI. The wall of the cavity is hypointense on T2WI and comprises myometrial tissue. Additionally, the uterine cavity is normal with normal bilateral cornua [10]. The normal endometrial cavity has no communication with the accessory cavity [11]. Surgical resection usually achieves optimal resolution of associated symptoms [12]. Laparoscopic minimally invasive surgery is the preferable treatment option [13]. Fertility preservation should be taken into consideration before surgical management. The lesions of ACUM are commonly located at the lateral aspect of the myometrium, beneath the insertion of the round ligament, leading to the recommendation of anterior wall uterine incision rather than posterior wall incision, thereby reducing the possibility of injury to the ascending branch of the uterine artery. As the boundary between the lesions and myometrium is usually clearly demarcated, the resection is similar to laparoscopic myomectomy. Urinary tract anomalies must be ruled out before surgery as they are associated with Müllerian anomalies, reducing the chance of urological injuries [13]. Ultrasound-guided sclerotherapy has been described in these cases. The rationale behind this treatment is the destruction of the functional endometrium, thereby preventing the monthly accumulation of menstrual blood. Even though this technique is relatively new in the treatment of ACUM, the effectiveness of alcohol instillation and its safety have been demonstrated in the management of ovarian endometriomas. In addition, the use of lauromacrogol was recently introduced for the sclerotherapy of ACUM, which is also claimed to function as a local anesthetic. Lauromacrogol has been used for the sclerotherapy of hepatic cysts, and further research is needed to draw conclusions regarding its use in the treatment of ACUM. Ultrasound drainage followed by cycle suppression to prevent further bleeding can also provide symptomatic relief, although this seems to be a temporary solution since the ectopic endometrium remains functional [9]. The aim of this report is to describe a case of dysmenorrhea with ACUM and ovarian endometrioma.

3. Case Report

A 43-year-old woman presented to the gynecology appointment with irregular vaginal bleeding and increasing dysmenorrhea, significantly impacting her daily life for the past four years. She had used an intrauterine device followed by combined contraception, but without significant improvement.Her menarche occurred at the age of 11, and her menstrual cycles were normal. She had two pregnancies, both resulting in cesarean births. She had a history of kidney lithiasis and had undergone bilateral tubal ligation.A gynecological examination revealed normal development of the vulva and vagina. Abdominal palpation revealed pelvic pain primarily located in the left iliac fossa, but no tenderness or rebound tenderness was noted. The uterus was anteverted and of a normal size. A well-defined, movable, painless mass was palpated in the left uterine adnexa. There were no obvious abnormalities in the right uterine appendages.An ultrasound examination revealed a solid mass with a regular shape on the left side of the uterus measuring 50 × 30 mm and an avascular unilocular cyst containing low-level, homogeneous "ground-glass"-like internal echoes measuring 70 × 40 mm in the left ovary.MRI showed an accessory cavitated mass along the uterine wall under the left round ligament with a T2 hyperintense endometrial lining measuring 31 × 28 × 25 mm (Figure 1). There was an expansive, round lesion measuring 89 × 56 mm on the left lateral aspect of the pelvic excavation, with hyperintense content on T1 and hypointense on T2, suggesting an endometriotic cyst (Figure 2). MRI further demonstrated a normal uterus, cornua, and right ovary (Figure 3).

The patient underwent laparoscopic surgery under general anesthesia. During the surgery, the uterus and right ovary appeared normal (Figure 4). The left ovary was enlarged, with a purplish color (Figure 5), and an oval mass was observed in the left broad ligament (Figure 6). The mass was not connected to the uterus.The left adnexa and the mass were removed and sent for histological examination. The left ovary, the largest measuring 70 × 40 mm, appeared to have a solid-cystic structure with brownish liquid content. Histological examination identified several luteinized follicular cysts and typical images of cystic endometriosis.The resected mass measured 50 × 30 mm in total. Gross examination revealed that the cystic part of the mass was filled with a blackish-brown fluid and had thick walls resembling the uterine myometrium (Figure 7). No obvious signs of malignancy were found on gross examination. Light microscopic examination of the tissue specimen, after staining with hematoxylin and eosin, showed that the cyst center was lined by endometrial glands and

stromal cells, and the cyst walls were composed of smooth muscle cells. The findings were quite similar to those of a miniature uterus (Figure 8). The final histopathological diagnosis of the mass was consistent with an accessory and cavitated uterine mass.Both uterine horns, fallopian tubes, and ovaries appeared normal on visual inspection.The patient showed no recurrence of pelvic pain or dysmenorrhea during a two-year follow-up period.

