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Rare Tumors of Wolffian Duct Origin: Clinical Insights from a FATWO Case and Literature Overview

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Abstract

Female Adnexal Tumor of Probable Wolffian Origin (FATWO) is a rare neoplasm of the female reproductive system arising from embryonic mesonephric duct remnants. Due to its low incidence and nonspecific histological patterns, FATWO poses a diagnostic challenge and is often misclassified. There is a lack of standardized diagnostic criteria, and optimal treatment strategies remain unclear. We report the case of a 57-year-old postmenopausal woman with an incidental finding of a pelvic mass on routine ultrasound. Her clinical history included thyroidectomy, appendectomy, and myomectomy. Laboratory findings were within normal limits. Imaging revealed a right adnexal mass, and intraoperative assessment showed additional tumor implants on the peritoneum, mesentery, and omentum. Complete cytoreductive surgery was performed, including hysterectomy with bilateral salpingo-oophorectomy, pelvic peritonectomy, and omentectomy. Histopathological evaluation revealed an epithelial neoplasm with tubular and reticular growth patterns and minimal cytological atypia. Immunohistochemical analysis excluded other common epithelial tumors. Based on morphology, immunophenotype and tumor location, a diagnosis of FATWO was confirmed.

This case underscores the importance of considering FATWO in the differential diagnosis of adnexal masses, especially in patients with unusual tumor localization and morphology. Accurate diagnosis relies on a combination of histopathologic evaluation and immunoprofiling. Given the potential for recurrence and metastasis in rare malignant cases, long-term follow-up is essential. This study did not require trial registration. No external funding was received.

Keywords: FATWO; Wolffian duct remnants; rare adnexal tumor; mesonephric tumor; gynecologic oncology; immunohistochemistry

1. Introduction

The term Female Adnexal Tumor of Probable Wolffian Origin (FATWO) was first introduced by Kariminejad and Scully in 1973 [1]. Later studies supported the Wolffian duct origin of these tumors using immunohistochemical and electron microscopy methods [2]. Although mitotic activity and capsular invasion were observed, the tumor was initially classified as benign [2].

FATWO is an extremely rare neoplasm of the female reproductive tract, thought to arise from remnants of the mesonephric (Wolffian) duct-embryological structures that are anatomically independent from Müllerian derivatives. These remnants are most commonly located in the broad ligament but can also be found in the mesosalpinx, fallopian tubes, ovaries, or omentum [2,3]. In most cases, FATWO is asymptomatic and discovered incidentally during imaging or surgical procedures performed for unrelated reasons. Some patients, however, may present with abdominal pain or a palpable mass [3]. The age of onset ranges from 15 to 83 years, with a median age of diagnosis around 50 [3]. Although the majority of FATWO cases exhibit benign clinical behavior, approximately 11% may show disease progression within two years of diagnosis [4].

Recurrent or metastatic behavior has been documented, suggesting a low malignant potential in some cases [4]. To date, no standardized treatment protocols exist for recurrent or metastatic FATWO [2]. This study presents a rare clinical case of FATWO and aims to contribute to the limited body of literature regarding its diagnostic challenges and management approaches.

2.Materials and Methods

Patient Information

A 57-year-old non-smoking, married female was referred to a specialized oncology center following the incidental discovery of a right ovarian tumor during routine evaluation. Her medical history included thyroid resection for nodular goiter, appendectomy, and laparoscopic myomectomy in 2014. The patient was postmenopausal for three years at the time of presentation.

Laboratory Evaluation

All serum tumor markers were within reference ranges: CA-125 - 22.32 U/mL, HCE - 12.35 ng/mL, AFP - 1.57 ng/mL, β-hCG - 6.83 mIU/mL, HE4 - 56.14 pmol/L. The ROMA index was calculated as 9.8% in premenopausal and 16.41% in postmenopausal settings.

Imaging Studies

Multislice computed tomography (MSCT) and pelvic magnetic resonance imaging (MRI) revealed multiple dense implants up to 15 mm along the uterine ligaments, pelvic peritoneum, and hypogastric region (Figure 1). A large cystic-solid mass measuring 102×119×72 mm was visualized separately from the ovaries, with a heterogeneous internal structure and clearly demarcated borders. T2-weighted imaging demonstrated mixed high- and low-intensity signal regions (Figure 2).

