**CEOG** Clinical and Experimental Obstetrics & Gynecology

**Case Report**

# A case report of acute ovarian cyst torsion by female adnexal tumor of probable Wölffian origin

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**Summary**

*Objective:* The authors report a rare case of female adnexal tumor of probable Wölffian origin ( FATWO), presenting with acute ovarian torsion and treated by laparoscopic surgery while preserving fertility in a young woman. *Case Report:* A 30-year-old woman was referred to this clinic for evaluation of an adnexal mass. The tumor was found to have originated from the right tubal fimbria and had torqued three times. When the frozen section proved negative for malignancy, a right salpingectomy was performed. Based on the pathological and immunohistochemical findings, the final diagnosis was concluded to be a FATWO. Adjuvant therapy was not administered because the patient wanted to preserve her fertility. No evidence of recurrence has been observed during the past five years. *Conclusion:* Fertility preserving treatment should be considered in young women with FATWO.

*Key words:* Ovarian cysts; Adnexal torsion; Wölffian ducts; Female infertility.

## Introduction

A female adnexal tumor of probable Wölffian origin (FATWO) is a rare, poorly understood lesion first described in 1973 by Kari and Scully, which originates from the remnant of the mesonephric duct [1]. It mainly originates from a broad ligament but can arise in other sites with remnants of a Wölffian duct such as the ovaries, mesosalpinx, fallopian tube, and peritoneum. The mesonephric duct is a paired organ that is divided in the male reproductive system. In females, owing to the absence of an anti-Müllerian hormone, the mesonephric duct degenerates and forms the broad ligament, lateral walls of the cervix, vagina, and uterine corpus. The mean age at diagnosis is approximately 50-years-old. However, FATWOs have been diagnosed in women 18 to 81 years in age [2] [3]. The preoperative diagnosis of a FATWO is very difficult owing to the rarity of the disease and the limited literature available. Positive immunoreactivity to pancytokeratin (CK), CAM 5.2 CK antibody, CK7, and vimentin, and negative immunoreactivity to estrogen receptor (ER), progesterone receptor (PR), epithelial membrane antigen (EMA), and CD20 may contribute to FATWO carcinogenesis [4, 5]. Although most cases follow a benign clinical course, they have the potential to recur or metastasize. Herein, the authors report the case of a 30-year-old woman with a combination of acute ovarian torsion and the rare ovarian neoplasm of FATWO arising from the right tubal fimbria which was treated laparoscopically while preserving her fertility.

## Case Report

A 30-year-old woman, gravida 0 para 0, without prior medical or surgical history came to the emergency unit of Pusan National University's Yangsan Hospital with an abrupt daylong onset of lower abdominal pain. She presented with lower abdomen pain with watery diarrhea and had tenderness in her right infra-umbilical region without rebound tenderness. A gynecological examination revealed right adnexal tenderness with an absence of abnormal cervical discharge. Blood samples were collected for laboratory tests. Her white blood cell count was 12,400/mm$^3$ with 8% bands and C-reactive protein (CRP), 1.06 mg/dL. Function tests of her pancreas and liver showed normality. A pelvic transvaginal ultrasound revealed a normal sized uterus, with a thin and regular endometrial layer. The left ovary was normal, but the right ovary was enlarged with a 4.0×3.0-cm sized cystic mass. The authors took an enhanced abdominal and pelvic computed tomography scan. There was a thin walled smoothly emarginated multi-locular cystic mass and twisted pedicle in her right adnexa, continuous with the cystic mass, apart from the ovary.

The levels of serums CA 19-9 and CA 125 were 29.7 U/mL (0-35 U/mL) and 31.0 U/mL (0-35 U/mL), respectively. The patient underwent emergent explorative laparoscopy. Upon entering the pelvic cavity, the laparoscopy showed a dilated and torqued mass in the right adnexa. The mass was 4 cm in size and twisted clockwise three times (Figure 1). The mass was removed, and a frozen biopsy was conducted. When the frozen biopsy proved negative for malignancy, a right salpingectomy was applied. The postoperative period was uneventful, and the patient was discharged on day 3 after surgery. An anatomic study revealed that the tumor measured 29.3 grams in weight and was 4.1×3.0×1.9 cm in dimensions. A pathological examination identified a sieve-like pattern, with round to oval hyperchromatic nuclei. Immunohistochemistry was performed for inhibin (Figure 2A), WT1 (Figure 2B), calretinin, CK7, CK20, ER, PR, and PAX8. The tumor