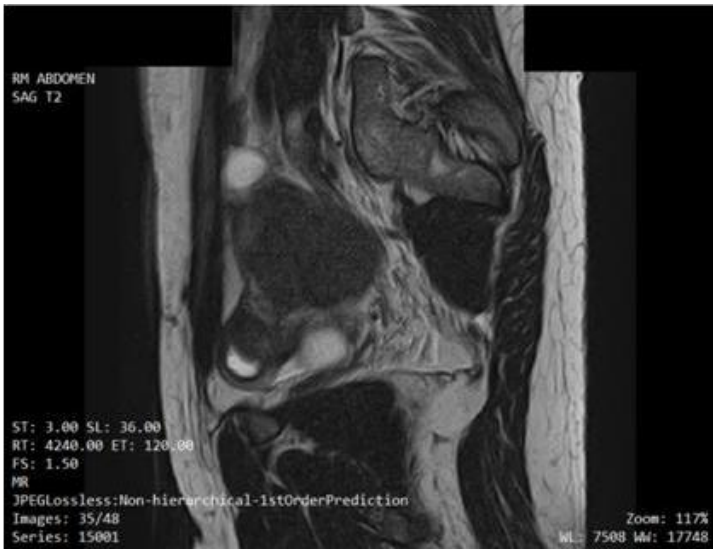

Figure 1: Accessory cavitated mass with T2 hyperintense endometrial lining.

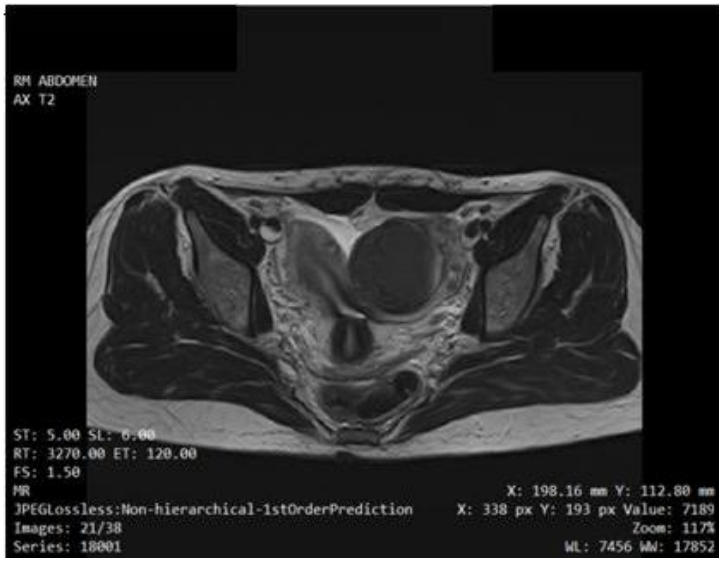

Figure 2: Left endometriod cyst.

