

# Multiloculated cystic Mullerianosis of uterus: A case report

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## Abstract

We are reporting a case of Mullerianosis, which presented as a multiloculated cystic mass on the serosal surface of the fundus of the uterus. Clinically and radiologically, this was interpreted as an ovarian tumor. Mullerianosis is a very rare benign tumor-like lesion. Awareness of this lesion is necessary to avoid misdiagnosis by clinicians, radiologists, and pathologists. (J Turk Ger Gynecol Assoc 2014; 15: 197-200)

**Key words:** Fundus of uterus, Mullerianosis, multiloculated cystic mass

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## Introduction

Mullerianosis was defined by Young and Clement (1) in 1996 as “a lesion seen at any site containing admixtures of endosalpingiosis, endometriosis, and endocervicosis -the three Mullerian glandular epithelia of tubal, endometrial, and endocervical type.” For diagnosis of Mullerianosis, presence of an admixture of at least two types of Mullerian epithelium is necessary.

If only one type of epithelium is seen, the lesion is referred to as endometriosis, endosalpingiosis, or endocervicosis as per the type of lining cells present. Of these, endometriosis is a very common lesion and may be seen in any site, the most common site being the ovary. Endosalpingiosis and endocervicosis are rarer and have been reported mostly in the urinary bladder. Mullerianosis in which at least two types of Mullerian epithelium should be present is still rarer, and only a few cases have been reported in the English literature. Mullerianosis has been reported in sites, like the pelvic peritoneum, urinary bladder, ureter, and inguinal lymph node. But, Mullerianosis of the uterine fundus is very rarely reported.

Here, we report a case of Mullerianosis located in the fundus of the uterus presenting as a multiloculated cyst, clinically and radiologically mimicking an ovarian malignancy.

## Case Presentation

A 48-year-old female presented with swelling in the lower abdomen. The clinical diagnosis was ovarian tumor. Imaging studies showed an ovarian mass. Total abdominal hysterectomy with bilateral salpingo-oophorectomy was done. The specimen received showed a uterus with both adenexae and

a polypoid cystic mass attached to the fundus of the uterus. The uterus measured 8x5.5x2.5 cm. The cystic mass measured 9x8x9 cm. The cystic mass, on cutting open, showed a multiloculated cyst with locules of varying sizes filled with clear fluid. The wall was thin with a smooth inner surface. No solid areas were identified (Figure 1). Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A cut section of the uterus showed two intramural fibroids, the larger one measuring 1 cm at the greatest diameter. The endometrium, endocervix, and ectocervix were unremarkable. The left ovary measured 3x1.5x1 cm and showed a cyst attached to one pole measuring (m/s) 3 cm at the greatest diameter, which on opening showed a thin-walled uniloculated cyst filled with clear fluid. The right ovary and both fallopian tubes were grossly unremarkable.

Microscopic examination of the fundal cyst showed a multiloculated cyst with microcystic spaces. The locules of the cysts were lined by variable types of epithelium, the predominant being ciliated columnar epithelium of the tubal type (Figure 2). Some of the locules were lined by flat to cuboidal epithelium (Figure 3). Focal areas showed closely packed glandular structures with stratification of the lining epithelium (Figure 4). Cellular atypia was not found in these areas. No mitotic activity was seen; the low proliferative nature was confirmed by Ki67 immunostaining. The cystic spaces were filled with eosinophilic secretion with neutrophil infiltrate in some spaces. Focally, endometrial-type small glandular structures were seen (Figure 5). The stroma was also variable in nature. It was myomatous, fibrous, and myofibromatous in areas (Figure 5, 6). Multicystic mesothelioma was a differential diagnosis grossly and histologically. The calretinin immunostaining was negative in the glandular lining cells, which ruled out this differential diagnosis.