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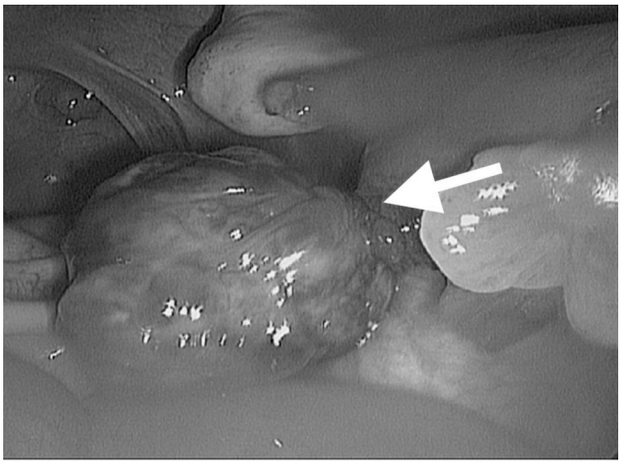
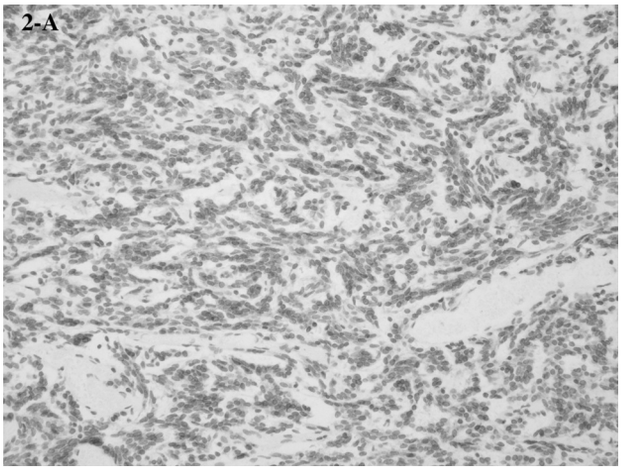


Figure 1. — Explorative laparoscopic finding shows an approximate 4cm-sized right adnexal mass with torsion. The white arrow indicates the direct the knot which was twisted by 1,080 degrees.



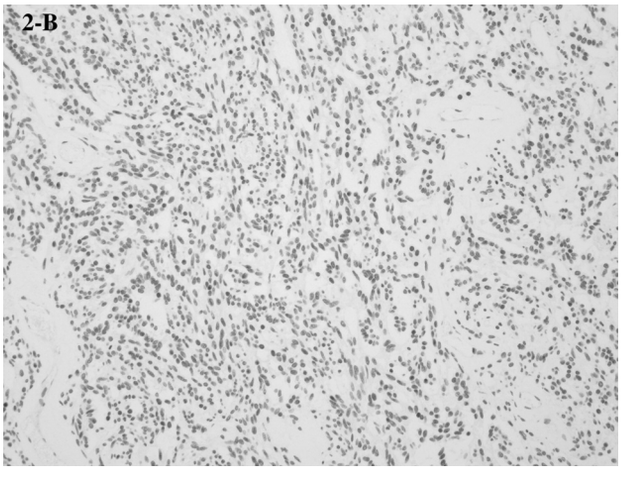




Figure 2. — Immunohistochemistry study of the mass. 2A: Represents positive staining of tumor cells for inhibin and 2B for WT-1.

cells were positive for inhibin and focal positive for WT1, but negative for calretinin, CK7, CK20, ER, PR, and PAX8. Based on pathological and immunohistochemical findings, a FATWO diagnosis was confirmed. The patient was warned about the risk of malignancy of this tumor and was recommended to undergo a total abdominal hysterectomy and bilateral salpingo-oophorectomy. However, the patient wanted to preserve her fertility and received a close follow up, which included a clinical examination, tumor markers, abdominal and pelvic ultrasound, and an MRI. Five years after surgery she remains asymptomatic. No evidence of recurrence was observed throughout this period.

## Discussion

A FATWO is a rare disease that can originate with persisting remnants of the mesonephric duct from the ovary, broad ligament, mesosalpinx, ovarian hilus, pelvis, paratubal site, and paravaginal area (2). A FATWO originates from the embryologically remnants of the mesonephric duct, also known as the Wölffian duct, which degenerates in females (3). A FATWO can occur anywhere in sites originating from the Wölffian duct. The sites of origin described in the literature are the ovary, broad ligament, mesosalpinx, ovarian hilus, pelvis, paratubal site, and paravaginal area. Through the many sites of origin, paratubal origin is rare, and usually asymptomatic. However, in such cases, the patients present abdominal pain owing to a torsion of the paratubal mass. As in the present case, FATWOs generally show a benign behavior. According to a previous study, however, one-fifth of FATWO cases show malignant outcomes such as a metastasis or recurrence [6].

The rarity and variable location of a FATWO make its diagnosis difficult. Usually, ultrasonographic findings show a pelvic semisolid mass with high vascularization. A slightly hyper-intense adnexal mass with cystic degeneration is a common MRI finding of this tumor, and it is difficult to differentiate its nature from a subserosal leiomyoma or ovarian cancer [7]. The pre-operational serum CA-125 levels of all patients have been normal. In the present case, the patient was admitted to the emergency unit and underwent ultrasonography and a CT scan. The putative diagnosis was ovarian cyst torsion, and surgery was conducted accordingly. The serum CA-125 of the patient was also within the normal range.