Surgical Procedure

The patient underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy, pelvic peritonectomy, omentectomy, and resection of metastatic lesions from the sigmoid mesentery and

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anterior abdominal wall. Intraoperatively, the uterus was normal in shape and measured 8×6×5 cm; adnexa appeared macroscopically unremarkable. A retroperitoneally extending tumor-like mass measuring 12×10×7 cm was identified anterior to the uterus and bladder. Additional nodules were detected in the Douglas pouch (3×4 cm), on pelvic peritoneum, sigmoid mesentery (up to 3 cm), anterior abdominal wall, and greater omentum (up to 1 cm) (Figure 3). Complete primary cytoreduction was achieved.

Macroscopic Pathology

The tumor had an uneven, lobulated surface and a cystic-solid consistency. Hemorrhagic fluid was present in cystic spaces, while the solid areas were grey-white to grey-yellow (Figure 4).

Histopathological Analysis

Histological examination of the main tumor and peritoneal, mesenteric, omental, and abdominal wall deposits revealed a uniform morphology. The tumor was composed of tubular structures with slit-like lumina, along with trabecular and solid growth patterns. The stroma was composed of hyalinized and sclerotic connective tissue (Figure 5). Tumor borders were well-defined, with no evidence of infiltrative growth. The ovarian parenchyma was uninvolved, and the tumor was separated from it by a thin fibrous pseudocapsule (Figure 6).

Cytology and Immunohistochemistry

Tumor cells exhibited minimal cytological atypia and were predominantly cuboidal to columnar in shape. Some elongated cells had indistinct cell borders. Cytoplasm was eosinophilic or vacuolated; nuclei were round to oval with smooth contours and fine chromatin. Nucleoli were inconspicuous, and mitotic figures were not observed (Figure 7).

Immunohistochemical profiling revealed strong, diffuse expression of vimentin, pancytokeratin, and WT1; focal weak expression of α-inhibin and calretinin; and nuclear progesterone receptor expression in isolated cells (Figure 8). Tumor cells were negative for CK7, CK5/6, CK20, PAX8, CD10, CD117, EMA, ER, GATA3, EpCAM, D2-40, HMW, CD34, CD99, CD56, chromogranin A, synaptophysin, Melan-A, and β-catenin. The Ki-67 proliferation index was <1% (Figure 9).

Additional Findings

Examination of the uterus revealed endometrial glandular hyperplasia without atypia and an uterine wall intramural leiomyoma. Pelvic lymph nodes were free of metastases but contained areas of endosalpingiosis. Both fallopian tubes and one ovary were not involved by the tumor.

3. Results

Complete cytoreduction was achieved intraoperatively. Imaging identified a large extraovarian mass and multiple small peritoneal and mesenteric implants. Histologically, the tumor demonstrated well-differentiated epithelial architecture with tubular and trabecular patterns, a low proliferative index, and absence of high-grade atypia. Immunohistochemistry supported an epithelial origin not related to Müllerian structures (PAX8-negative) and most consistent with a tumor of probable Wolffian origin (FATWO).

Figure 1: *CT- imagination.*

a - The arrows indicate solid implants with a maximum size of up to 15mm along the ligaments of the uterus, pelvic peritoneum and hypogastrium; b - The arrow shows a cystic-solid tumor conglomerate in size 102*119*72 mm.