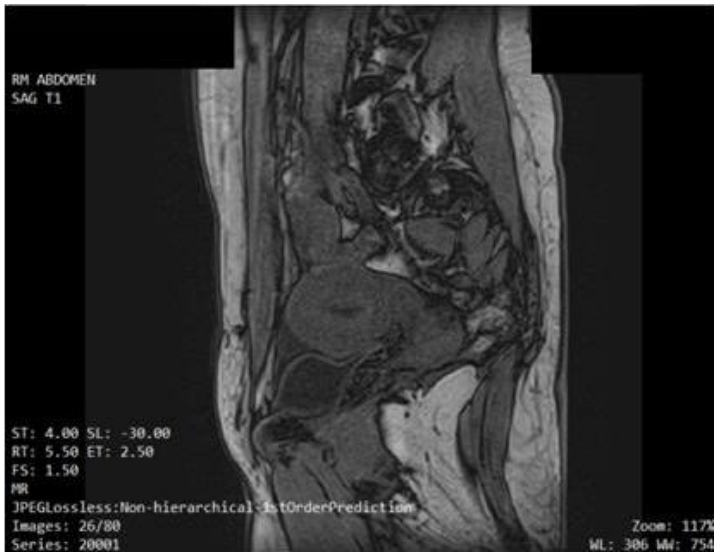

Figure 3: Uterus.

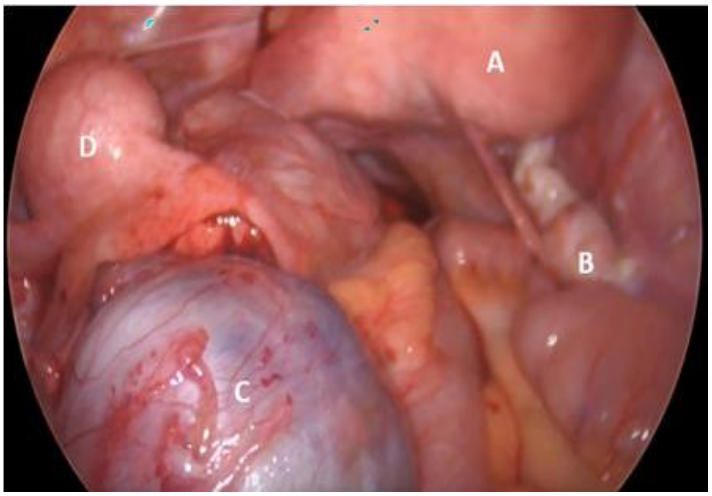

Figure 4: Surgical findings. A – Uterus; B – Right ovary; C – Cyst in the left ovary (ovarian endometrioma); D – Mass in the left broad ligament (ACUM).

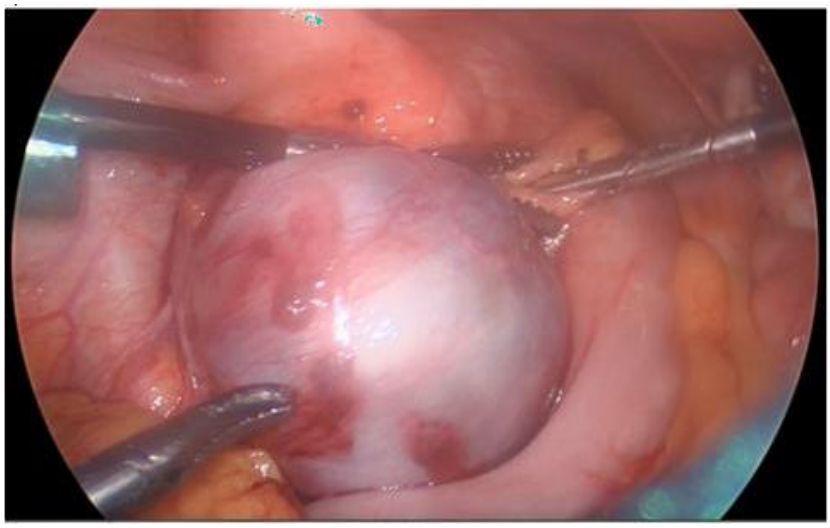

Figure 5 "Cyst in the left ovary (ovarian endometrioma).