Figure 1. Cut section of uterus showing multiloculated cystic mass attached to the serosa of the fundus

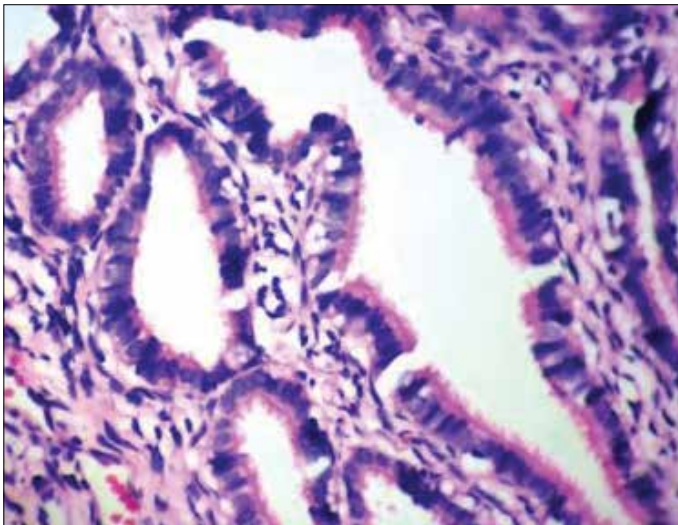


Figure 2. Locules and glandular spaces lined by ciliated tall columnar epithelium of tubal type (H&E x400)

The uterus showed 2 small intramural leiomyomas, proliferative endometrium, and chronic cervicitis. The left ovary showed a simple serous cyst. The right ovary and both fallopian tubes were histologically unremarkable.

Immunohistochemistry for cytokeratin (CK) was strongly positive in the lining cells. Calretinin was negative. Ki-67 showed no proliferative activity in the glandular epithelium (Figure 7).

## Discussion

Mullerianosis was defined by Young and Clement in 1996 as a lesion seen at any site containing Mullerian glandular epithelia of tubal, endocervical, and endometrial type (1). At least any two of the Mullerian tissues should be present for the diagnosis of Mullerianosis. Mullerianosis was considered a choristoma of the Mullerian rest (2). This is a benign tumor-like lesion and

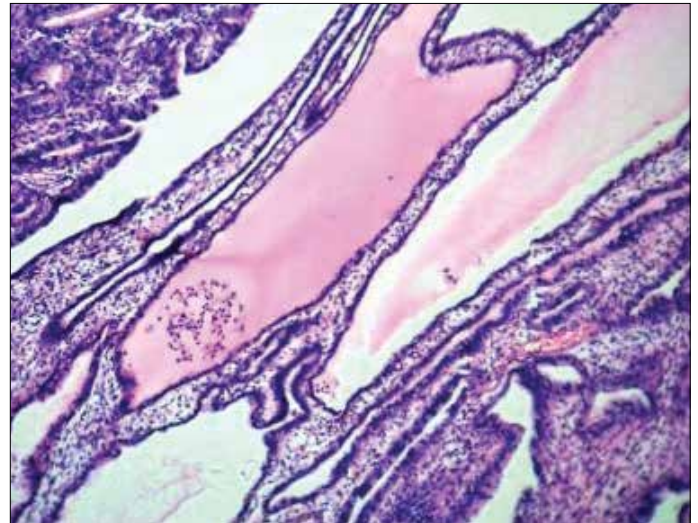


Figure 3. Locules of the cyst lined by flat to low cuboidal epithelium. Lumen shows eosinophilic secretion and neutrophil collection (H&E x100)

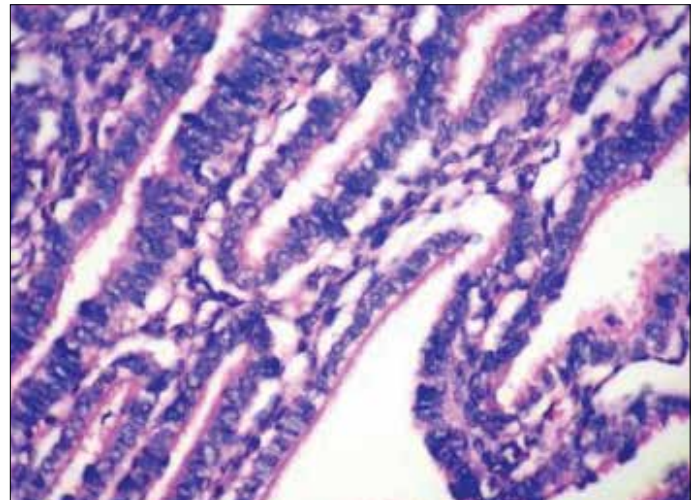


Figure 4. Crowded glandular structures showing stratification of nuclei (H&E x400)

has been reported in the urinary bladder, mesosalpinx, pelvic peritoneum, and inguinal lymph nodes (3-5).

