

## Case Report

# Florid cystic endosalpingiosis associated with a retroperitoneal leiomyoma mimicking malignancy: a case report

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**Abstract:** Florid cystic endosalpingiosis (FCE) is a rare type of endosalpingiosis that presents as a mass-like lesion. Here we report an unusual case of FCE associated with a retroperitoneal leiomyoma. A 46-year old female presented with a palpable abdominal mass. A pelvic CT revealed a 23.5×16.3×9.4 cm sized multilocular cystic and solid mass in the retroperitoneum. Surgical excision of the mass was performed. Microscopically, the cystic spaces were lined by a single layer of ciliated tubal epithelium. The solid areas consisted of thick bundles of spindle cells. There were no cytologic atypia, mitosis or necrosis. The spindle cells were positive for actin and desmin, and were negative for c-kit, CD34, S100 and HMB-45, confirming the diagnosis of FCE associated with retroperitoneal leiomyoma.

**Keywords:** Florid cystic endosalpingiosis, endosalpingiosis, retroperitoneal mass, leiomyoma, mimicker

## Introduction

Endosalpingiosis refers to the presence of ectopic tubal-like ciliated epithelial cells and is one of the triad forms of Mullerianosis [1, 2]. Although it is mostly asymptomatic, florid cystic endosalpingiosis (FCE) is a rare type of endosalpingiosis that presents as a mass-like lesion [1]. Here we report an unusual case of FCE associated with a retroperitoneal leiomyoma.

## Case presentation

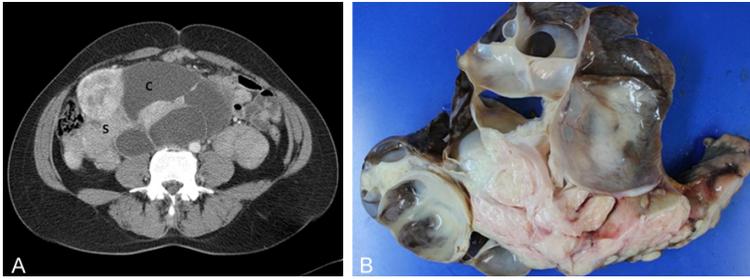
A 46-year-old female presented with a palpable abdominal mass. She previously had a total hysterectomy without salpingo-oophorectomy fourteen years ago, due to a 5.0×5.0×5.0 cm sized intramural leiomyoma. Pelvic CT revealed a 23.5×16.3×9.4 cm sized lobulated mass with multiloculated cysts and solid portions (**Figure 1A**). The mass was located on the right side of pelvic cavity extending superiorly to the right anterior pararenal space, which was in the retroperitoneum. The retroperitoneal mass was excised. On gross examination, a 23.5×16.3×9.4 cm sized gray to white colored lobulated mass was noted. On cut section, multilocular cysts

were mainly located at the peripheral area, and white colored multinodular solid areas with whirling features were noted at the central area (**Figure 1B**). On microscopic examination, the cystic spaces were lined by a single layer of tubal epithelium consistent with endosalpingiosis. The solid areas consisted of thick bundles of spindle cells with elongated cigar shaped nucleus. There were no cytologic atypia, mitosis or necrosis. On immunohistochemistry, the lining of the cyst were positive for cytokeratin 7. The solid areas were positive for actin and desmin, and were negative for c-kit, CD34, S100 and HMB-45, confirming the diagnosis of florid cystic endosalpingiosis in a leiomyoma (**Figure 2**).