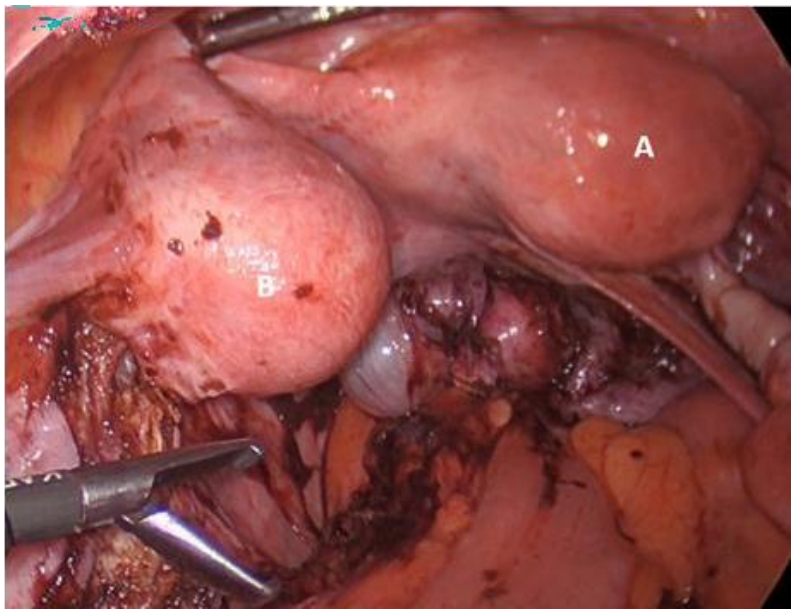

Figure 6: After left adnexectomy. A – Uterus; B - Mass in the left broad ligament (ACUM).

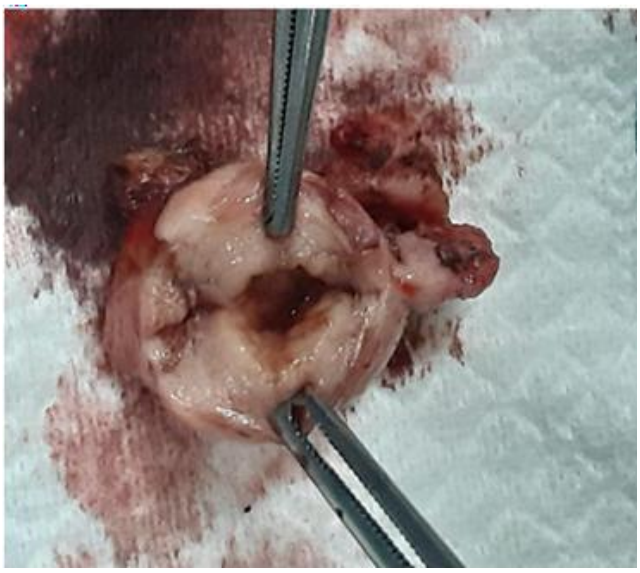

Figure 7: Resected mass from the left broad ligament. It was cut lengthwise, exposing a cavity inside.

