Case Report

Accessory and cavitated uterine mass: A rare cause of dysmenorrhea and chronic pelvic pain

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ABSTRACT

Accessory and cavitated uterine mass (ACUM) is a rare developmental uterine anomaly that is said to develop due to duplication of ductal Mullerian tissue. It is an accessory cavity lying along the lateral wall of the uterus near the site of attachment of the round ligament and presents in young females with pelvic pain and dysmenorrhea. Histologically, the cavity is lined with endometrium and smooth muscle bundles similar to the myometrium. Ultrasound is helpful in the identification of the lesion. MRI of the pelvis is the imaging modality of choice for diagnosis. Laparoscopy and histopathology remain confirmatory. This report describes a case of ACUM in a young female who was managed by surgical excision of the mass.

Keywords: Accessory and cavitated uterine mass, Dysmenorrhea, Mullerian anomalies

The female reproductive system (uterus, cervix, upper vagina, and fallopian tubes) develops from a pair of Mullerian ducts through a staged process. A disruption in this process results in a variety of malformations known as Mullerian anomalies. Accessory and cavitated uterine mass (ACUM) is an uncommon developmental anomaly of the uterus, not classified under the Mullerian anomalies. ACUM is an accessory cavity lying along the lateral wall of the uterus near the site of attachment of round ligament and is said to develop due to duplication of Mullerian tissue [1,2]. The cavity is lined with endothelium and is surrounded by smooth muscle bundles resembling myometrium. It presents in young females with pelvic pain and severe dysmenorrhea. ACUM has been described in the previous literature with different names such as accessory cavitated masses and juvenile cystic adenomyoma. It remains an underdiagnosed entity and is often misdiagnosed on ultrasound as cystic adnexal mass, degenerated subserosal or broad ligament fibroid, cystic adenomyoma, or obstructed rudimentary horn. Proper knowledge of clinicoradiological features of ACUM is warranted to make a proper diagnosis and rule out other imaging mimics.

This report describes a case of ACUM in a young female and aims to discuss detailed clinical, radiological, and pathological features for a better understanding of this rare but treatable cause of dysmenorrhea.

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CASE REPORT

A 21-year-old nulliparous, not sexually active female, presented to our gynecology OPD with complaints of severe dysmenorrhea and pain abdomen for the past 3–4 years. The patient had taken multiple injectables and oral analgesics in the past for the same complaints. Her menstrual cycle was regular with normal flow.

On examination, her vitals were stable. General examination revealed no significant abnormality except for mild pallor. Tenderness was present in the right iliac fossa region. The rest of the systemic examination was unremarkable.

The previous ultrasound examination had reported subserosal/broad ligament fibroid with cystic degeneration. We performed a repeat transabdominal ultrasound that revealed a well-defined thick-walled cystic lesion of approximately size 12×10 mm with central echogenicity, along the right lateral uterine wall and separated from the right ovary. Magnetic resonance imaging (MRI) pelvis was performed for better characterization of the lesion. MRI demonstrated a well-defined round intrauterine cystic lesion along the right lateral wall of the uterus. The cyst contents were bright on T1W consistent with hemorrhagic products. A thick T2 hypointense rim of myometrium was noted (Fig. 1). No communication was seen between the lesion and the uterine cavity. Uterus otherwise appeared normal with normal-appearing bilateral adnexal structures and ovaries. Based on the imaging features on MRI and clinical profile, ACUM and obstructed rudimentary horn were considered as differential diagnoses.

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Diagnostic laparoscopy was performed followed by exploratory laparotomy under general anesthesia. Ovaries and fallopian tubes were found to be normal bilaterally. Both the cornual regions appeared normal ruling out the possibility of the obstructed rudimentary horn. A nodule of approximately size 1×1 cm was felt on the uterine surface near the right round ligament (Fig. 2). An incision was given on the uterine surface over the nodule. The incision was then given on the cystic lesion and chocolate-colored fluid was drained out followed by the excision of the cyst. Myometrial defect and abdomen were closed in layers. There were no immediate or early post-operative complications and the patient was discharged on post-operative day 3 in a satisfactory condition.