If only the tubal-type epithelium is present in the lesion, the condition is termed endosalpingiosis. Endosalpingiosis was first described by Sampson in 1930 (6).

Endosalpingiosis is usually an incidental finding. But, sometimes, it may present as a tumorous mass. Clement and Young described four cases of florid cystic endosalpingiosis presenting as a tumor-like mass (7). The fifth case of florid endosalpingiosis had been reported in the uterine fundus as a multiloculated cystic mass (8), grossly similar to our case. But, histologically, only endosalpingiotic tissue was present and lacked the endometrial-type glands, as seen in the present case. In the case of florid cystic endosalpingiosis reported by M Heatley et al. (9), there were multiple small cysts in the uterus extending into the parametrium and broad ligament. Batt et al. (2) suggested that for the diagnosis of Mullerianosis, the following criteria

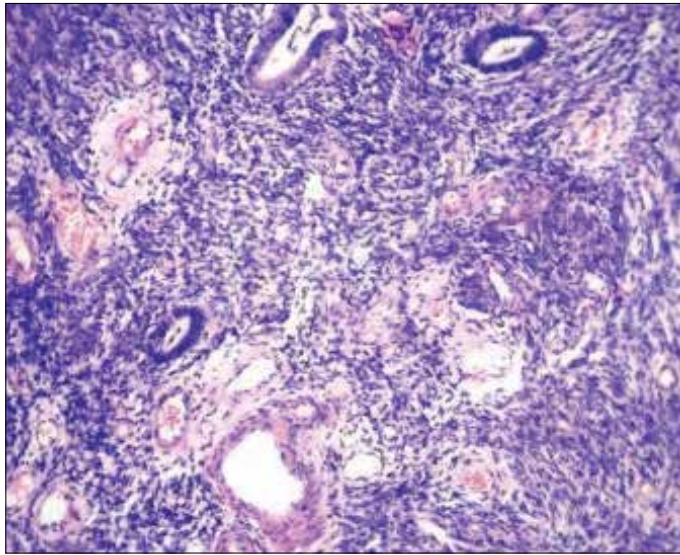


Figure 5. Endometrial type glands in a fibrous stroma with prominent hyalinized vessels (H&E x100)

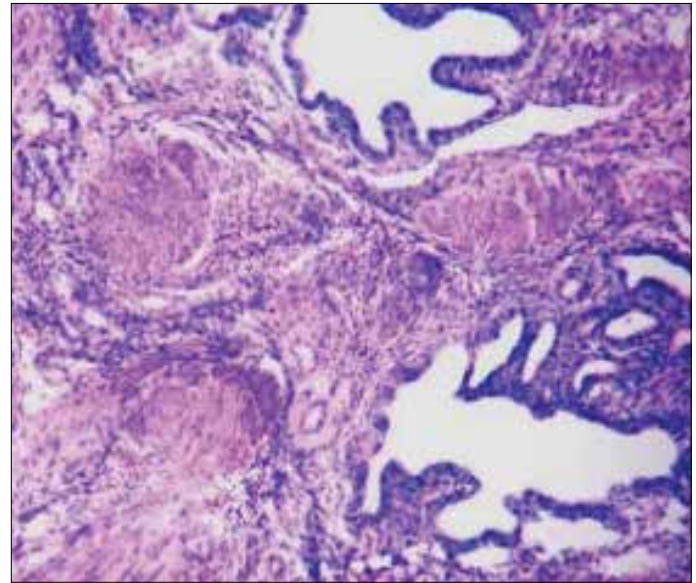


Figure 6. Smooth muscle stroma (H&E x100)

