SCIENCEFORECAST
A digital science library

Case Report
Published: 17 Sep, 2021

Journal of Gynecology and Obstetrics Forecast

Accessory and Cavitated Uterine Mass-Communicating Type: A Rare Mullerian Anomaly

Aswathy P, Priya PV and Sankar S*

Department of Pathology, Government Medical College, Kottayam, Kerala, India

Abstract

Background: Accessory and cavitated uterine mass (ACUM) is a rare clinical entity. Reported cases of ACUM have no communication with main uterine cavity. Here we report a case of mullerian anomaly resembling ACUM but having communication with main uterine cavity.

Case Presentation: 47 year old lady presented with lower abdominal distension and pain. Radiological evaluation: large pelvic cyst suggestive of pyometra or a peritoneal cyst. Laparotomy finding: a fully developed normal uterus and a large cystic mass near the left broad ligament suggestive of a degenerating broad ligament leiomyoma. The mass was removed along with uterus. Histopathological finding: uterus showed normal morphology and the broad ligament mass showed a circumscribed cystic lesion with morphology similar to uterus with a narrow communication between two. Diagnosis: Accessory and Cavitated uterine mass- communicating type.

Conclusion: ACUM is a rare Mullerian anomaly often misdiagnosed as degenerating leiomyoma or adenomyoma and so may be under reported.

Keywords: Accessory and cavitated uterine mass; Mullerian anomaly; Uterus like mass

Introduction

Accessory and cavitated uterine mass (ACUM) in a woman with an otherwise normal appearing uterus, is an extremely rare mullerian anomaly without uterine malformation, caused by duplication and persistence of ductal Mullerian tissue at the insertion of round ligament [1]. ACUM is not categorized as a distinct class in ASRM classification of anomalies of mullerian ducts [2]. Generally clinical symptoms develop at younger age, before 30 years [3]. ACUM is a treatable cause of infertility. Other usual symptoms are dysmenorrhea and pelvic mass. As age advances, the mass becomes larger and often misdiagnosed as degenerating broad ligament leiomyoma oradenomyomas. MRI is the investigation of choice and diagnosis is made by histopathological study of the lesion [4]. 68 cases of ACUM have been reported in literature till date [5-9]. But no cases of communicating ACUM have been reported so far.

Case Report

A 47 year old multiparous lady presented to gynecology outpatient department with history of amenorrhea and pelvic pain for 2 years and lower abdominal distension for 4 months. USG study of abdomen showed a pelvico-abdominal large cystic mass and suggested the possibility of pyometra or a peritoneal cyst. CT abdomen showed a large cystic mass lesion measuring 16.8x11.5x12.3 cm with enhancing wall, which closely abutting the uterus. The clinical diagnosis considered was broad ligament fibroid with degenerative change and the clinician proceeded with laprotomy. Preoperatively surgeon found a large cystic mass measuring 20x10x10 cm attached to the posterolateral surface of uterus near the left broad ligament. Uterus was normal shaped and fully developed and showed a small subserosal fibroid near the medial end of left fallopian tube. Both fallopian tubes and ovaries were seen in normal relation to uterus. Cystic mass was removed along with uterus, both fallopian tubes and both ovaries. No free fluid or endometriotic focus was identified in abdominal or pelvic cavity.

Gross examination of surgical specimen showed a large cystic mass attached to left postero-lateral surface of uterus. Cystic mass had a muscular wall with shaggy inner surface and 750 ml of tarry dark brown fluid within the cavity. A small fibroid measuring 2.8x2x2 cm was noted in the muscular wall of this cyst in one focus. Left fallopian tube showed minimal hematosalpinx. No abnormality was seen in other fallopian tube and both ovaries. Uterus showed normal endometrial thickness

OPEN ACCESS

**Correspondence:*
Aswathy P, Department of Pathology, Government Medical College, Kottayam, Kerala, India.
E-mail: aswathybalan23@gmail.com

Received Date: *04 Aug 2021*
Accepted Date: *13 Sep 2021*
Published Date: *17 Sep 2021*

Citation: *Aswathy P, Priya PV, Sankar S. Accessory and Cavitated Uterine Mass-Communicating Type: A Rare Mullerian Anomaly. J Gynecol Obstet Forecast. 2021; 4(1): 1015.*

Figure 1: Uterus with attached bilateral tubes and ovaries and the large cystic mass attached to its lower part.