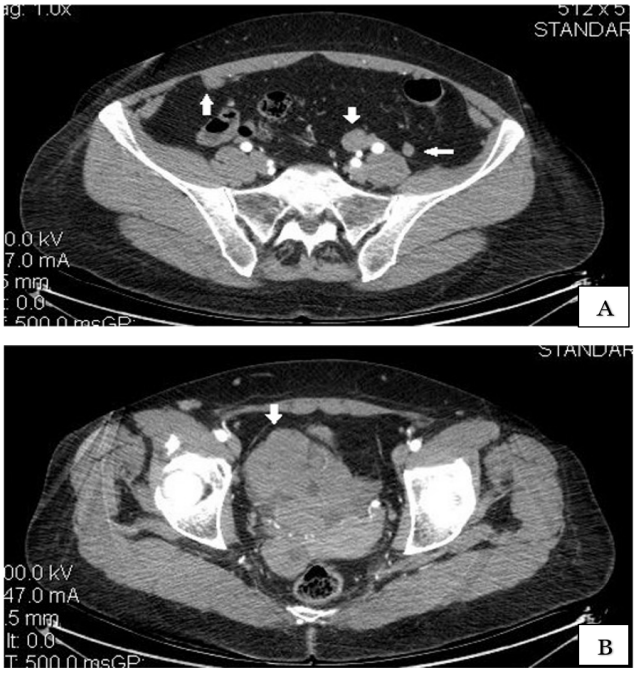


Figure 2: *The T2-weighted magnetic resonance imaging image visualizes a clearly limited oval formation characterized by a heterogeneous structure with areas of both increased and decreased signal intensity, located in anatomical areas not associated with ovarian tissues.*

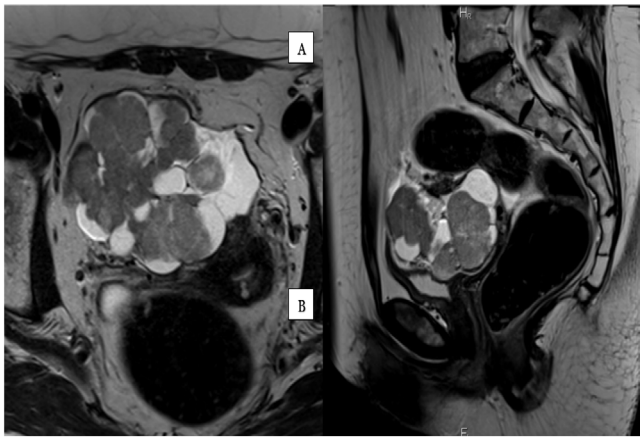


Figure 3: *The arrows indicate single nodes on the mesentery of the sigmoid colon and in the large omentum.*

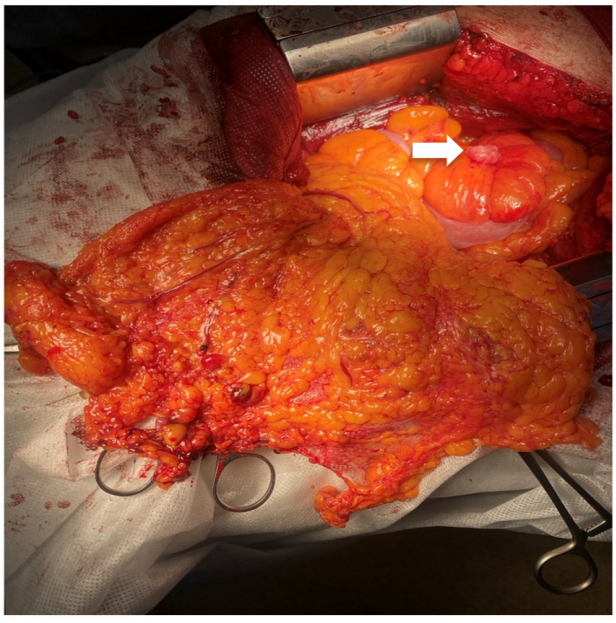


Figure 4: *Intraoperative image of the tumor.*

Figure 4: *Macroscopic image of the removed preparation: the tumor is characterized by a bumpy surface, partially cystic structure with areas of hemorrhage; dense gray-white and yellowish tissues corresponding to histologically heterogeneous components of the neoplasm are visualized on the incision.*