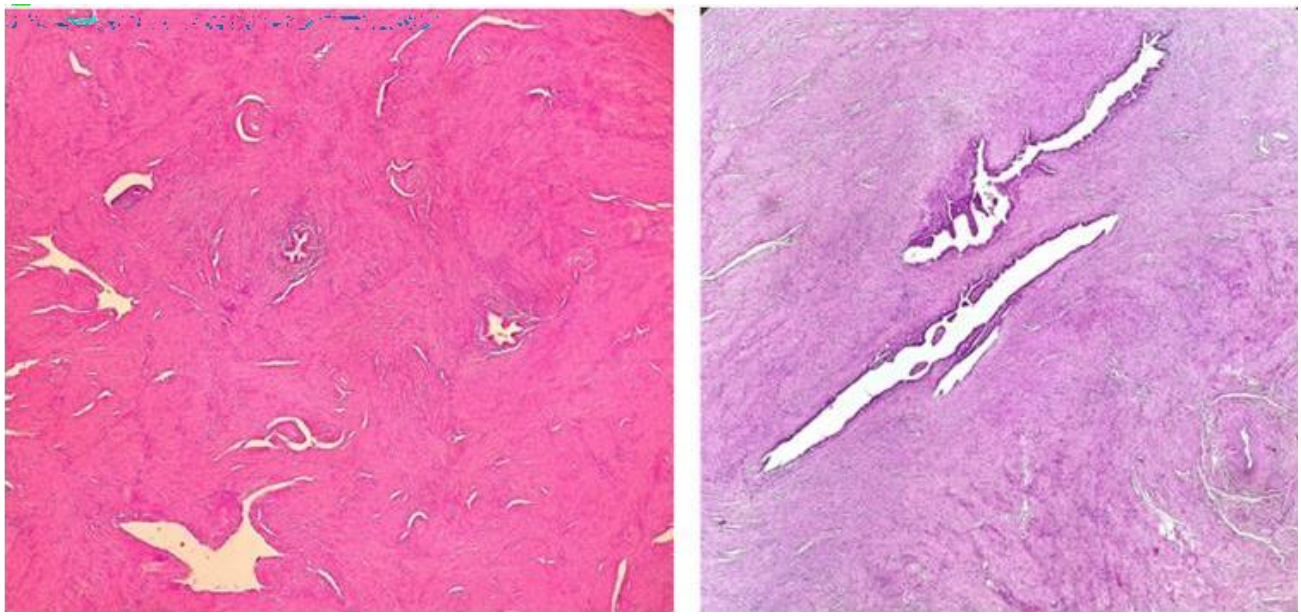

Figure 8: Histologic examination showing functional endometrium, with glands and stroma lining the cavity wall surrounded by irregularly arranged smooth muscle cells, resembling myometrium.

4. Discussion

All patients diagnosed with ACUM had a history of dysmenorrhea or noncyclic abdominal pain. These symptoms started soon after menarche in most cases and were probably caused by the accumulation of blood in the ACUM, raising the pressure inside the cavity, as seen in other obstructive congenital anomalies [14]. This patient presented with an atypical clinical course. After being asymptomatic for years under medical treatment, symptoms began to manifest at the age of 44. Patients with an undiagnosed ACUM that were successfully treated with combined oral contraceptives are less likely to undergo further diagnostic work-up and receive a later diagnosis. It is therefore unknown if age at treatment initiation, ACUM size, or other morphological characteristics are associated with successful medical treatment [9].

Intraoperatively, two lesions were identified on the left side of the uterus, which were histologically confirmed as ACUM in the broad ligament and an ovarian endometrioma. The ACUM has a size within the range typically described. Most lesions of ACUM commonly measured ≤50mm, according to certain reports [14,15]. Endometriotic lesions can be found in patients diagnosed with ACUM on various locations, including the uterosacral ligaments, the posterior peritoneum of broad ligaments, rectum, uterine surface, and even in ovaries [18,19], indicating that ACUM should kept in mind as a diagnosis in childbearing age women with severe dysmenorrhea, even if diagnosis of endometriosis is considered. As a result of the broad differential diagnosis and relatively low index of suspicion among clinicians, the diagnosis of ACUM may be significantly delayed [12].

5. Conclusion

ACUM has been reported more frequently during the past two decades, although it is still an extremely rare lesion [20]. This anomaly may be underestimated and underdiagnosed due to its rare presentation, non-specific symptoms, and the small volume of the mass. ACUM should be considered in patients with severe dysmenorrhea resistant to medical management. Early diagnosis and appropriate management via laparoscopic minimally invasive surgery can significantly improve the patient's quality of life [13]. It is important to select an appropriate individualized treatment plan based on the patient's clinical condition. The prognosis of this disease remains unknown, and clinicians must perform careful long-term follow-up of patients with ACUM [20].In reporting this rare case of an ACUM, we aim to offer clinicians a perspective that ACUM should be considered and investigated as a differential diagnosis in atypical cases of pelvic pain in women with pelvic lesions. However, we lacked long-term follow-up data in this case, necessitating the accumulation of further cases to obtain a more comprehensive understanding of the treatment prognosis [20]. This anomaly may be underestimated and underdiagnosed due to its rare presentation, non-specific symptoms, and the small volume of the mass. ACUM should be considered in patients with severe dysmenorrhea resistant to medical management. Early diagnosis and appropriate management via laparoscopic minimally invasive surgery can significantly improve the patient's quality of life [13].

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