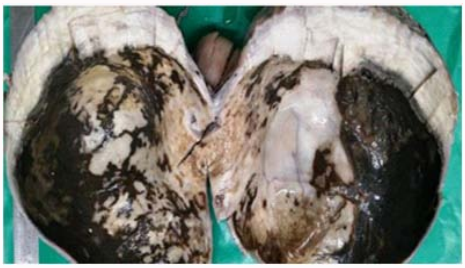

Figure 2: Cut section of the cystic mass showing shaggy and grey white inner wall and thick and thin areas.

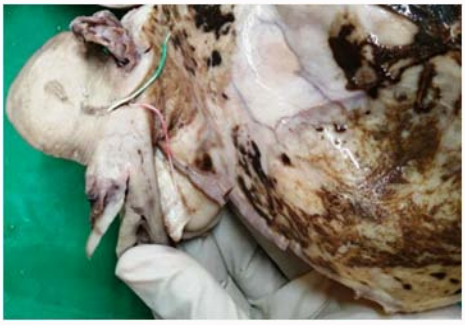

Figure 3: Shows communication of mass with uterine cavity (red thread-connection between uterine cavity and cervix, green thread-communication between mass and uterine cavity).

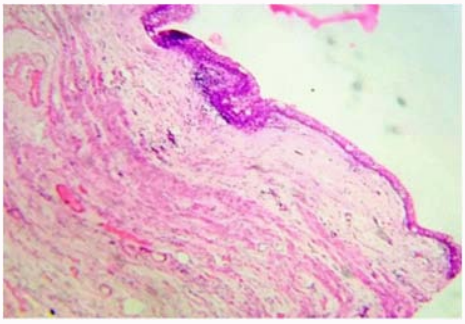

Figure 4: Microscopy of the cyst shows focal lining by columnar to cuboidal epithelium with squamous metaplasia.

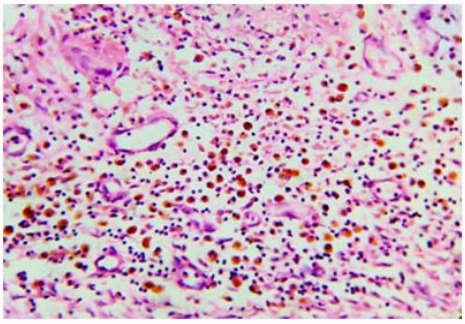

Figure 5: Microscopy of the cyst focally lined by hemosiderin laden macrophages and underlying smooth muscle (40X).

and two small fibroids in the myomerium. A narrow connection between cavity of uterus and the cystic mass was identified near the lower uterine segment. Microscopy of the cystic mass showed outer thick layer of orderly placed smooth muscle bundles and inner thin layer of epithelial lining by cuboidal/columnar cells with extensive ulceration, and focal squamous metaplasia. The muscular layer of cystic mass showed a focus of small leiomyoma. Microscopy of uterus showed small leiomyomata. No focus of adenomyosis was seen. Left fallopian tube showed findings consistent with hematosalpinx. No abnormality was seen in opposite fallopian tube, both ovaries and cervix. Considering the location and morphology of the large cystic mass lesion and the presence of an otherwise normal appearing uterus in this patient, we made a diagnosis of mullerian anomaly possibly accessory and cavitated uterine mass having communication with uterine cavity (communicating ACUM).

Discussion

Mullerian ducts are the primordial anlage of the female reproductive tract, which differentiate to form fallopian tubes, uterus, cervix and upper two-third of vagina. Any failure in its normal development leads to Mullerian malformation. Congenital anomalies of the mullerian system occur due to discrete disturbances in the embryologic development of mullerian ducts during fetal life. The ESHRE-ESGE classification of Mullerian anomalies is designed to include all the diseases resulting from a defect in the formation, fusion or absorption process of normal embryological development of uterus. ESHRE-ESGE class 6 is meant for the still unclassified infrequent uterine anomalies, duplication defects and ectopic Mullerian tissue anomalies [5,6].

Accessory and cavitated uterine mass (ACUM) is a mullerian anomaly caused by duplication and persistence of ductal Mullerian tissue at the insertion of round ligament, believed to be due to the gubernaculums dysfunction ACUM is a mass which grossly and microscopically resembles uterus. Usually ACUMs seen as a cystic mass filled with chocolate coloured contents. This may be due to the cyclical changes occurring in the endometrial lining during each menstrual cycle. Histopathology of the cyst wall shows the presence of endomyometrial lining [1].