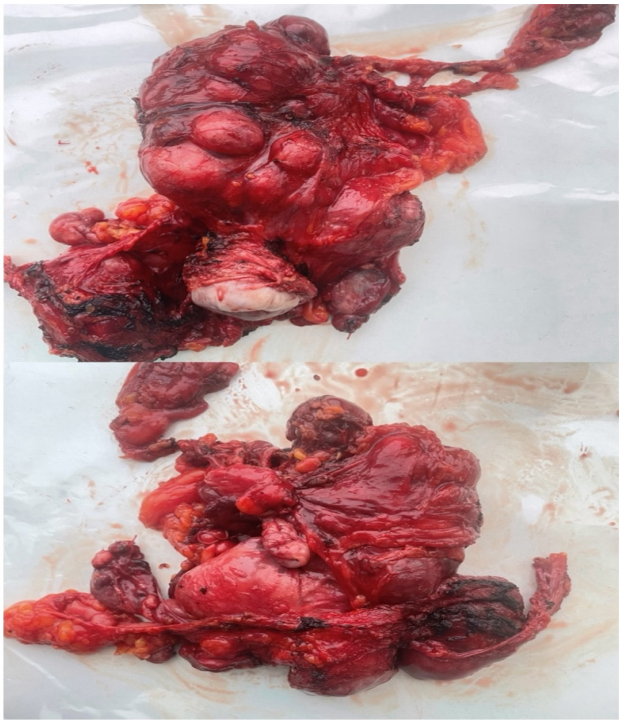


Figure 5: *Microscopic view of the tumor (scanning magnification, x5): areas of tubular and trabecular growth patterns.*

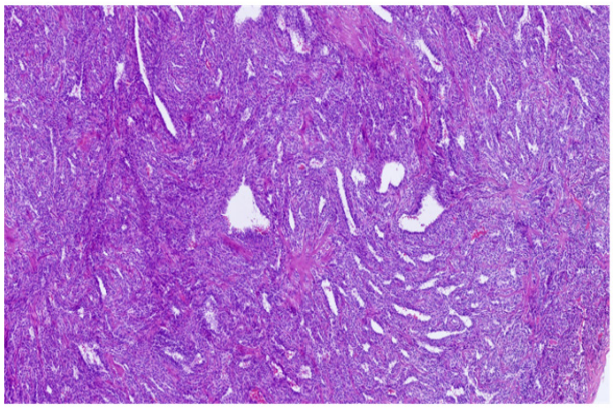


Figure 6: *Fibrous pseudocapcule detached the ovary parenchyma from the tumor.*

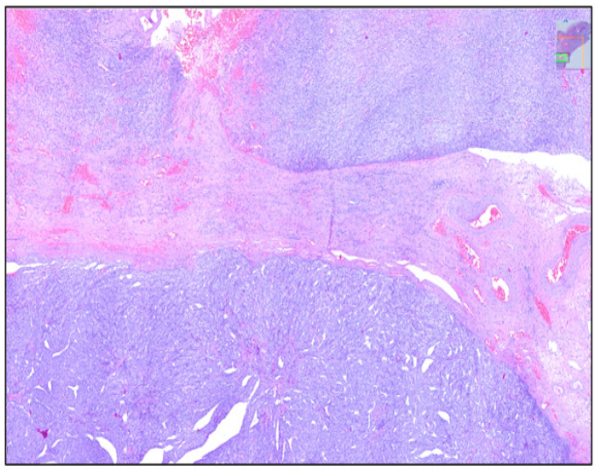


Figure 7: *Microscopic view of the tumour (at x20 magnification): cuboidal tumor cells with ovoid, monomorphic nuclei with dispersed chromatin and barely discernible nucleoli.*

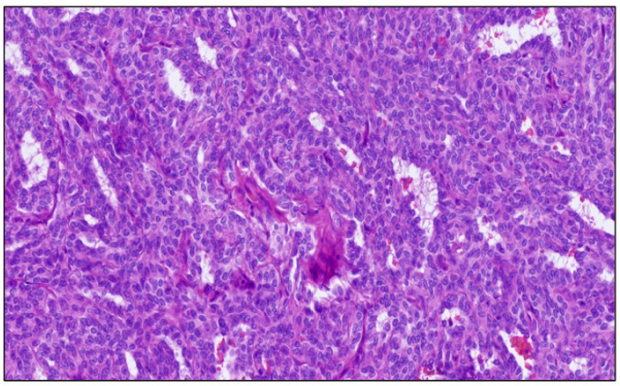


Figure 8: *IHC-study results: tumor cells show expression of Vimentin, PanCK, WT1, a-Inhibin and Calretinin (weak staining), Pr (focally).*