The histopathological section revealed an epithelial-lined cavity with embedded endometrial glands and stroma. Numerous smooth muscle bundles arranged in whorls and fascicles with intervening thin-walled blood vessels were noted. Pathological results were consistent with the clinicoradiological diagnosis of ACUM. The patient reported significant improvement in dysmenorrhea and pelvic pain in her menstrual cycle during the subsequent follow-up OPD visit.

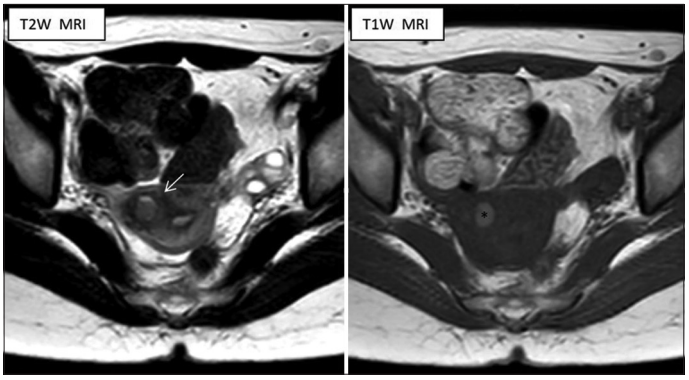

Figure 1: T2-weighted MR image of the pelvis (left) shows a well-defined round intrauterine intermediate signal intensity cystic lesion (arrow) along the right lateral wall of uterus surrounded by thick T2 hypointense rim of myometrium. The cyst contents appear hyperintense (asterisk) on T1-weighted sequence (right) likely representing hemorrhage

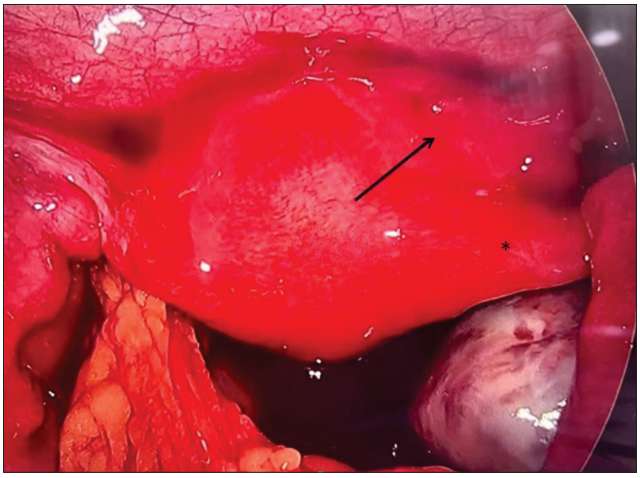

Figure 2: Intraoperative image shows a bulge (arrow) on the uterine surface near the right round ligament separate from the cornua (asterisk). An incision was given over the nodule and chocolate colored fluid drained out followed by excision of cyst

DISCUSSION

The female reproductive system (uterus, cervix, upper vagina, and fallopian tubes) develops from a pair of Mullerian ducts through a staged process. A disruption in this process results in a variety of malformations known as Mullerian anomalies. The American Society of Reproductive Medicine has classified Mullerian Anomalies into the nine categories which include cervical agenesis, unicornuate uterus, uterus didelphys, bicornuate uterus, septate uterus, longitudinal vaginal septum, transverse vaginal septum, and complex anomalies [3].

ACUM is a rare developmental anomaly of the uterus, not classified under the Mullerian anomalies. It is an accessory cavity located lying along the lateral wall of the uterus near the site of attachment of the round ligament [1]. It does not communicate with the main uterine cavity and the uterus remains otherwise normal. ACUM is considered to develop due to gubernaculum dysfunction and duplication of Mullerian tissue near the site of attachment of round ligament [2,4]. The cavity is lined with endothelium and is surrounded by smooth muscle bundles resembling myometrium.