The diagnosis of a FATWO depends largely on its histological features. FATWOs have been characterized microscopically as having a solid, diffuse, sieve-like tubular arrangement of the spindle or epithelial cells, retiform, a multicystic pattern or closed packed tubules, and uniform round or oval-shaped nuclei [8]. According to one report, tumor cells are positive for AE1/3, CK1, CAM 5.2, CK7, and 20, high molecular weight cytokeratin, keratin 903, epithelial membrane antigen (EMA), ER, PR, androgen receptor, inhibin, calretinin, and vimentin [9], but negative for monoclonal carcinoembryonic antigen, cytokeratin 20, Ki-67, and KIT protein [5]. In this case, however, the tumor

cells were positive for inhibin and focal positive for WT1, but negative for calretinin, CK7, CK20, ER, PR, and PAX8.

Because there have been few cases of malignancy, pathologic or immunohistologic findings of potentially malignant tumors are currently under study. FATWOs with high mitotic activity, cellular atypia, and necrosis usually behave in an aggressive manner [10, 11]. These findings are good indicators of being malignant, although a FATWO with very low mitotic activity and minimal nuclear atypia may also occur. Recurrence can take place in the absence of aggressive histological findings, and even after several years following the initial diagnosis. In this case, no such findings occurred, and there has been no recurrence or metastasis after surgery.

In general, a patient will show a recurrence or metastasis more than six years after diagnosis, or after living with the disease for a long period of time [6]. Recurrence and metastasis can occur within two years, and are found in 11-20% of cases. In FATWO cases, the liver and lungs are the most common organs for metastasis [11-13]. According to the literature, recurrence usually occurs between 13 and 96 months, with a median period of 48 months. Recurrence will occasionally be found even after a long period of time after diagnosis. In this case, the patient underwent surgery five years ago, has remained asymptomatic, and has shown no problems suggesting a recurrence.

There are three main differential diagnoses with a FATWO: well-differentiated endometrioid adenocarcinoma, Sertoli-Leydig cell tumors, and sex-cord stromal tumors. A FATWO and endometrioid adenocarcinoma occur in different places [8]. A FATWO occurs in the broad ligament and ovaries outside the fallopian tube, whereas endometrioid adenocarcinoma occurs in the fallopian tubes. In comparison with a FATWO, endometrioid adenocarcinoma has a more pronounced degree of nuclear atypia and mitotic activity. In addition, endometrioid adenocarcinoma shows immunoreactivity to whole cytokeratin, WT1, and EMA. Only a-inhibin appears negative in endometrioid adenocarcinoma [2]. Although a FATWO and Sertoli-Leydig cell tumor are morphologically similar, a FATWO tends to not exhibit endocrine symptoms, which are characteristic of Sertoli-Leydig cell tumors. In addition, unlike a FATWO, Sertoli-Leydig cell tumors have not been reported in a paratubal site or in the broad ligament. In a FATWO, a-inhibin is localized positive, whereas in a sex-cord stromal tumor, a-inhibin is generally positive. CK7 is generally absent in sex-cord stromal tumors, and AE1/3 is positive in 30-37% of cases [7].

A complete resection is considered the best treatment. In general, a total hysterectomy with bilateral salpingo-oophorectomy is recommended for a malignant FATWO. Most patients with recurrent tumors undergo only a tumor resection [12]. Despite these facts, a tumor resection alone is a suitable treatment. In this case, only pelviscopic paratubal mass detorsion and excision surgery were performed to maintain the patient's fertility.

Because of the rarity of occurrence, no definitive chemotherapy methods are currently available for dealing with a FATWO. A variety of studies are underway in the fields of radiotherapy, chemotherapy, and molecular-targeting therapy. Various chemotherapeutic methods for treating metastatic and recurrent FATWOs, such as cisplatin/cyclophosphamide, etoposide/ifosfamide/carboplatin, cisplatin/paclitaxel, carboplatin/paclitaxel with leuprolide, and irinotecan alone, have been used in clinical practice. Among them, paclitaxel and carboplatin are the most applied methods [6]. Only a combination of paclitaxel and cisplatin can temporarily stabilize the disease. Gleevec has been administered in some patients who were c-kit positive [5]. The effects of adjuvant chemotherapy or radiation therapy are still controversial. Although there are insufficient data on the use of Gleevec for the treatment of these types of tumors, it is worth considering the use of target molecular therapy for a recurrent, C-kit positive, FATWO. In this case, adjuvant therapy was not used to preserve the patient's fertility [14].

In conclusion, the patient underwent emergent exploratory laparoscopic surgery because of the torsion of the FATWO. The tumor was removed through surgery, and a frozen biopsy was conducted. When the frozen biopsy showed that the tumor was negative for malignancy, a right salpingectomy was performed. The patient was informed of the risk of malignancy of the tumor and was advised to undergo a total abdominal hysterectomy and bilateral salphingo-oophorectomy. The patient received a close follow-up to maintain her fertility.

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