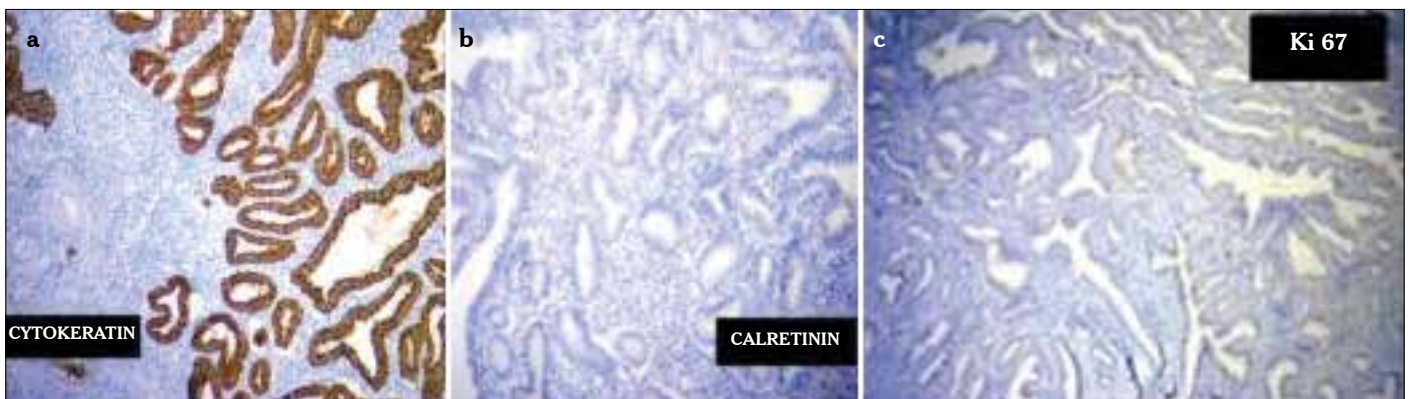


Figure 7. a-c. Immunostains (x100) showing cytokeratin positivity in the glandular epithelial cells (a). Calretinin is negative, ruling out mesothelial cyst (b). Ki-67 shows low proliferative index, ruling out an adenocarcinoma (c)

should be met: the patient should have no evidence of pelvic endometriosis; no history of surgery to the reproductive organs; and no direct communication of the lesion with the endocervix, endometrium, or endosalpinx. In our case, all these criteria were satisfied.

The pathogenesis of this lesion is still in debate. A developmental theory (2), an implantation theory (10), and a metaplastic theory (11) have been put forward. In our case, the patient was a middle-aged female, and no associated developmental anomalies were seen to propose a developmental theory. The metaplastic theory seems to be more appropriate in this case. The tumor might have originated from the serosal (peritoneal) covering of the uterine fundus, probably by a metaplastic conversion of the mesothelial cells to Mullerian epithelium.

The clinical importance of this lesion is that it must be distinguished from malignancy, because the glandular structures and cell/nuclear stratification may resemble an adenocarcinoma. Mullerianosis is a benign lesion having no invasion to the deeper tissue, whereas adenocarcinoma is invasive to the adjacent tissue. Adenocarcinomas show cellular features of malignancy

and high proliferative index. In our case, no invasion to the adjacent fundal myometrium was seen. Though there were some focal glandular crowding and nuclear stratification, mitotic activity was not seen. Ki-67 immunostaining showed no proliferative activity. This lesion is also a close mimicker of mesothelioma, which may present as a multiloculated cystic mass and may be seen attached to the uterine serosal surface. But, microscopically, the tubal- and endometrial-type epithelium is characteristic of Mullerianosis. Moreover, in our case, calretinin immunostaining was negative, ruling out mesothelioma

To conclude, Mullerianosis is a very rare benign tumor-like lesion. The fundus of the uterus is a rare site, and the previously reported sites have been the pelvic peritoneum, urinary bladder, mesosalpinx, and inguinal lymph nodes. Awareness of this rare lesion is helpful for the clinician and radiologists to avoid a misdiagnosis of ovarian tumor. Pathologists should not misdiagnose this lesion as a malignancy and should differentiate it from adenocarcinoma and mesothelioma with appropriate immunostaining if the morphological diagnosis is difficult.

**Ethics Committee Approval:** N/A.

**Informed Consent:** Written informed consent was obtained from patient who participated in this case.

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