It presents typically in young females, with chronic pelvic pain and dysmenorrhea (painful menstruation) due to repeated hemorrhage within the endometrium-lined cavity [5]. Azuma *et al*. reported severe dysmenorrhea non-responsive to any form of hormonal therapy, including low-dose estrogen-progestin combinations or progestin alone in their case report [6].

Ultrasound can be a helpful screening modality to identify the lesion but is not diagnostic. It is seen as a thick-walled cystic lesion with echogenic contents lying along the uterine wall and separated from the ovaries. A fluid level may or may not be present. Sometimes, the lesion is misdiagnosed as an adnexal lesion rather than uterine on ultrasound due to its peripheral location in the uterus. Jain and Verma reported a case of ACUM, which was reported as an adnexal mass on the previous ultrasound [2]. Unicornuate uterus with obstructed rudimentary horn, degenerated broad ligament or subserosal fibroid, and cystic degeneration in adenomyoma are other imaging differentials on ultrasound. Malhotra and Bajaj described a case of ACUM which had been previously reported as subserosal fundal fibroid on ultrasound [7]. MRI of the pelvis is a problem-solving tool. On MR imaging, it is seen as a cystic lesion within the uterus along the uterine wall and in close relation to the attachment site of the round ligament. The cystic contents are hyperintense on T1-weighted images consistent with hemorrhagic nature and are lined by T2 hyperintense endometrium. The cavity is surrounded by a thick T2 hypointense rim likely representing a hypertrophied myometrium. Normal visualization of the rest of the uterus and bilateral cornua is essential to rule out unicornuate uterus with an obstructed rudimentary horn as it remains a potential imaging differential [2,8]. This differentiation is, at times, difficult on MRI like in our case. In such cases, direct visualization on laparoscopy remains the only option for confirming the diagnosis.

Acién *et al*. had suggested criteria for the diagnosis of ACUM [9] which consists of the following features: Accessory

cavitated mass usually located under round ligament, normal uterus, fallopian tubes, and ovaries, surgical case with excised mass and pathological examination, accessory cavity lined by endometrial epithelium with glands and stroma, chocolate brown-colored fluid contents, and no adenomyosis in the resected uterus (small foci of adenomyosis in the myometrium of the accessory cavity may be seen).

The histopathological specimen shows an epithelial-lined cavity with embedded endometrial glands and stroma that are surrounded by smooth muscle bundles. These smooth muscle cells resemble myometrium and show positivity for estrogen and progesterone receptors (ER and PR positivity).

Since dysmenorrhea in patients with ACUM is usually non-responsive to conventional analgesics and hormonal therapies, surgical treatment is indicated. The treatment consists of surgical (laparoscopic) excision of the mass [10].

In our case, the patient presented with the typical complaints of dysmenorrhea for the past 3–4 years and had been taking over-the-counter analgesics for the same. Using ultrasound and MR imaging, we were able to narrow down the differential diagnoses to ACUM and obstructed rudimentary horn. Visualization of a cavitated mass under the round ligament with chocolate brown-colored contents, with normal appearance of bilateral cornua, fallopian tubes and ovaries on laparoscopy, and typical histopathological findings confirmed the diagnosis of ACUM.

CONCLUSION

We described ACUM, a rare developmental anomaly of the uterus causing chronic pelvic pain and dysmenorrhea. The entity is often misdiagnosed on ultrasound as cystic adnexal mass, degenerated subserosal or broad ligament fibroid, cystic adenomyoma, or obstructed rudimentary horn. MRI pelvis is the imaging modality of choice for diagnosis. Laparoscopy and histopathological examination are confirmatory. The condition is managed by the surgical excision of the mass.

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