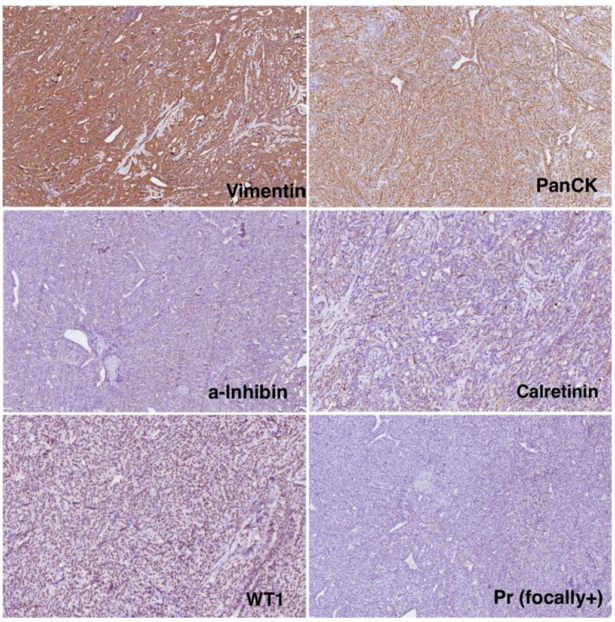
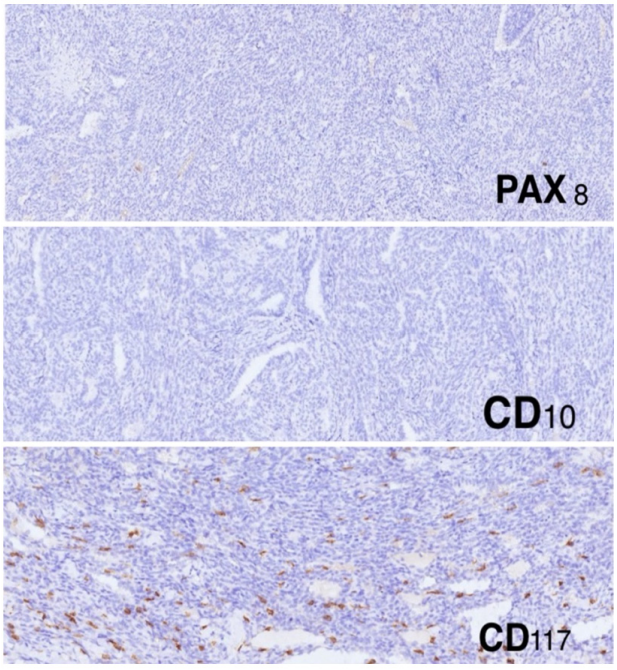


Figure 9: *IHC-study results: tumor cells show no expression of PAX8, CD10 and CD117.*



4. Discussion

Female adnexal tumors of probable Wolffian origin (FATWO) are rare neoplasms derived from remnants of the mesonephric duct, generally considered to have low malignant potential. However, a subset of cases with aggressive clinical behavior has been described [5]. Most FATWOs behave in a benign manner and do not recur following surgical resection, though approximately 20% demonstrate malignant progression [6,7], with a mean time to recurrence of about 48 months (range: 7-96 months). Recurrences and metastases have been documented in 11% of patients within the first two years after diagnosis [3,8-10].

FATWOs develop from regressed mesonephric structures, and the underlying molecular mechanisms remain poorly understood. Reported gene alterations include mutations in STK11, APC, and MDB4, as well as variants in KMT2D [6], although the clinical significance of these findings remains unclear.