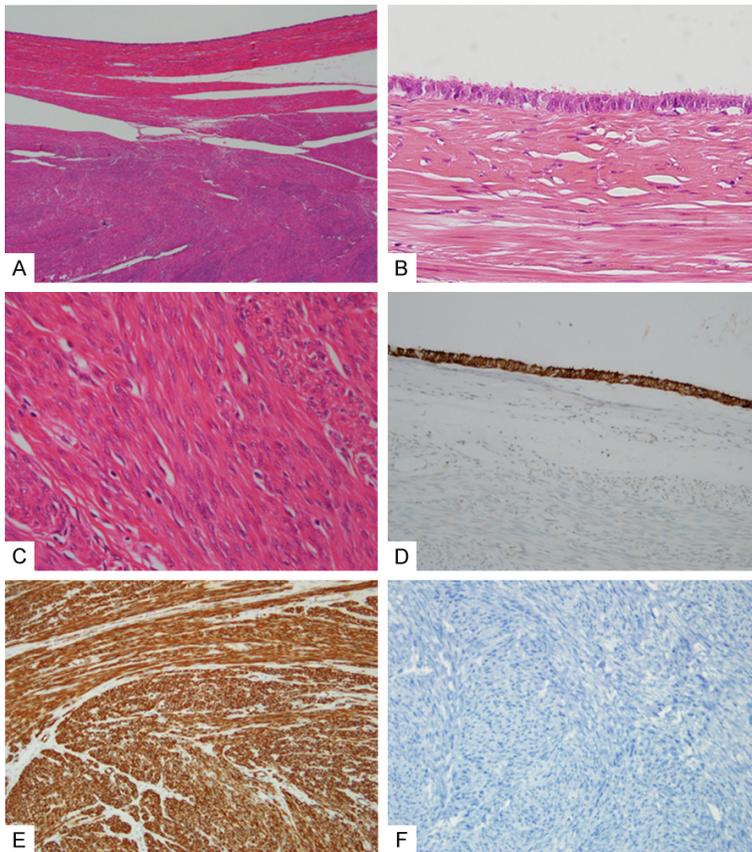
## Discussion

Endosalpingiosis is one of the triad of non-neoplastic secondary Mullerian lesions, with the others being endocervicosis and endometriosis [1]. Endosalpingiosis is mostly an asymptomatic lesion. However, its rare tumor-like presentation has been reported as florid cystic endosalpingiosis (FCE) by Clement and Young [1]. FCE has been reported in various areas,

## FCE associated with a retroperitoneal leiomyoma



**Figure 1.** Radiologic and gross findings. A. Pelvic CT reveals a 23.5×16.3×9.4 cm sized retroperitoneal mass at the right anterior pararenal space. It has a lobulated contour with multiloculated cystic (C) and solid (S) portions. B. Gross examination reveals a gray to white colored, lobulated solid and cystic mass, measured 23.5×16.3×9.4 cm. On cut section, multilocular cysts filled with clear serous fluid were mainly located at the peripheral area, and white colored multinodular solid areas with whirling features were present at the central portion.



**Figure 2.** Microscopic findings. A. Solid mass lesion at the lower field with cystic area at the upper field. B. The cysts were lined with a single layer of ciliated tubal epithelium. There are no endometrial stroma-like areas near the epithelial lining. C. The solid area consisted of thick bundles of spindle cells with elongated cigar shaped nuclei. D. On immunohistochemistry, the epithelial lining were positive for cytokeratin 7. E. The spindle cells were positive for actin. F. The spindle cells were negative for HMB-45.

leen, colon and appendix [1, 3-5]. It most commonly involves the uterus [4, 6, 7]. In the present case, the patient had a previous total hysterectomy, and the location of the lesion was not only extrauterine, but also retroperitoneal. The unusual location, large size and solid and cystic appearance warranted a retroperitoneal sarcomatous lesion. However, absence of cytologic atypia, mitosis and necrosis ruled out malignancy. Differential diagnosis included lesions that could have biphasic features of epithelial and spindle cell components. The single layer of ciliated columnar epithelium without any endometrial stroma-like areas ruled out the possibility of endometrial lining, and endometriosis and extrauterine adenomyoma were excluded. The epithelial component was thought to be FCE. Differential diagnosis of the spindle cell-sarranged in thick bundles included adenofibroma, extra gastrointestinal stromal tumor, angiomyolipoma, lymphangiomyomatosis (LAM) and leiomyoma. Extra gastrointestinal stromal tumor was ruled out due to negative staining results for c-kit and CD34. Immunoreactivity for actin and desmin in the spindle cells confirmed a smooth muscle differentiation and adenofibroma was ruled out. The lack of adipose cell component and HMB-45 negativity ruled out the possibility of angiomyolipoma. The lack of slit-like vascular spaces and HMB-45 negativity ruled out the possibility of LAM. The final diagnosis of this case was FCE associated with a retroperitoneal leiomyoma.

such as the uterus, ovaries, fallopian tubes, paraovarian areas, urinary bladder, ureter, sp-

There are two interesting points noted in the present case of FCE; its occurrence within a

## FCE associated with a retroperitoneal leiomyoma

**Table 1.** Review of literature about florid cystic endosalpingiosis (FCE) involving leiomyomas and endosalpingiosis presenting as retroperitoneal mass lesions