Often ACUM is confused with non-communicating rudimentary uterine horn, true cavitated adenomyoma and degenerating fibroid, so histopathological study is essential for diagnosis. The diagnostic criteria for ACUM are presence of 1. An isolated accessory cavitated mass, 2. A normal uterus with endometrial cavity, fallopian tubes and ovaries, 3. a surgical case with an excised mass and a pathological examination, 4. accessory cavity lined by endometrial epithelium with glands and stroma, 5. chocolate brown coloured fluid content, and no adenomyosis in normal uterus (if the uterus has been removed) but there be small foci of adenomyosis in the myometrium adjacent to the accessory cavity [7] (Figure 1,2,3,4 and 5). In contrast to ACUM the

non-communicating rudimentary uterine horn is usually associated with uterine malformation (unicornuate uterus, bicornuate uterus). True cavitated adenomyoma can be differentiated by absence of internal epithelial lining, lack of uterus like smooth muscle organization in the cystic cavity and diffusely spread adenomyotic foci in the uterus corpus. Degenerating fibroid also lacks internal epithelial lining [1].

Our patient had a fully developed normal uterus with attached bilateral tubes and ovaries. The large mass closely abutted on the left lateral side of the normal uterus showed chocolate coloured hemorrhagic fluid in the cavity, an inner epithelial lining and an outer smooth muscle wall, which were compatible with the diagnostic criteria of ACUM. No focus of adenomyosis or endometriosis was observed. Areas of ulceration and squamous metaplasia in luminal epithelial lining and communication with endometrial cavity of normal uterus were the additional findings we observed in our case, which were not previously reported so far.

Excision of mass with or without hysterectomy is the treatment of choice of ACUM. Harith *et al.,* reported that conception can occur in ACUM, however, it is very rare and may be difficult to diagnose.

Conclusion

ACUMs are rare Mullerian anomalies; only 68 cases are reported till date. Clinically most of the cases are misdiagnosed as broad ligament leiomyomas with degeneration or adenomyomas and histopathological examination is diagnostic. The atypical finding observed in our case was communication of ACUM with endometrial cavity of developed normal uterus, which is not reported so far.

References

1. Supermaniam S, Thye WL. Diagnosis and laparoscopic excision of accessory cavitated uterine mass in young women: Two case reports. Case Rep Womens Health J. 2020; 26: e00187.
2. The American Fertility Society classifications of adnexal adhesions, distal tubal occlusion, tubal occlusion secondary to tubal ligation, tubal pregnancies, müllerian anomalies and intrauterine adhesions. Fertil Steril. 1988; 49: 944-955.
3. Malhotra K, Bajaj B. A case report on ACUM: a rare mullerian anomaly. Int J Reprod Contracept Obstet Gynecol. 2020; 9: 1310-1314.
4. Acien P, Bataller A, Frenandez F, Acien MI, Rodriguez JM, Mayol MJ. New cases of accessory and cavitated uterine masses (ACUM): a significant cause of severe dysmenorrhea and recurrent pelvic pain in young women. Hum Reprod. 2012; 27: 683-694.
5. Grimbizis GF, Gordts S, Di Spiezio SA, Brucker S, De Angelis C, Gergolet M, et al. The ESHRE/ESGE consensus on the classification of female genital tract congenital anomalies. Hum Reprod. 2013; 28: 2032-2044.
6. Ludwin A, Ludwin I. Comparison of the ESHRE-ESGE and ASRM classifications of Müllerian duct anomalies in everyday practice. Hum Reprod. 2015; 30: 569-580.
7. Acién P, Acien M, Fernandez F, Jose MM, Aranda I. The cavitated accessory uterine mass: a Mullerian anomaly in women with an otherwise normal uterus. Obstet Gynecol. 2010; 116: 1101-1109.
8. Alkhateeb HM, Yaseen EM. Twin pregnancy in an accessory cavitated non-communicating uterus. Int J Surg Case Rep. 2015; 10: 45-48.
9. Vani M, Sonia D, Smiti N, Meenakshi C, Vandhana B. Accessory and cavitated uterine mass: Is it a mullerian duct anomaly. Gyne Surg J. 2020; 36: 350-356.