Clinically, FATWOs typically present unilaterally and arise in the broad ligament or from adnexal tissue on a narrow stalk. To date, no specific immunohistochemical markers exist for unequivocal diagnosis. A major diagnostic challenge is distinguishing FATWO with tubular growth patterns from low-grade endometrioid carcinoma. The latter usually arises from endometriotic lesions and shows a cribriform glandular pattern with frequent squamous or morular metaplasia, the presence of secretion in glandular lumens, tumor cells with basally located nuclei. Immunohistochemically, endometrioid carcinoma expresses PAX8, ER, and PR [7].

The differential diagnosis also includes mesonephric-like adenocarcinoma, sex cord-stromal tumors such as Sertoli-Leydig cell tumors, and cellular fibromas with spindle-cell morphology. Mesonephric-like adenocarcinoma typically expresses GATA-3 and PAX8, but not ER or PR. Sertoli-Leydig tumors feature tubular arrangements of Sertoli cells and Leydig cell clusters with Reinke crystals, and are usually negative for cytokeratins but positive for α-inhibin and calretinin [7]. Recurrences may occur many years post-diagnosis, even in histologically benign-appearing tumors. Malignant FATWOs are characterized by extra-organ spread, recurrence, and distant metastases—commonly to the liver and lungs [11]. Notably, recurrences tend to occur in patients who underwent local excision only [12]. Importantly, clinical behavior often does not correlate with histomorphologic features [13].

Features associated with aggressive behavior include necrosis, capsular invasion, high mitotic activity, marked nuclear pleomorphism, CD117 positivity, and a high Ki-67 index [14]. Due to the extreme rarity of FATWO, standardized treatment guidelines are lacking. Radical surgery, including hysterectomy, bilateral salpingo-oophorectomy, and complete cytoreduction, is currently the most effective primary approach. Recurrence is more frequent in patients managed with fertility-sparing or localized surgeries such as cystectomy or mass excision [3,13,14].

While sporadic reports of partial responses to adjuvant therapies exist, the efficacy of radiation, chemotherapy, hormonal, and targeted treatments remains uncertain in malignant FATWO. A personalized treatment approach is thus critical in cases with aggressive clinical behavior. Chemotherapeutic regimens for recurrent and metastatic FATWO have included various combinations, most notably paclitaxel plus carboplatin. Tyrosine kinase inhibitors like imatinib have also been explored in cases with c-Kit expression [2,3]; however, clinical benefit has been limited, underscoring the need for further studies to identify effective treatment strategies for malignant FATWO.

Limitations and Future Directions

Given the rarity of FATWO, current knowledge is largely based on case reports and small case series. The absence of prospective studies, standardized diagnostic markers, and therapeutic protocols limits the generalizability of findings. Future research should focus on molecular characterization, development of reliable diagnostic biomarkers, and evaluation of targeted therapies in larger, multi-institutional cohorts to improve outcomes for patients with aggressive FATWO.

Broader Implications

This case adds to the growing body of evidence that FATWO, although often indolent, can exhibit malignant potential. Comprehensive surgical management and long-term follow-up remain critical. The development of molecular and immunohistochemical diagnostic tools and targeted treatment options could significantly advance the management of this rare entity.

5. Conclusion

Female adnexal tumor of probable Wolffian origin (FATWO) is an extremely rare gynecologic neoplasm, presumed to arise from mesonephric duct remnants. While most cases exhibit indolent behavior, rare malignant variants have been reported with aggressive clinical progression.

Diagnosis of FATWO requires comprehensive morphological assessment and an extended panel of immunohistochemical markers. To date, there are no validated histological features or serum biomarkers that reliably predict clinical behavior. Therefore, long-term follow-up is strongly recommended for all patients, regardless of the initial histological findings.

Radical surgical excision remains the mainstay of treatment. The role of adjuvant chemotherapy or radiotherapy in managing recurrent or malignant FATWO remains uncertain and is a subject of ongoing debate. A systematic review of existing clinical cases and continuous accumulation of treatment outcome data are essential for developing evidence-based management strategies.

Future research should focus on identifying reliable diagnostic and prognostic markers and evaluating the efficacy of systemic therapies for aggressive disease variants. These steps are critical to improving personalized care and long-term outcomes for patients with this rare tumor type.

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