| <b>FCE associated with leiomyoma</b>                               |     |                                                                                                     |                        |                                                                                                                                           |                                                                                                                                                                                                                                                                                                                                                                                              |
|--------------------------------------------------------------------|-----|-----------------------------------------------------------------------------------------------------|------------------------|-------------------------------------------------------------------------------------------------------------------------------------------|----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Authors                                                            | Age | Presentation                                                                                        | Surgery                | Gross findings                                                                                                                            | Microscopic findings                                                                                                                                                                                                                                                                                                                                                                         |
| Suarez-Vilela et al. [8]                                           | 51  | Metrorrhagia<br>Endometrioid carcinoma, FIGO grade 2                                                | TAH with BSO           | 5×3 cm sized cystic area in a<br>8×7×5 cm sized subserosal mass                                                                           | Cysts lined by tubal epithelium. Walls of the cysts and stroma around the glands were composed of fibrovascular connective tissue. Stromal component was sharply delimited of the muscular fascicles of the subserosal uterine myoma at the periphery.                                                                                                                                       |
| Driss et al. [9]                                                   | 47  | Vaginal bleeding and pelvic mass                                                                    | TAH with BSO           | Numerous 1-2 mm cysts sprinkled on a 13×13×10 cm sized deep red colored exophytic mass extending from the uterus to the broad ligament    | Cotyledonoid dissecting leiomyoma associated with numerous glands and cysts lined by a ciliated tubal type epithelium. No cellular atypia or increased mitotic activity<br>No necrosis.                                                                                                                                                                                                      |
| <b>Endosalpingiosis presenting as retroperitoneal mass lesions</b> |     |                                                                                                     |                        |                                                                                                                                           |                                                                                                                                                                                                                                                                                                                                                                                              |
| Authors                                                            | Age | Presentation                                                                                        | Surgery                | Gross findings                                                                                                                            | Microscopic findings                                                                                                                                                                                                                                                                                                                                                                         |
| Matsui et al. [11]                                                 | 37  | Incidental finding during 4 <sup>th</sup> Cesarean section<br>Pulmonary LAM diagnosed 6 years later | Mass excision          | Two encapsulated masses measuring 20cm and 1.5 cm. The larger mass was composed of multiple cysts filled with pale yellow-tan milky fluid | Heterogenous population of LAM cells present throughout the solid areas in fascicles separated with slit-like vascular channels. Stratified ciliated epithelial cells lining the cystic spaces.<br>Positive for HMB-45.<br>Diagnosis: Endosalpingiosis associated with LAM in a retroperitoneal lymph node                                                                                   |
|                                                                    | 50  | Abdominal discomfort                                                                                | Exploratory laparotomy | Encapsulated retroperitoneal cystic mass, measured 4×4×3 cm<br>Soft, yellow-tan lobulated mass with cystic changes on cut sections        | Heterogenous population of LAM cells present throughout the solid areas in fascicles separated with slit-like vascular channels. Stratified ciliated epithelial cells lining the cystic spaces.<br>Positive for HMB-45.<br>Diagnosis: Endosalpingiosis associated with LAM in a retroperitoneal lymph node                                                                                   |
| Fukunaga et al. [12]                                               | 25  | Painless palpable abdominal mass<br>No history of tuberous sclerosis                                | Mass excision          | Multilocular retroperitoneal cystic mass, measured 4.0×3.5×3.5 cm                                                                         | Three components; multiple cysts lined by ciliated columnar epithelium, condensation of small stromal cells immediately subjacent to the cystic epithelium and thick exterior wall composed of plump spindle cells in a fascicular pattern and slit-like vascular spaces<br>Immunoreactive for HMB-45, smooth muscle actin, and h-caldesmon<br>Diagnosis : LAM arising from endosalpingiosis |

TAH: total abdominal hysterectomy, BSO: bilateral salpingo-oophorectomy, LAM: lymphangi leiomyomatosis.

leiomyoma, and its unusual location in the retroperitoneum. Two cases of FCE associated with a uterine leiomyoma have been previously reported in the literature. Suarez-Vilela *et al.* reported a case of FCE within a uterine subserosal leiomyoma [8]. In the case report of Driss *et al.*, FCE was associated with a cotyledonoid leiomyoma extending from the uterus to the broad ligament [9]. The details of the previously described cases are reviewed in **Table 1**. Other than uterine leiomyoma, Bermejo *et al.*, reported a case of FCE associated with peritoneal leiomyomatosis [10]. As there had been no other mass-like lesion visible at the time of total hysterectomy, the leiomyoma in the present case is considered truly retroperitoneal, and not a parasitic leiomyoma. Three cases of endosalpingiosis have been reported to present as retroperitoneal mass lesions in the literature [11, 12]. However, all three cases were associated with extrapulmonary lymphangi-oleiomyomatosis (LAM). Matsui *et al.* reported two cases of endosalpingiosis associated with LAM in a retroperitoneal lymph node [11]. Fukunaga *et al.* reported a case of retroperitoneal LAM associated with endosalpingiosis [12]. The detailed clinical features are reviewed in **Table 1**. To our knowledge this is the first case of a FCE associated with a retroperitoneal leiomyoma.

There are three major theories for the etiology of endosalpingiosis [2]. Metaplastic change of the celomic epithelium into tubal epithelium is the most widely accepted theory [2, 10]. Others suggest the theory of implantation, occurring through transplantation of tubal mucosa to peritoneal surfaces during surgery [2]. Lastly, some suggest the lymphatic pathogenesis [13]. As the cystic areas were mostly located beneath the surface of leiomyoma, and the patient had a previous history of abdominal surgery, both celomic metaplasia and implantation theory would be acceptable in this case.

We report a rare case of FCE associated with a retroperitoneal leiomyoma, which may clinically mimic a retroperitoneal malignancy with its unusual presentation and location. Full understanding of the histologic features and awareness of this unusual clinical setting will facilitate accurate diagnosis and prevent over-treatment.

### Disclosure of conflict of interest

None.

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## FCE associated with a retroperitoneal leiomyoma

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